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Case reports in aortic surgery and endovascular repair: 2023

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Case report: Surgery combined with extracorporeal membrane oxygenation for a patient with type A aortic dissection complicated with myocardial infarction after percutaneous coronary intervention

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Background: Aortic dissection (AD) is a severe cardiovascular disease characterized by aortic rupture, aortic valve insufficiency, aortic branch lumen stenosis, and occlusion. Acute ST-segment elevation myocardial infarction may be the primary manifestation when aortic dissection affects the coronary artery, leading to delayed or missed diagnosis of aortic dissection, and preventing patients from receiving timely and comprehensive treatment. Simultaneous aortic repair and coronary artery bypass grafting surgery are controversial because of their high mortality rates. Personalized and optimal treatment plans for patients should be taken seriously based on their different conditions and treatment options.

Case presentation: A 42-year-old man who experienced 1 h of persistent precordialgia was admitted to a local second-level hospital for emergency treatment. Electrocardiogram (ECG) showed evidence of ST-segment elevation, and myocardial enzyme levels were CK-MB 18.35 ng/ml and troponin 0.42 ng/ml. The patient was treated for acute myocardial infarction (AMI) and urgently sent to the interventional catheter room. Coronary angiography showed stenosis of the starting part of the right coronary artery trunk. Thus, stent implantation was performed, and the stenosis section recovered patency; however, postoperative precordialgia was not alleviated. Computed tomography angiography (CTA) revealed a type A AD. The patient was immediately transferred to a higher-level hospital, underwent emergency surgery with cardiopulmonary bypass (CPB) ascending aorta replacement, SUN's procedure (total arch replacement and stented elephant trunk implantation), and simultaneous implantation of extracorporeal membrane oxygenation (ECMO), and regained consciousness within intensive care unit care. ECMO was discontinued when hemodynamics stabilized. The patient ultimately recovered well and was discharged.

Abbreviations
AD, aortic dissection; AMI, acute myocardial infarction; CTA, computed tomography angiography; CPB, cardiopulmonary bypass; SUN's procedure, total arch replacement and stented elephant trunk implantation; ECMO, extracorporeal membrane oxygenation; PCI, percutaneous coronary intervention; CABG, coronary artery bypass grafting.

Conclusion: This case demonstrated that precordialgia is not limited to myocardial infarction but may also be accompanied by aortic dissection. Percutaneous coronary intervention (PCI) can timely and effectively restore coronary artery perfusion, strive for the opportunity of aortic repair surgery, and can overcome pump failure caused by myocardial infarction, cardiopulmonary bypass, heart block time, and myocardial ischemia-reperfusion injury. Personalized treatment is crucial for patients with complex type A aortic dissection.

KEYWORDS

case report, aortic dissection, acute myocardial infarction, percutaneous coronary intervention, SUN's procedure, extracorporeal membrane oxygenation

Introduction

Acute type A AD is a severe cardiovascular disease that may present with aortic rupture, aortic branch lumen stenosis, or occlusion. Hypoperfusion of the aortic branches caused by dissection of the false lumen can occur in all branches from the coronary artery opening to the abdominal aortic bifurcation (1). If the coronary artery is affected by acute type A AD, AMI may be the primary manifestation, and AD may be diagnosed late or go undetected if not fully considered (2).

Therapeutic methods for patients with AD complicated by AMI remain controversial. If a patient undergoes a one-stage surgery, including aortic repair and coronary artery bypass grafting (CABG), the mortality rate is often extremely high because of a severe setback of cardiac function, including existing myocardial infarction, CPB, heart block time, and myocardial ischemia-reperfusion injury. Owing to the high mortality rate, some scholars believe that implementing PCI treatment first is beneficial for such patients because it allows them to undergo second-stage aortic repair surgery and can reduce the mortality rate (3–5).

The etiology of acute chest pain in patients is not only limited to AMI but also includes the possibility of AD leading to coronary artery involvement and AMI (6). Early diagnosis can provide comprehensive treatment guidance. Notably, myocardial infarction may lead to heart pump failure. If a patient needs to undergo CPB aortic repair surgery again, the myocardial damage will undoubtedly worsen, leading to the inability to detach from the CPB. ECMO should be an indispensable treatment to maintain effective circulation and allow time for myocardial recovery.

In this article, we report a rare case of acute chest pain in a patient who was initially diagnosed with AMI and underwent PCI; however, the diagnosis of AD was delayed. After AD was confirmed, emergency aortic repair surgery was performed, and ECMO was implanted owing to heart pump failure. Based on relevant treatment experiences and a comprehensive literature review, we aimed to elucidate the clinical diagnosis and treatment of this unique and complex combined disease.

Case description

A 42-year-old man (height 170 cm, weight 75 kg) who experienced severe precordialgia, which persisted for 1 h, was admitted to a local second-level hospital for emergency treatment. Although he had a history of hypertension for 5 years, he did not take antihypertensive drugs according to the advice of doctors. He had no trauma, surgery, or family history. There was no apparent cause of the sudden precordialgia. His blood pressure was 115/76 mmHg, and his heart rate was 105 bpm at admission. ECG showed ST-segment elevation (II/III/aVF) (**Figure 1A**), and myocardial enzyme levels were CK-MB 18.35 ng/ml and troponin 0.42 ng/ml. No scheduled cardiac ultrasound examination was performed, and the patient was treated for acute ST-segment elevation myocardial infarction, received 300 mg of aspirin and 180 mg of ticagrelor, and was immediately sent to the interventional catheter room. Coronary angiography showed severe stenosis of the starting part of the right coronary artery trunk (**Figure 1B**). Subsequently, PCI and coronary stent implantation were performed, as well as stenosis section recovery patency (**Figure 1C**). However, precordialgia was not alleviated after PCI, and no improvement was observed after the tirofiban injection. Moreover, hemodynamics became unstable, oxygenation function decreased, blood pressure dropped to 80/50 mmHg, peripheral blood oxygen saturation was 85%, and the heart rate was130 bpm, leading to mechanical ventilation 22 h after PCI and vasoactive drugs to maintain blood pressure. At this point, clinical physicians began to consider the possibility of AD, and an aortic CTA examination confirmed the diagnosis of type A AD (**Figures 2A–D**). The rupture of the aortic intima was located in the descending thoracic aorta, aortic dissection was reverse-tearing to the ascending aorta, a false lumen was extending to the upper edge of the right coronary artery opening, the hematoma inside the false lumen was very thick, and the true lumen of the ascending aorta became smaller due to compression by the false lumen. Local hospital staff urgently transferred the patient to a higher-level hospital, and emergency surgery was performed, including CPB (right axillary artery perfusion), ascending aorta replacement, and SUN's procedure (total arch replacement and stented elephant trunk implantation). It was difficult to detach the CPB during surgery because of right ventricular failure caused by preoperative myocardial ischemia. Therefore, VA-ECMO (left femoral artery and vein) was simultaneously implanted. The CPB time was 337 min, cardiac arrest time was 132 min, circulatory arrest time was 21 min, and intraoperative minimum temperature was 28°C. Owing to preoperative antiplatelet therapy, coagulation function had been severely affected, posing significant difficulties for surgical hemostasis. Platelets, cryoprecipitate, and fibrinogen were also infused. Fortunately, there was no severe progressive bleeding,

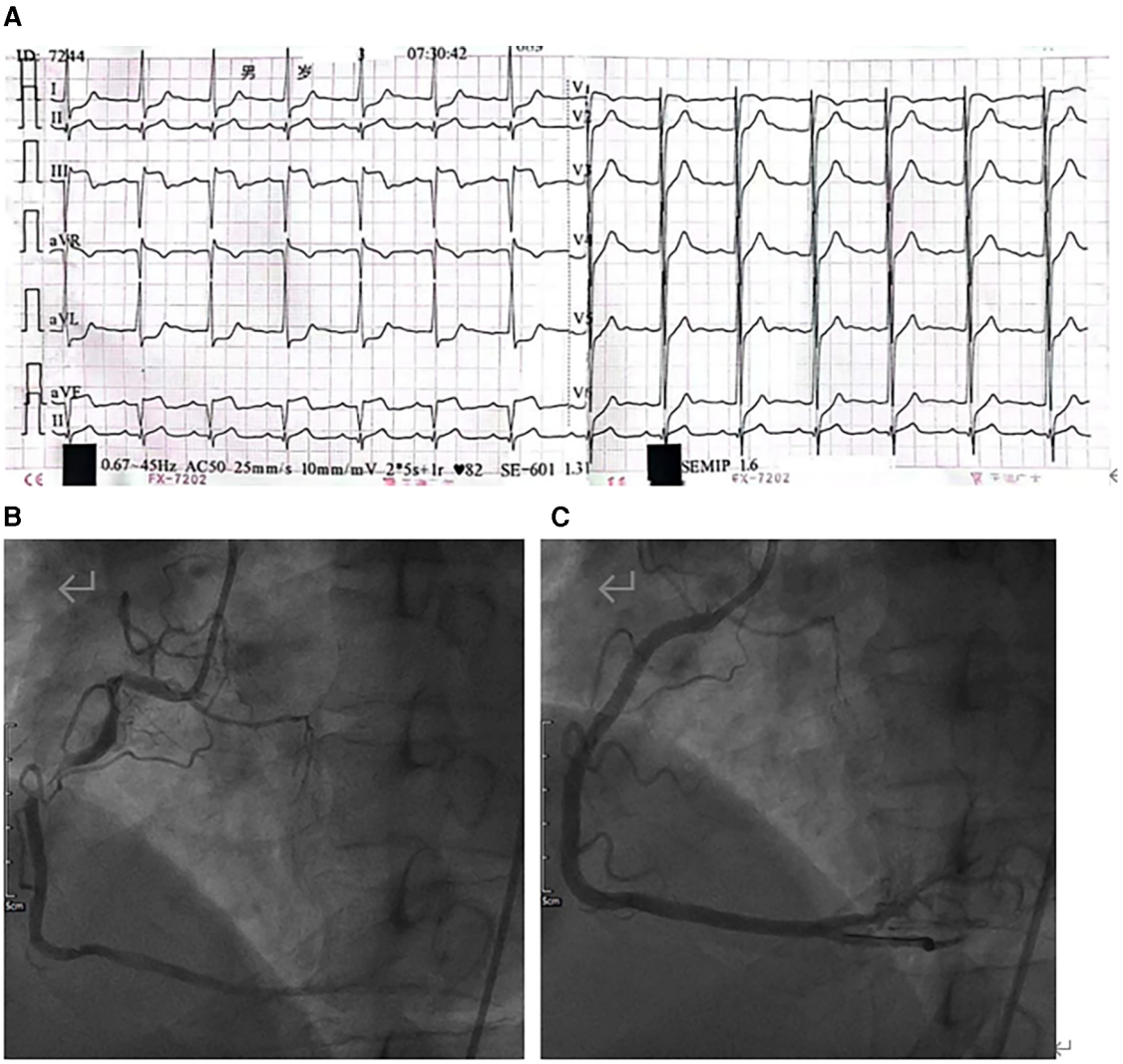

FIGURE 1
(A) Electrocardiogram (ECG) showing ST-segment elevation (II/III/aVF) and inferior acute myocardial infarction. (B) Coronary angiography showing irregular long-segment stenosis of the right coronary artery from the opening to the middle segment. (C) After stent implantation, the right coronary artery shows good morphology and an unobstructed vascular lumen.

and thoracic drainage on POD1 after the surgery was 500 ml. To prevent coronary stent thrombosis after PCI, platelet inhibitors on POD2 were re-administered. The patient regained consciousness on POD4 in the intensive care unit. Cardiac troponin and myocardial enzyme levels continued to decrease after surgery, circulation gradually stabilized, and the dosage of vasoactive drugs gradually decreased. ECMO was discontinued on POD5 when the hemodynamics returned to stability. Tracheotomy was performed on POD10 because of moderate pulmonary infection, and the postoperative mechanical ventilation time was 276 h. The tracheostomy was closed on POD20. Postoperative aortic CTA revealed that the artificial vessels were unobstructed (**Figure 3A**). The thoracic stent was well dilated without internal leakage (**Figure 3B**). The opening of the right coronary artery was unobstructed, the coronary stent implantation was visible (**Figure 3C**), and smooth morphology of the thoracic aorta was observed (**Figures 3D**, **4**). The heart rate and blood pressure of the patient were stable, and heart function was normal through rehabilitation training after 6 months. Moreover, a cardiac ultrasound examination indicated normal cardiac structure and valve function, with a left ventricular ejection fraction of 55%.

Discussion

Acute type A AD can be multifactorial and lead to rupture of the aortic intima and entry of circulating blood into the aortic wall, resulting in the formation of a false lumen. Acute type A AD is the most critical complication of cardiovascular surgery. The mortality rate of acute type A AD increases by 1% every hour within 48 h of onset. Conservative treatment has a high mortality rate of 50% within 2 days. Surgery is the preferred treatment for acute type A AD (7), whereas SUN's procedure is the standard procedure for Stanford type A AD (8).

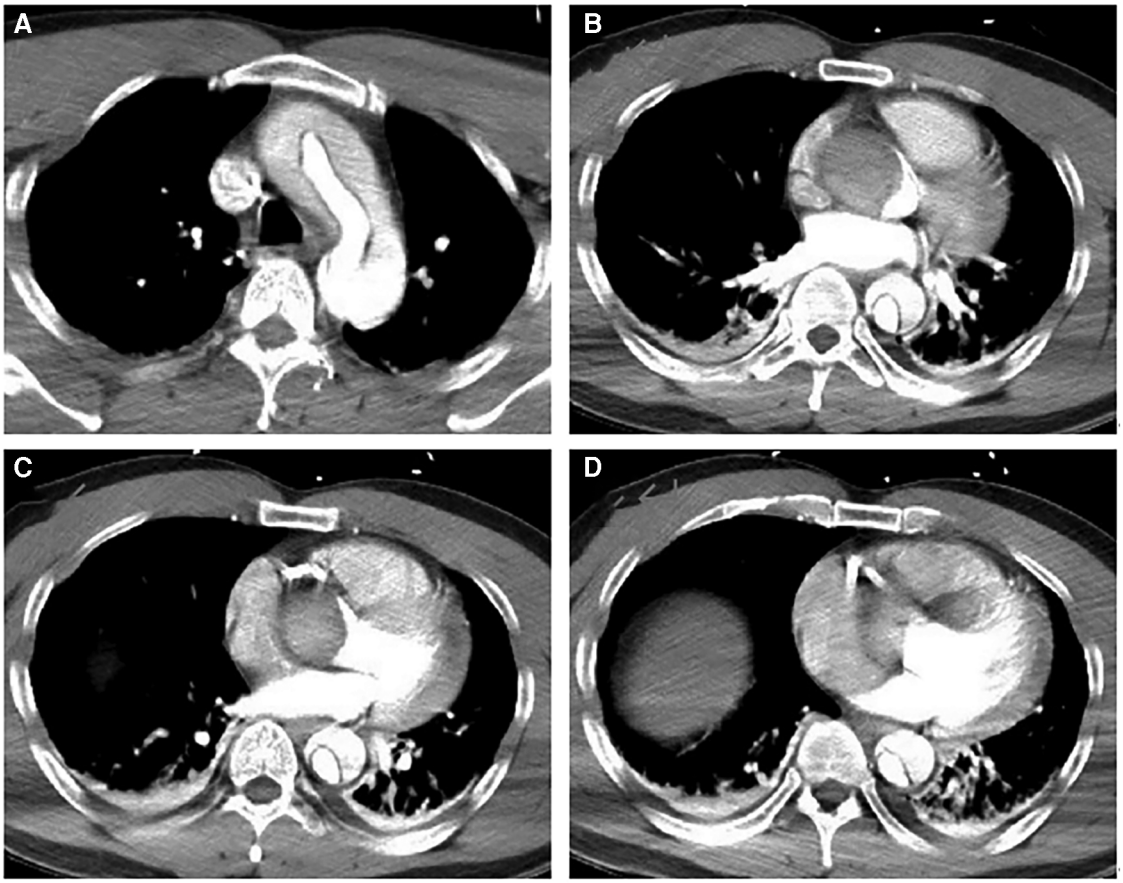

FIGURE 2
Preoperative aortic CTA image. (**A–D**) CTA shows the false lumen of the ascending aorta compressing the true lumen, extending above the opening of the right coronary artery. They indicate a thick hematoma in the false lumen compressing the right coronary artery.

The most common causes of death are dissection rupture, aortic valve insufficiency, ischemia of vital organs caused by aortic branch blood supply occlusion, and organ necrosis (9). The low perfusion of branch vessels is primarily due to the false lumen of the branch vessel intimal tear, which receives most of the blood; therefore, the false lumen expands and compresses the true lumen, resulting in insufficient blood supply to the organs. Acute type A AD can affect almost all aortic branches, from the coronary artery to the femoral artery. The incidence and severity of hypoperfusion differ among the various organs. Poor preoperative organ perfusion is a vital factor that significantly affects the prognosis of patients with acute type A AD (10). Accurate AD classification provides precise guidance on surgical treatment and evidence for disease prognosis (11). However, surgical treatment strategies are controversial when complex AD involves organs, such as the brain or other important branch arteries (12). In patients with AD complicated by AMI, preoperative myocardial ischemia, intraoperative cardiac blockade time, and ischemia-reperfusion injury have multiple impacts on cardiac function. The 5-year mortality rate in patients with AD and myocardial infarction is high, and surgical treatment can help reduce this mortality rate (13). However, concomitant CABG performed in acute type A AD repair surgery is associated with high in-hospital mortality (14–16).

The primary clinical manifestations of AD are persistent chest and back pain, and if complicated with AMI, precordium pain may persist. Acute type A AD is misdiagnosed or delayed because only acute coronary syndrome is considered when chest pain occurs. Therefore, the possibility of patients with AMI having AD should be considered (17–19). The acute coronary syndrome is not limited to simple coronary artery disease. AMI may also be the first manifestation of acute type A AD, which can be rapidly diagnosed through cardiac ultrasound and requires aortic CTA examination if necessary. Point-of-care ultrasound (POCUS) can quickly diagnose type A AD and change treatment strategies in patients with ST-segment elevation myocardial infarction (20). AD with concomitant AMI is prone to misdiagnosis, and transesophageal echocardiography (TTE) and D-dimer examination can help detect AD (21–23). Moreover, surveys have shown that for such patients, if a TTE examination is only performed after antithrombotic treatment, there is a correlation with a high surgery mortality rate (24). During the diagnosis and treatment of this case in a local secondary hospital, a cardiac ultrasound examination was overlooked. Therefore, we believe that even if a patient is diagnosed with simple AMI, a cardiac ultrasound examination should be performed before emergency intervention.

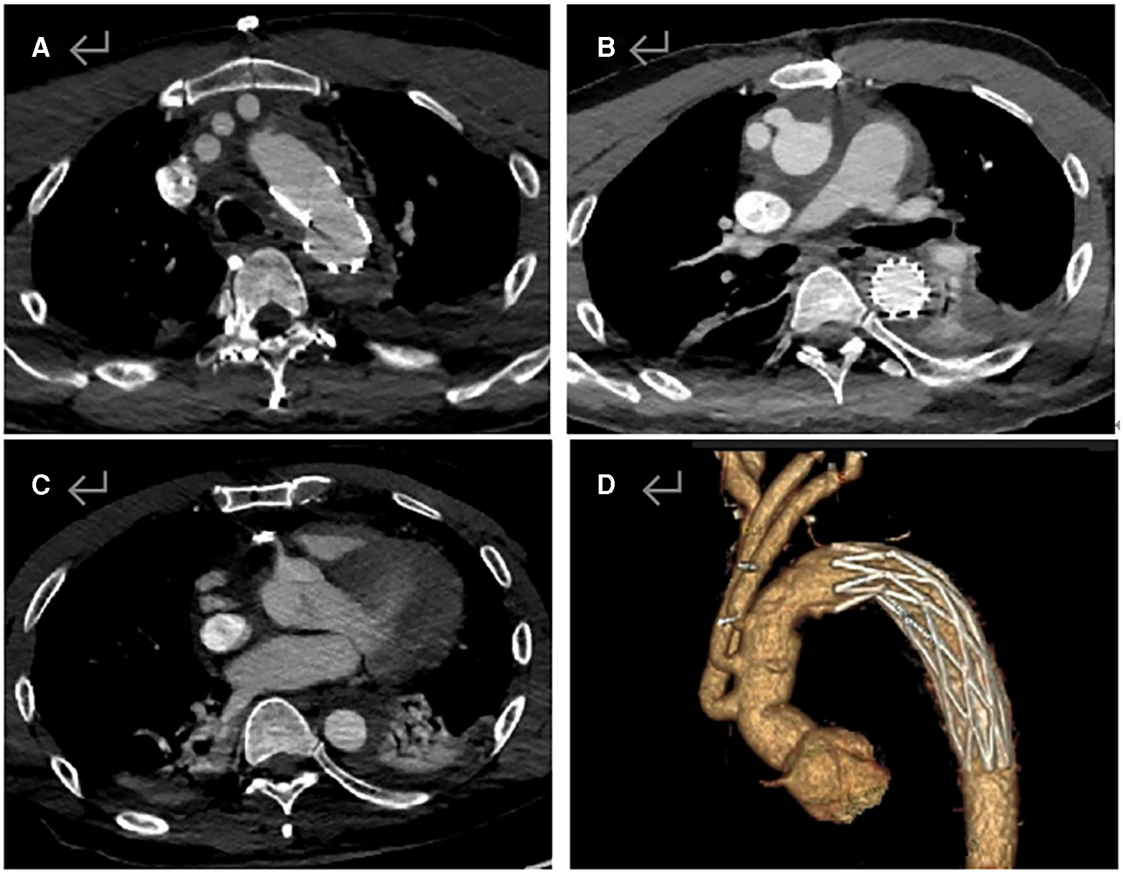

FIGURE 3
Postoperative aortic CTA image. (A) Artificial vessels in the aortic arch and the cephalobrachial artery are unobstructed. (B) Ascending aorta is unobstructed, and the stent is well dilated without internal leakage. (C) Opening of the right coronary artery is unobstructed, and stent implantation is visible. (D) Smooth morphology of the thoracic aorta.

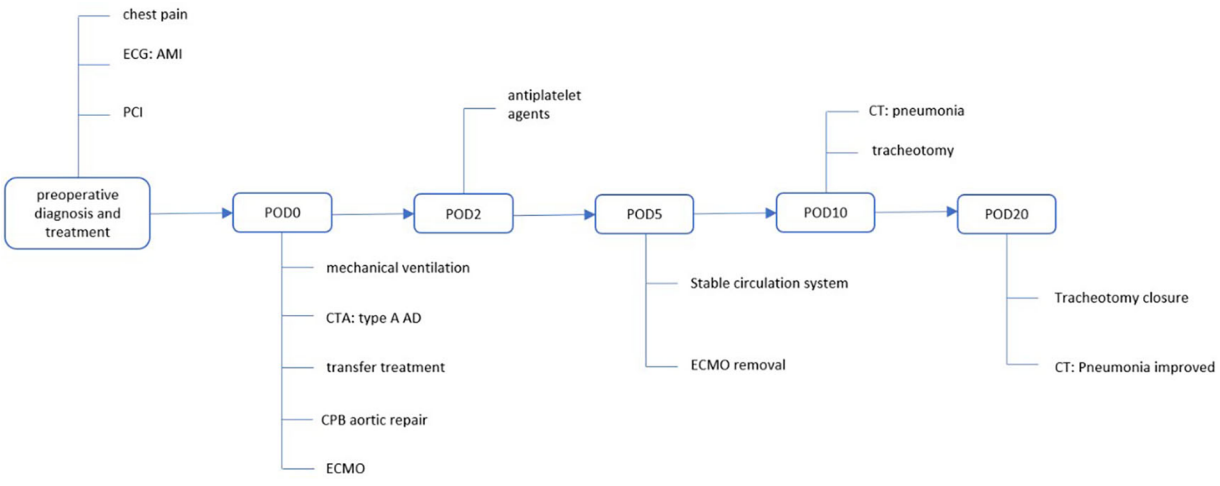

FIGURE 4
Timeline of the case.

In addition, aortic CTA can provide detailed information on the morphology of AD, including the location of intimal rupture and the involvement of branching vessels (25).

If AD diagnosis is missed, coronary angiography and PCI for patients with AMI are understandable, although interventional catheterization carries the risk of aortic rupture. However, if AD

complicated by AMI can be accurately diagnosed simultaneously, PCI can be considered a priority treatment without urgent surgical procedures; some studies have been conducted accordingly (26, 27). The hospitalization mortality rate of patients with AD complicated by coronary artery involvement is as high as 40%. The first-stage PCI treatment strategy is of great help to patients undergoing aortic repair surgery and can reduce the mortality rate of such patients (28, 29). Cho et al. believed that using only guidewires to restore coronary artery perfusion could be an innovative interventional method as the first-stage treatment for coronary malperfusion caused by acute type A AD (30). In this case, although the diagnosis of AD was initially missed at a local secondary hospital, PCI did benefit the patient as it restored the right coronary artery supply and secured opportunities for referral and aortic repair surgery. However, it is equally difficult to determine the interval from PCI to aortic repair because patients with type A AD face the risk of aortic rupture at any time, even if first-stage PCI is successful. Therefore, we believe that aortic repair should be performed immediately after successful PCI. Similarly, not all types of coronary artery involvement caused by type A AD can be corrected using PCI. Some types can only be resolved through surgical repair of both the aorta and coronary arteries or through simultaneous CABG. Kreibich et al. recommended surgical strategies based on different coronary artery lesions, such as CABG for type C lesions and repair of coronary artery openings for type A and B lesions (31–33). Tong et al. reported that repair surgery is still possible even if the opening of the coronary artery intima is torn (34).

Due to the anatomical location of the coronary artery, right coronary artery involvement is more common than left coronary artery involvement in acute type A AD (35). Before undergoing AD center repair surgery, whether PCI is required in patients with concomitant right coronary artery hypoperfusion should be comprehensively evaluated, including concomitant cardiac tamponade, left and right ventricular function, and surgical team efficiency (36). Intraoperative coronary angiography is feasible if hospital conditions permit (37).

Intra-aortic balloon pump treatment is inappropriate because of aortic dissection etiology. ECMO should be used as an essential treatment for cardiac dysfunction to ensure that patients can be weaned from CPB and transferred to the intensive care unit (38, 39). Before VA-ECMO implantation, it is necessary to fully evaluate whether the artery to be intubated is affected by dissection, ensuring that the artery is supplied by the true lumen. Using the axillary artery for arterial catheterization ensures anterograde blood flow to the aorta (40–46). However, in this case, the right axillary artery was intubated for CPB, and the right femoral artery had just undergone a puncture for PCI. Fortunately, the left femoral artery of the patient was not affected by the dissection and was supplied by the true lumen of the aorta. Of course, it was also possible to consider intubating the branch of the aortic artificial vessel, which has the advantage of obtaining anterograde blood flow to the aorta. However, based on previous experience, in patients who had been treated with platelet inhibitors, if ECMO was established through branch intubation of artificial vessels, the impact of ECMO blood flow would lead to severe leakage of blood vessels. Therefore, left femoral artery catheterization was performed to establish ECMO. ECMO can significantly improve the survival rate of patients with cardiogenic shock caused by myocardial infarction or aortic repair surgery (47–53). With improvements in surgical technology and team efficiency, personalized and optimal treatment plans should be provided for patients based on their different conditions and treatment options.

Conclusions

Our case demonstrated that caution regarding possible aortic dissection is required in patients with AMI. It also indicated that although PCI can serve as a transitional treatment if emergency aortic repair surgery cannot be performed, the aortic repair is fundamental, and ECMO must be prepared to overcome pump failure.

Data availability statement

The original contributions presented in the study are included in the article, further inquiries can be directed to the corresponding author.

Ethics statement

Written informed consent was obtained from the patient for the publication of this case report and accompanying images.

Author contributions

JY and WY reviewed the literature and drafted the manuscript. HZ provided the case resources. JG processed the data. JX edited and revised the manuscript. All authors contributed to the article and approved the submitted version.

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Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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Hybrid surgery management challenges of a Behcet's disease patient with recurrence of aortic aneurysms: a case report

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Background: Behcet's disease is a vasculitis of unknown origin that can involve multiple organs or tissues. Aneurysm or pseudoaneurysm, also one of the complications of Behcet's disease, is usually accompanied by a poor prognosis. Surgery is usually accompanied by a high risk of complications, such as the recurrence of anastomotic pseudoaneurysms and blockage of the target vessel. Using hybrid surgery, we successfully treated a complex and recurrent abdominal aortic pseudoaneurysm in a patient with BD.
Methods: We report a 32-year-old female diagnosed with Behcet's disease with recurrent thoracoabdominal aortic aneurysm. Adequate immunotherapy was given during the perioperative period. The splanchnic artery branches were reconstructed, and the aneurysm was sequestered with endovascular repair. The patient recovered uneventfully and was discharged from the hospital 8 days after hybrid surgery. At the 60-month follow-up, no aneurysm was observed, the stent had no displacement or internal leakage, and the reconstructed blood vessels were unobstructed.
Conclusion: Hybrid surgery could be a feasible and effective strategy for BD aneurysms. Adequate preoperative and postoperative immunotherapy with arterial anastomosis away from the diseased artery may be the key to success.

KEYWORDS

behcet's disease, hybrid surgery, recurrent aneurysms, endovascular aneurysm repair, open surgery repair, revascularization, bypass

Introduction

Aortic aneurysms are common in elderly individuals but rare in young individuals and are mostly caused by vascular tissue degradation, trauma, inflammation, infection, fibromuscular dysplasia and Marfan syndrome. Recurrent aneurysms are relatively rare, especially in young people. Recurrent aneurysms have been reported in Behcet's disease (BD) (1, 2), which is a systemic variable vessel vasculitis that involves the skin, mucosa, joints, eyes, arteries, veins, nervous system and the gastrointestinal system. BS mainly occurs in countries located between 30 and 45° north latitude through the Mediterranean basin, the Middle East, and the Far East regions, but it is rare in Europe and North America (3). It is the only primary vasculitis that affects both arteries and veins of any size (4). However, arterial lesions are relatively rare and manifest mostly in the form of aneurysms (5). BS is commonly seen in patients aged 20 and 40 years and has a male preponderance with a male to female ratio of 5.02 in patients with vascular involvement, although the overall patient ratio is 4.4 (6, 7). Medical treatment with cyclophosphamide and corticosteroids is necessary before intervention to repair both aortic and peripheral

artery aneurysms. Surgery or stenting should not be delayed if the patient is symptomatic, according to the 2018 ELUAR recommendations (8).

Treatment for aortic aneurysms involving the visceral arteries is a difficult surgical challenge. Surgery is still the prime treatment for juxtarenal aortic aneurysms (9–11), while in today's minimally invasive era, approximately 80% of patients undergo endovascular aneurysm repair (EVAR) or fenestrated or branched EVAR surgery (12–14). Our case aimed to report hybrid surgery for a patient with recurrent abdominal aneurysm and Behcet's disease and to conduct a literature review.

Case presentation

A 32-year-old female presented with a chief complaint of tearing pain in the back for 2 weeks with persistent worsening. The patient was thin with a malnourished appearance. Physical examination indicated a postoperative scar of the abdomen. Biochemical indexes indicated that serum albumin was 36 g/L, hemoglobin 102 g/L, erythrocyte sedimentation rate (ESR) 35 mm/h, and procalcitonin 0.3 ng/ml had no other abnormalities. Computerized topographic angiography (CTA) reported that a thoracoabdominal aortic aneurysm and an abdominal aortic aneurysm were located in the proximal and distal artificial vessel (5.5 cm and 3.0 cm, respectively, **Figure 1A**, left one), respectively. In addition, she had no smoking or drinking history and no dental or genital ulcer. However, the patient had a very complex and repeated history of aneurysms and surgery.

She had undergone left nephrectomy and renal artery ligation approximately 19 years ago because of a giant left renal aneurysm with a diameter of 10 cm at a local hospital. Unfortunately, 13 years ago, she underwent another open repair for a juxtarenal abdominal aortic aneurysm (6.0*5.3 cm in diameter, **Figure 2A**. **Figure 2B**-postoperative CTA). She was diagnosed with Vasculo-Behcet's disease after the second aneurysm procedure and accepted immunosuppressive therapy. However, 7 months after this operation, the patient experienced epigastric pain and vomiting after eating, which were relieved by symptomatic treatment. Ultimately, the patient developed diffuse peritonitis, and open exploration confirmed small bowel necrosis due to intestinal volvulus, and resection of the necrotic small bowel was performed. It was thought to be caused by multiple abdominal operations resulting in adherent ileus and then leading to strangulated ileus. Despite multiple episodes of aneurysm, immunological tests did not reveal abnormalities.

To minimize the occurrence of postoperative complications, preoperative immunosuppressive therapy and nutritional support were intensified. During this period, the patient's vital signs and symptoms were closely monitored in preparation for emergency surgical treatment at any time. For this recurrent aneurysm, completely open repair was challenging due to the history of multiple surgeries. Facing the celiac trunk, superior mesenteric artery, and right renal artery, fenestrated endovascular repair or chimney stents are difficult and very costly in terms of medical care. Due to thoracoabdominal aortic aneurysm, reconstruction is time consuming, and the organ has a high risk of ischemic damage and even necrosis in traditional open surgery. The patient was young and had undergone multiple surgeries. To shorten the duration of aortic cross-clamping and end-organ ischemia, hybrid

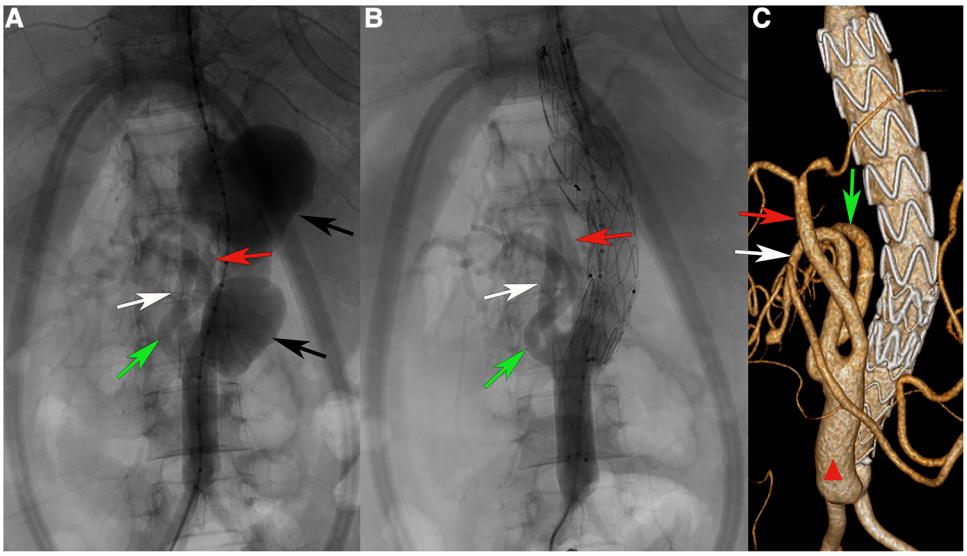

FIGURE 1
(**A**) Angiography after completion of splanchnic artery branch reconstruction. (**B**) Angiography after completion of stent placement. (**C**) After 5 years of follow-up, CTA showed that the pseudoaneurysm cavity completely disappeared, and the stent and branch arteries were patent. Green arrow, right renal artery. White arrow, superior mesenteric artery. Red arrow, celiac artery. Black arrow, abdominal aortic pseudoaneurysms. Red triangle: Artificial blood vessel.

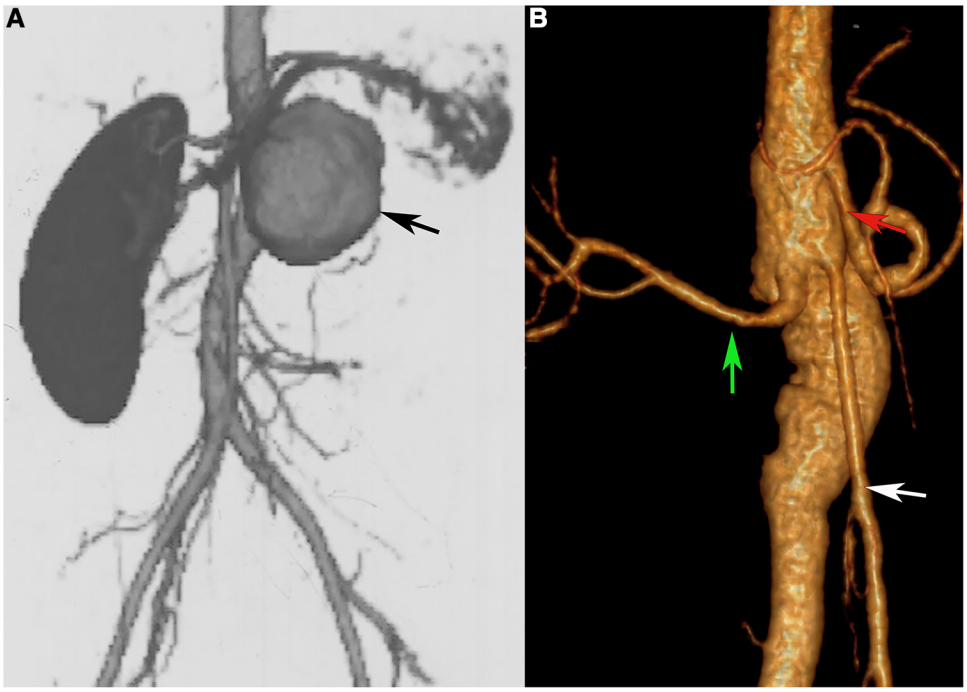

FIGURE 2
(A) abdominal aortic pseudoaneurysm (black arrow). (B) CT Angiography after pseudoaneurysm resection and abdominal aorta reconstruction. Green arrow, right renal artery. White arrow, superior mesenteric artery. Red arrow, celiac artery. Black arrow, abdominal aortic pseudoaneurysms.

surgery was performed (artificial vessel shaping *in vitro*-bypass-endovascular stent repair). When the internal environment improved, we started the surgical treatment. Prior reconstruction used an artificial vessel stent to reconstruct the abdominal trunk, the right renal artery, and the superior mesenteric artery (16 × 8 mm bifurcated artificial vessel was reshaped into three bifurcations), anastomosed the artificial vessel to the aorta at the normal vessel underneath the second abdominal aortic aneurysm, and then reconstructed the branch arteries sequentially, which greatly reduced the organ ischemia time. After reconstruction was completed (**Figure 1A**, left), the first stent (Medtronic ENEW2020C80) was released above the artificial vessel anastomosis. The second stent (VAMC2622C150TE) was released in the middle of the first stent, with the proximal end of the stent located to the normal aorta located superior to the thoracoabdominal aortic aneurysm (**Figure 1A**, right).

The patient was discharged on antiplatelet therapy (100 mg aspirin and 40 mg glucocorticoid) and regular immune therapy in the rheumatology ward. During her 60-month follow-up, the graft had no displacement or internal leakage, and the reconstructed blood vessels were unobstructed (**Figure 1C**).

Discussion

Behcet's disease (BD) is considered a syndrome rather than a disease and is a systemic variable vessel vasculitis that involves many systems (8, 15). It has the highest prevalence along the ancient silk road, which stretches from the Mediterranean through the Middle East to East Asia (16). BD usually starts during the second or third decade of life, and an early age of disease onset seems to be a poor prognostic factor in addition to male sex (17). The majority of patients (75%) experience their first vascular event within 5 years of disease onset (6). Vascular disease develops in up to 25% to 35% of patients and has a definite male preponderance (6, 18).

Immunosuppressive therapy, such as cyclophosphamide, corticosteroids, and even monoclonal anti-TNF antibodies, is the main medical treatment for BD aneurysms (8, 19). Medical therapy should be started as early as possible to provide a stable internal environment for surgical treatment and reduce the incidence of postoperative complications (20). The timing of surgery depends on the size of the aneurysm and the patient's symptoms.

At present, open surgery or endovascular repair is still the main surgical method for aneurysms or pseudoaneurysms. Medical treatment combined with endovascular aneurysm or pseudoaneurysm exclusion in BD is feasible and effective (21–23). The graft occlusion rate despite immunosuppressive treatment was approximately 40% in patients who received bypass surgery (18). In addition, the incidence of pseudoaneurysm formation at the anastomosis, stent anchoring area or access site for stent graft introduction and graft occlusion is high. Several patients accepted intervention again because pseudoaneurysms or aneurysms occurred at other sites (24, 25). Previous studies indicated that most patients died from aneurysm or pseudoaneurysm rupture after surgery (21, 25, 26). Therefore, reducing the occurrence of postoperative aneurysms and

pseudoaneurysms is the key to improving the prognosis. Mousa et al. proposed that mechanical prosthetic wrapping for vascular anastomoses could reduce postoperative pseudoaneurysm formation after aortic aneurysm in BD, and his study showed a 6.3% (1/16) incidence of postoperative anastomotic pseudoaneurysm. The surgeries in BS patients had a large risk of complications such as wound dehiscence, infection, or graft failure. Implementing immunosuppressive treatment can reduce the occurrence of postoperative complications. The results of the pathology test can help to guide the initiation of postoperative immunotherapy (27). Hong et al. reported a successful case-hybrid endovascular repair of thoracic aortic aneurysm in a patient with Behcet's disease following right to left carotid-carotid bypass grafting in 2011 (28).

In our case, the patient underwent three procedures for aneurysms: the first was resection of the large renal aneurysm and left kidney, the second was open surgery on the proximal renal abdominal aortic aneurysm, and the third was hybrid surgery for the pseudoaneurysm of the proximal and distal anastomosis. We speculated that the postoperative pseudoaneurysm that formed after the second procedure was due to the inadequate length of the proximal and distal resected abdominal aorta and the active inflammatory period, which resulted in the abnormal vessel still existing at the anastomosis. To avoid the formation of another pseudoaneurysm at the anastomosis, adequate preparation was performed before surgery. The hybridization procedure, retrograde visceral artery bypass and endovascular repair, was performed. This surgical approach allowed for less clamping of the suprarenal abdominal aorta and could better maintain intraoperative circulatory stability. After reconstruction of the major visceral artery, the length of the stent anchorage zone could be increased. However, it requires a normal abdominal aorta distal to the aneurysm, and retrograde bypass can cause changes in hemodynamic forces that may cause a slowing of blood flow. In addition, the location of the arterial bypass anastomosis is at risk of reappearing as a pseudoaneurysm and consequently a fatal attack.

According to current reports (29, 30), fenestrated-branched endovascular aortic repair presents another option for treating complex abdominal aortic aneurysms. Despite the high technical success rate and low perioperative mortality, unsatisfactory medium- and long-term prognosis as well as high re-intervention rates have been observed (30). The overall cost of total endovascular surgery without further intervention is already very expensive (31). In view of the above situation, we choose the hybrid operation program. And the excellent follow-up results of the patient's five years after surgery-no recurrence of aneurysm or pseudoaneurysm, no graft infection, and no target vessel occlusion suggested that a hybrid surgical approach can also become a treatment option for Behcet's disease-associated aneurysms.

Conclusion

Hybrid surgery may be a feasible and effective strategy for BD aneurysms. Adequate preoperative and postoperative immunotherapy with arterial anastomosis away from the diseased artery may be the key to success.

Data availability statement

The original contributions presented in the study are included in the article/Supplementary Material, further inquiries can be directed to the corresponding author.

Ethics statement

Written informed consent was obtained from the individual for the publication of any potentially identifiable images or data included in this article.

Author contributions

GC and JW are joint first authors and wrote the manuscript. JW was responsible for patient follow-up. CC was responsible for reviewing manuscripts. BH, the corresponding author, was responsible for the review and submission of the manuscript. All authors contributed to the article and approved the submitted version.

Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

Publisher's note

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Thoracic endovascular aortic repair under venoarterial extracorporeal membrane oxygenation for acute aortic dissection patients: a case report

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Background: Open repair and replacement of the diseased aorta is still the standard treatment for type A aortic dissection (TAAD) in most patients. In endovascular treatment alone, ensuring adequate blood supply to the brain while covering the dissection with a stent is difficult.
Case presentation: This study includes a 71-year-old male patient with type A aortic dissection presented at a recent follow-up examination after having undergone thoracic endovascular aortic repair (TEVAR) plus left subclavian artery chimney stent reconstruction for descending aortic dissection 5 years ago. Preoperative computed tomographic angiography, computed tomographic perfusion, and transcranial Doppler showed an intact cerebral arterial ring and good collateral circulation. We successfully performed an endovascular repair of the thoracic aorta with venoarterial extracorporeal membrane oxygenation (V-A ECMO) to protect the craniocerebral blood supply, greatly increase the safety of the operation, and ensure a good prognosis.
Conclusion: TEVAR under V-A ECMO protection is beneficial for patients with TAAD because of its minimal trauma, rapid recovery, few complications, and low mortality.

KEYWORDS

thoracic endovascular aortic repair, V-A ECMO, aortic dissection, type A aortic dissection, case report

Background

Thoracic endovascular aortic repair (TEVAR) is less invasive and has a faster recovery, fewer perioperative complications, and lower mortality (1). It is effective in preventing thrombosis of the false lumen and aortic dissection enlargement, as observed in a long-term follow-up. Therefore, TEVAR has been identified as the first-line treatment for complex type B aortic dissection (cTBAD). In contrast, acute type A aortic dissection (TAAD) is usually treated with open thoracotomy and total arch replacement in the early stage, but the risks of trauma and perioperative mortality are high. This type of dissection has long been unsuitable for TEVAR (2). In recent years, the use of lumen therapy for the treatment of ascending aortic lesions has become a debated and complicated topic.

Abbreviations
TEVAR, thoracic endovascular aortic repair; cTBAD, complex type B aortic dissection; TAAD, type A aortic dissection; CTA, computed tomographic angiography; CTP, computed tomographic perfusion; TCD, transcranial Doppler.

In this case, we describe a patient with acute TAAD undergoing TEVAR in Ningbo Huamei Hospital, University of Chinese Academy of Sciences, under the cooperation of a vascular surgery team and an emergency ECMO team.

Case presentation

A 71-year-old male patient was followed up at irregular intervals. When his chest pain symptoms gradually worsened, he was followed up in a local hospital by a CT scan, which revealed problems before coming to our hospital. TEVAR and left subclavian artery fenestration stent implantation were performed for his descending aortic dissection 5 years ago in our hospital. He had a history of rheumatoid arthritis and had been taking steroids for a long time. The follow-up computed tomographic angiography (CTA) revealed TAAD with proximal occlusion of the brachiocephalic trunk artery due to obvious compression of the false lumen (**Figures 1A,B**). The patient had no obvious symptoms or clinical manifestations of cerebral ischemia before surgery, and CTA, computed tomographic perfusion (CTP), and transcranial Doppler (TCD) examinations of the cerebral arteries showed that the cerebral ring was intact with good collateral circulation. After fully informing the patient of the advantages and disadvantages of thoracotomy and interventional surgery, the patient again strongly requested endovascular treatment and refused thoracotomy. After discussions between the vascular surgery team and the ECMO team, TEVAR under venoarterial extracorporeal membrane oxygenation (V-A ECMO) protection was planned. The procedure is as follows.

After successful anesthesia, routine disinfection and towel laying were performed, followed by bilateral neck and left elbow incisions to expose the bilateral common carotid artery and left brachial artery, respectively, and the proximal and distal rubber circles were retained for use. After systemic heparinization (5000 IU), bilateral carotid arteries were punctured by Seldinger's method with an 8-F vascular sheath. The proximal innominate artery was occluded in the true lumen of the carotid artery and the innominate artery, as shown by right carotid sheath angiography. Using Seldinger's method, we punctured the left brachial artery with a 6-F vascular sheath and inserted an indwelling pigtail catheter into the ascending aorta.

The left femoral artery was successfully punctured using Seldinger's method, and an 11-F arterial sheath was indwelled. The right femoral vein was punctured successfully using Seldinger's method, and a venous drainage catheter (Maquet®, 21 F) was inserted into the right atrium opening of the inferior vena cava. An arterial perfusion catheter (Maquet®, 15 F) was inserted through the right axillary artery to the proximal end of the right subclavian artery, and ECMO was successfully started at a flow rate of 1 L/min.

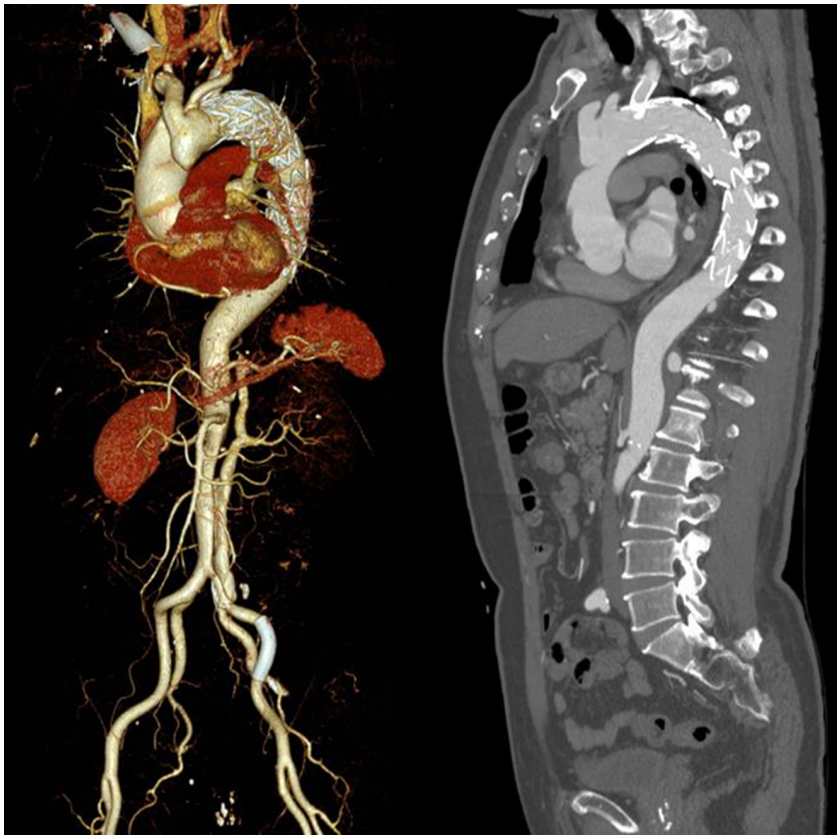

FIGURE 1
Follow up examination of aortic CTA.

The left brachial artery was punctured successfully by Seldinger's method using a 6-F vascular sheath, followed by a pigtail catheter to the ascending aorta, and the sheath was indwelled. A 4-F labeled angiography catheter was pushed up to the ascending aorta through the left femoral artery sheath. The aortic angiography showed a dissecting aneurysm in the greater curvature of the aortic arch, involving the innominate artery, the right common carotid artery, and the right subclavian artery (**Figure 2A**). A covered stent (GORE® TGU454520) was inserted through the right femoral artery of the thoracic aorta and pushed upward under fluoroscopy so that the proximal marker of the stent was positioned in the ascending aorta, approximately 2 cm away from the coronary artery opening, and then released to fully cover the diseased segment.

The liver puncture needle was inserted through the vascular sheath of the left carotid artery, following the V-18 guide wire. After the successful puncture of the covered stent, the guide wire was inserted into the ascending aorta and then dilated with a 4-mm balloon. After exchanging the superhard guide wire, 6- and 10-mm high-pressure balloons were used for expansion. After swapping into the 11-F sheath, an 11 mm × 50 mm Gore VIABAHN covered stent was implanted. The proximal end exceeded the main stent of the thoracic aorta by about 1 cm, and the stent was released under fluoroscopy. In this way, the left common carotid artery *in situ* fenestration stent placement was completed. In the same way, innominate artery *in situ* fenestration stent implantation was performed by implanting a 9 mm × 50 mm Gore VIABAHN covered stent. The angiography was satisfactory.

After smooth bilateral carotid blood flow was achieved, the ECMO flow was slowly downregulated, and the whole procedure ensured that the oxygen saturation of both brains was greater than 65%. The puncture needle was inserted through the left brachial artery access, followed by a V-18 guide wire, and then the guide wire was inserted into the distal part of the descending aorta. After stepwise dilation of the puncture orifice with a balloon, the 11 mm × 50 mm Gore VIABAHN covered stent was inserted; the stent was released approximately 1 cm beyond the proximal left subclavian artery opening.

ECMO was stopped, and aortography and coronary arteriography were performed again. The results showed that the covered stent was positioned satisfactorily, the stent was close to the vascular wall, there was no twist or end leakage, and the covered stent was not blocking the coronary artery. The thoracic aortic dissection was completely isolated by covered stents, and contrast media could be seen in the innominate artery, right common carotid artery, right subclavian artery, left common carotid artery, and left subclavian artery (**Figure 2B**). The operation was completed, the ECMO arterial catheter was removed, and the left femoral artery puncture wound was successfully closed with an embedded ProGlide vascular suture device. The right axillary artery incision was closed with 6-0 Prolene. The right femoral vein was closed with the previously embedded ProGlide suture device, and the elastic band was applied for compression. On the second day after surgery, the patient awoke in the EICU with stable vital signs and good muscle strength in the extremities and was transferred to the ward. One week later, the patient was discharged and followed up for 6 months without obvious discomfort. CT angiography showed blood vessels in good condition (**Figure 3**). Informed consent was obtained from the patients and family members for case information reporting.

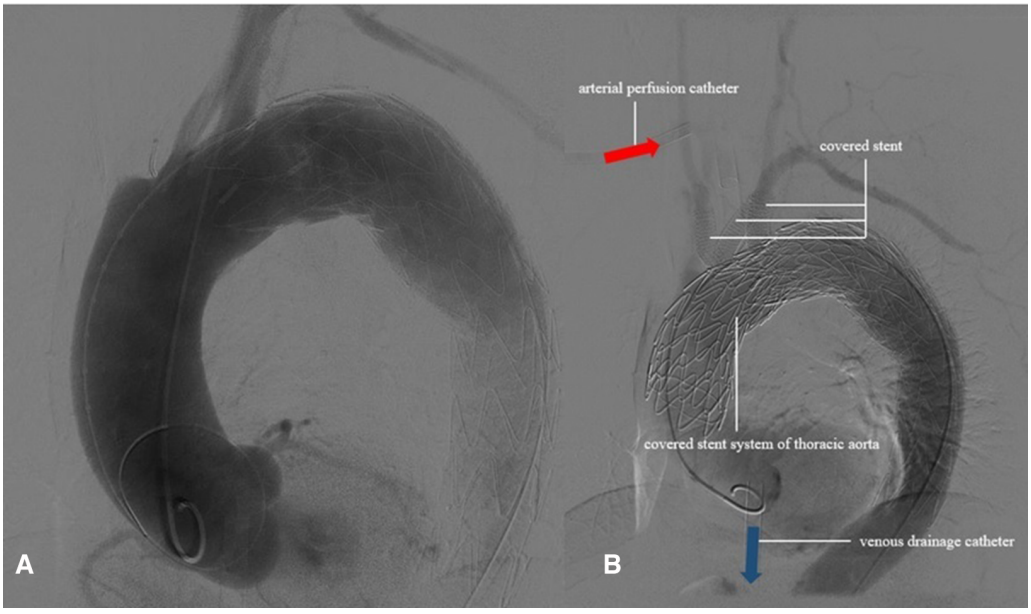

FIGURE 2
The aortic angiography showed that the dissecting aneurysm in the greater curvature of the aortic arch was formed.

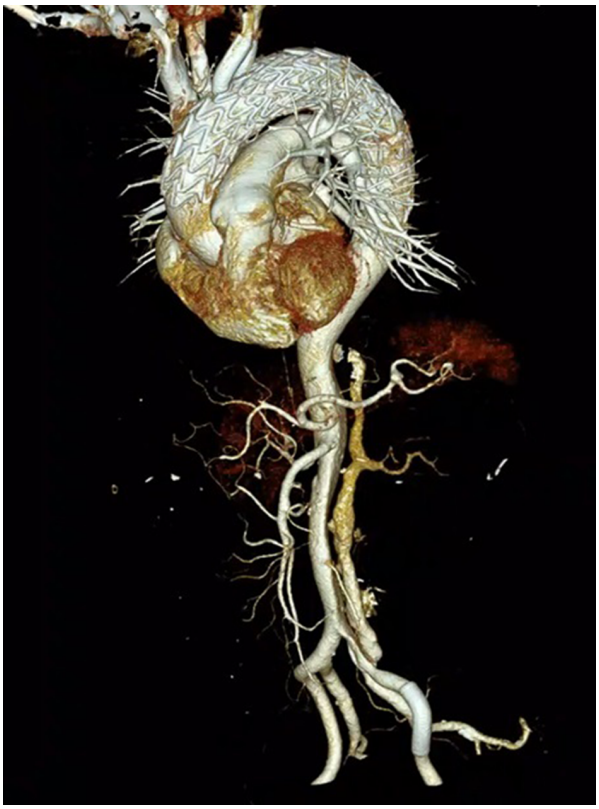

FIGURE 3
Aortography was performed at six months post-operative follow-up.

Discussion and conclusions

Acute TAAD is a cardiovascular emergency caused by weakness or tear of the intima leading to the formation of a false lumen in the media. Blood rushes into the false lumen, enlarging the tear at the proximal, distal, or both ends. When the false lumen compresses the aorta, it can lead to poor perfusion of the coronary arteries, brachiocephalic trunk, and their branches, aortic valve insufficiency, and aortic rupture (3). Open repair and replacement of the diseased aorta remain the standard of care for most patients. The primary objective of surgical treatment is to remove the origin of the intimal tear and reconnect the intima with the media and adventitia to eliminate the false lumen (4). TEVAR is an additional treatment mode for thoracic aortic diseases and can be an alternative treatment for high-risk or inoperable patients. Because stent transplantation is less invasive, TEVAR is well tolerated in an older and unwell patient population without needing thoracotomy, cardiopulmonary bypass, or deep hypothermic circulatory arrest.

In 2002, Mitchell et al. divided the thoracic aorta into five regions ranging from 0 to 4 according to the relationship between each part of the thoracic aorta and branches of the brachial and cephalic arteries (**Figure 4**) (5). This case is a complex case of aortic dissection. During CTA follow-up 5 years after TEVAR for aortic dissection in the Z4 region, proximal stent dissection was found, which progressed to TAAD, involving the Z0 region and proximal occlusion of brachial and cephalic trunk arteries by obvious false lumen compression. The difficulty of the treatment lies in ensuring the blood supply to the brain while covering the dissection rupture with the covered stent, especially when the brachiocephalic trunk artery, the left common carotid artery, and the left subclavian artery are covered at the same time. Moreover, the fenestration of the three branches is not completed, the blood supply to the bilateral brain is completely blocked, and the risk of cerebral hypoperfusion is high. However, the patient refused to allow the opening of his chest due to his advanced age, poor pulmonary function, and underlying diseases. Combined with CTA, the Circle of Willis was complete, and TCD showed that the collateral circulation of the anterior and posterior cerebral communicating arteries was satisfactory and the blood flow of the jugular vein was smooth. This provides an opportunity for V-A ECMO to protect cerebral perfusion by supporting the right cerebral blood supply via right axillary artery perfusion, and the left cerebral blood supply is temporarily provided by the right cerebral blood supply via the cerebral arterial ring (**Figure 3**).

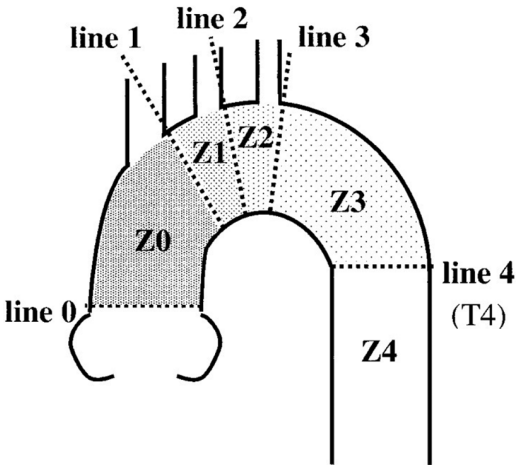

FIGURE 4
An anatomical map of each landing zone bordered by lines delineating the distal sides of the branch arteries of the aortic arch (5).

V-A ECMO is a well-developed and commonly performed technique to provide circulatory support to critically ill patients with refractory cardiogenic shock and cardiac arrest (6). However, due to the retrograde perfusion of the aorta, V-A ECMO has been considered relatively contraindicated in the field of aortic dissection. ECMO as a bridge to support this patient undergoing TEVAR eliminated the contraindication and provided better blood supply to the brain. The intubation strategy we adopted selected femoral vein drainage and right axillary artery perfusion, which can ensure better and more accurate flow support for the brain compared with femoral artery perfusion, and this method also prevents the femoral artery perfusion blood flow from becoming blocked by the thoracic aortic covered stent system.

Axillary artery cannulation does not require median sternotomy nor will it cause complications related to femoral artery cannulation. The literature supports that the limb can

tolerate 4–6 h without a blood supply (7). In this case, the tolerable operation time of the right upper limb under ECMO protection is still uncertain. We recommend limiting the operative time or taking other measures to improve blood supply to the right upper extremity (distal arterial sheath placement to open collateral circulation).

We believe that the most important condition for unilateral cerebral artery perfusion is the integrity of the Willis ring and collateral circulation in the patient's brain (8). In this case, the collateral circulation function of the cerebral artery ring was carefully investigated before surgery, but there was still the possibility of blood hypoperfusion in the distribution area of the left external carotid artery. At present, there is no good method to evaluate the blood supply of cervical pulp. Once high cervical pulp ischemia injury occurs, the prognosis of surgery can be seriously affected. In addition, there are no exact data on the specific flow under ECMO protection. The team estimated the possible range of cerebral blood flow based on the blood flow velocity and vessel diameter measured by TCD of bilateral common carotid arteries before surgery, which was approximately 20% of cardiac output in the calm state. We believe that in the future, intraoperative TCD monitoring of cerebral blood flow combined with cerebral oxygen monitoring can be implemented to obtain further research data.

Data availability statement

The original contributions presented in the study are included in the article/Supplementary Material, further inquiries can be directed to the corresponding author.

Ethics statement

The study was approved by the Ethics Committee of Hwa Mei Hospital. Informed written consent was obtained from the patient's legal representative for the publication of this report and any accompanying images. Written informed consent was obtained from the patient for the publication of this study.

Author contributions

YS designed the idea. KX and PL collected the clinical data. LZ and LD wrote the manuscript. LZ was a major contributor in writing the manuscript. All authors contributed to the article and approved the submitted version.

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Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

Publisher's note

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Case report: Pheochromocytoma complicated by type B aortic dissection

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Introduction: Pheochromocytomas combined with aortic dissections are rare. Treatment of aortic dissection can be complicated by the presence of pheochromocytomas.
Case presentation: we present the case of a 48-year-old male who visited the hospital with chest and back pain for 13 h. Enhanced computed tomography (CT) revealed a type B aortic dissection combined with a left adrenal mass (72 mm). Elevated 24-h urinary vanillylmandelic acid levels can aid in the diagnosis of pheochromocytomas. Aortic dissection due to unstable hypertension secondary to pheochromocytoma is rare and complicates the procedure. Thoracic endovascular aortic repair was performed, and antihypertensive treatments were administered after surgery. After hypertension was addressed and the patient was stable, laparoscopic resection of the adrenal mass was performed.
Conclusions: despite its rarity, it is important to consider pheochromocytoma as a differential factor for unstable hypertension when an aortic dissection is found.

KEYWORDS
pheochromocytoma, type B, aortic dissection, case report, computed tomography

Introduction

Aortic dissection is a life-threatening condition caused by a tear in the intima, where blood flow splits the intima and creates a false lumen in the aortic wall. Disruption of the normal blood flow may lead to poor organ perfusion, aortic dissection, and advanced aneurysm formation. Risk factors for aortic dissection include smoking, poorly controlled hypertension, male sex, age, and connective tissue disease (1, 2).

Type B aortic dissection (TBAD) occurs distal to the left subclavian artery (LSCA). The early prognosis for TBAD is good, with a 1-year survival rate of 65% for patients. However, the late prognosis is worse because of anatomy-related complications, with a 5-year survival rate of 50% (3). Thoracic endovascular aortic repair (TEVAR) is recommended as the first-line treatment in patients with acute complicated TBAD (4, 5).

Treatment of aortic dissection can be complicated by the presence of pheochromocytomas. Pheochromocytomas are tumors in which the adrenal medulla and other chromophobic tissues of the adrenergic system produce excessive amounts of catecholamines, which can lead to uncontrolled blood pressure (6). Here, we present a case of aortic dissection as the initial presentation of an undiagnosed pheochromocytoma.

Case presentation

A 48-year-old male patient presented to our clinic with chest and back pain that had persisted for 13 h. The patient complained of sudden onset of pain in the precordial

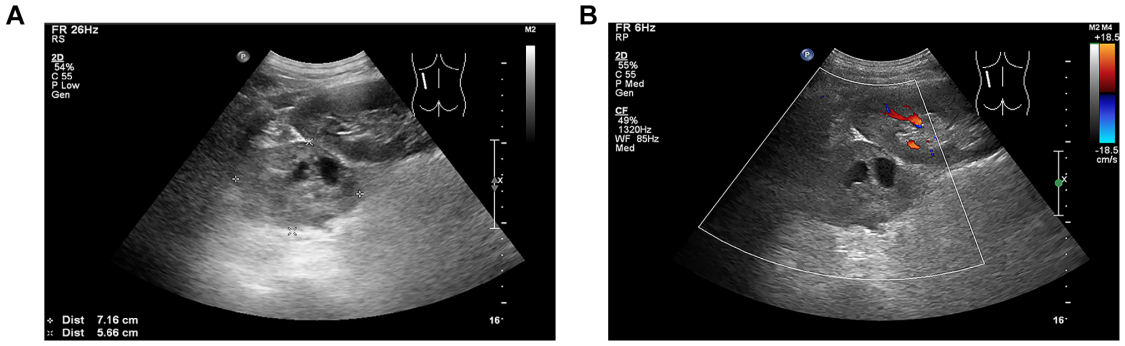

FIGURE 1
(A) Heterogeneous echogenic mass with a size of approximately 72 × 57 mm in the left adrenal gland with clear boundary and regular shape, in which patchy anechoic areas were observed. (B) No significant blood flow signal inside the left adrenal mass.

region more than 13 h prior during the course of a meal, which was persistent and dramatic, and then radiated to the abdomen and waist, with profuse sweating, posterior abdominal distension, vomiting of approximately 200 ml of dark brown fluid, no clots, no syncope, no diarrhea, no cough, or expectoration. The etiology of this condition remains unclear. The patient denied any history of hypertension, diabetes mellitus, tumor, trauma, or previous surgery, and had a 30-year history of smoking.

Physical examination revealed an acutely ill face, clear consciousness, poor spirit, heart rate of 125 beats/min, blood pressure of 210/94 mmHg, distended abdomen, drum sounds on percussion, no tenderness or rebound tenderness, and the presence of femoral and dorsalis pedis pulses. According to the aortic dissection detection risk scoring (ADD-RS) system proposed by the aortic diagnosis and treatment guidelines in 2014, ADD-RS = 1. The etiology of the patient was unknown; clinicians could not rule out aortic dissection, urinary calculi, or other related diseases; therefore, echocardiography and abdominal urinary ultrasonography were performed on the patient. Echocardiography showed normal findings, and abdominal ultrasonography revealed a mass in the left adrenal region (72 × 57 mm, Figure 1). Contrast-enhanced computerized tomography (CT) of the abdomen revealed that the intimal flap moved inward from the descending part of the thoracic aorta to the level of the bilateral common iliac arteries, the lumen showed "double-lumen" changes, the right renal artery opened into the false lumen, and the left renal artery opened into the true lumen, consistent with aortic dissecting aneurysm (Stanford type B). A soft tissue mass was observed in the left adrenal gland, with uneven density, approximately 61 × 53 mm in size, with a clear boundary, and with significant enhancement after contrast-enhanced scanning in which there was necrosis; pheochromocytoma was first considered (Figure 2). The patient subsequently underwent aortic computed tomographic angiography (CTA), which revealed Stanford TBAD (Figure 3). Laboratory tests showed a 24-h urine vanillylmandelic acid (VMA) level of 20.51 mg/24 h (reference value: ≤12 mg/24 h) and a D-dimer level of 6.75 mg/L (reference range 0.0–0.5 mg/L).

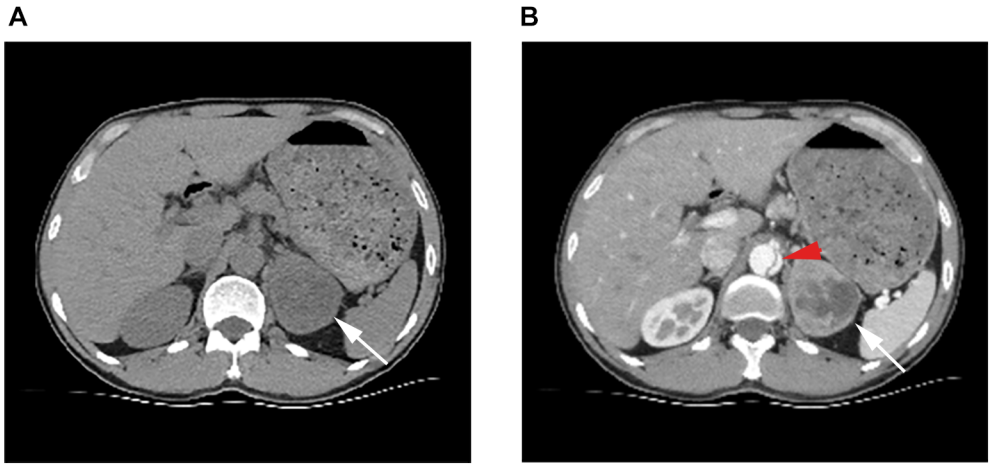

FIGURE 2
(A) Ct scan showing a soft tissue mass in the left adrenal gland with uneven internal density, clear borders, and regular shape. (B) White arrows point to the left adrenal mass in the arterial phase, which is significantly enhanced and necrotic inside. Red arrows point to dissection of the abdominal aorta.

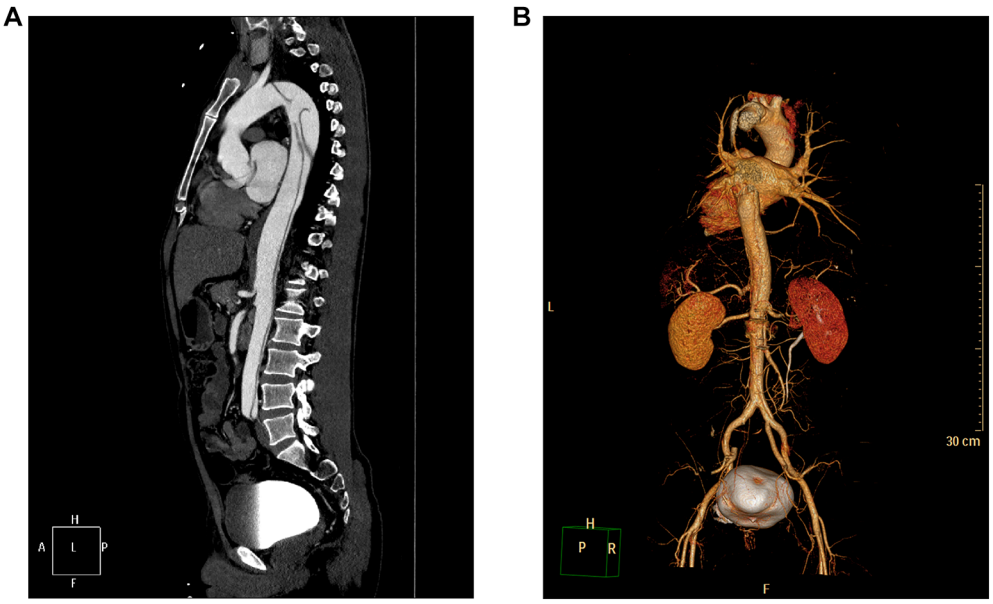

FIGURE 3
CTA showing medial migration of the intimal sheet from the descending thoracic aorta to the level of both common iliac arteries, and the lumen showing "double-lumen" changes, consistent with aortic dissecting aneurysms (Stanford type B). (A) Maximal intensity projection of the sagittal section of the aorta. (B) Three-dimensional reconstruction of the aorta.

Nitroglycerin and esmolol hydrochloride were administered to the patient for antihypertensive treatment after admission and was later replaced with a phentolamine drip due to persistent blood pressure fluctuation. Forty-eight hours after the onset of symptoms, blood pressure control was relatively stable after treatment, and because the patient developed abdominal pain and distension, vomiting of dark brown fluid, and the occult blood test was positive (+ + +) in vomitus, the clinician was concerned about aortic dissection involving visceral malperfusion. After communicating with the patient and his family, the patient actively requested surgery; therefore, thoracic endovascular aortic repair was performed.

Due to concern regarding adrenergic storm caused by intraoperative pheochromocytoma secreting a large amount of catecholamines, a phentolamine drip was given for active antihypertensive treatment before surgery, metoprolol was given to control heart rate, and preoperative blood transfusion preparation was actively performed. The phentolamine drip was continued during the operation and was closely observed by the anesthesiologist, without signs of hemodynamic instability. During the procedure, we sealed the dissection of the descending aorta, with good stent curvature. Multiple digital subtraction angiography (DSA) at the distal end of the stent revealed difficulty in opening the true lumen at the distal end of the graft. The patient's family was informed, and partial artificial blood vessel replacement of the abdominal aorta was recommended. Because of the high surgical risk, the patient's family refused, and requested conservative treatment. Nitroglycerine and esmolol were continued as antihypertensive treatments after surgery, and the blood pressure was stable at 101–154/51–80 mmHg.

After the patient was stable for 14 days, the adrenal mass was treated in the urology department and laparoscopic resection of the adrenal mass was performed. The postoperative tumor pathology report included (left adrenal gland) pheochromocytoma with focal hemorrhage, degeneration, and necrosis (size 6.5 × 5.5 × 3.5 cm). Immunohistochemical staining showed chromogranin A (CgA) (+), S-100 protein (S-100) (+), synaptophysin (Syn) (+), CD56 (+), epithelial membrane antigen (EMA) (−), vimentin (+), GFAP (−), and Ki-67 (+5%) (**Figure 4**). The patient recovered well after surgery; blood pressure and 24-h urine VMA levels returned to normal after re-examination, and the patient was discharged. The patient tolerated both procedures well. When the pheochromocytoma was removed, the improvement in blood pressure was significant, and the patient eventually stopped all antihypertensive drugs. Six months after surgery, we performed a telephone interview; the patient was in good general condition and had stable blood pressure, which was regularly reviewed at a local hospital, and he participated in light physical work five times a week.

Discussion

TBAD with pheochromocytoma or adrenal mass is very rare, and there is no relevant literature reporting the prevalence of the combination of the two. Previous studies suggest that pheochromocytomas are rare tumors that, although usually asymptomatic, can be fatal if left untreated (7). The annual incidence rate is approximately 0.6 per 100,000 individuals (2). Pheochromocytomas are tumors in which the adrenal medulla and other chromophobic tissues of the adrenergic system

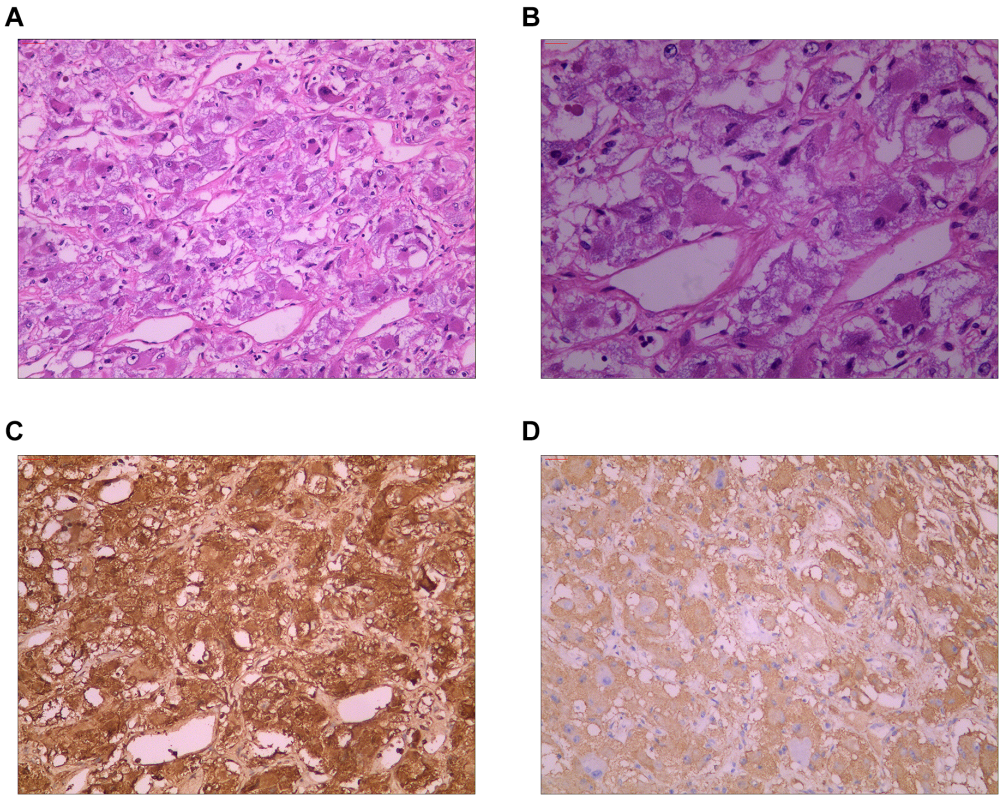

FIGURE 4
(**A**) Hematoxylin-eosin staining (HE) showing that the tumor cells were distributed in nests and beams, and thin-walled and reticular blood vessels were abundant between the cell nests (10 × 10). (**B**) Hematoxylin-eosin staining (HE) showing that the tumor cells were round or oval at high magnification, finely stained, granular, and evenly distributed, with no mitotic figures observed (10 × 20). (**C**) Immunohistochemical staining showing chromogranin A (CgA) (+). (**D**) Immunohistochemical staining showing synaptophysin (Syn) (+).

produce excessive amounts of catecholamines. The clinical signs of pheochromocytomas vary; however, the most common are paroxysmal hypertension or paroxysmal exacerbation of persistent hypertension, palpitations, headache, sweating, and other symptoms resulting from catecholamine release.

Aortic dissection secondary to uncontrolled hypertension due to pheochromocytomas is rare. In this case, the pheochromocytoma was discovered incidentally on abdominal ultrasonography and abdominal enhancement CT. The current treatment of choice for pheochromocytomas is surgical resection (8). However, the risk of surgery is greatly increased by cardiovascular accidents due to severe intraoperative blood pressure fluctuations. The role of hypertension in the development of aortic dissection is indisputable as approximately 80% of patients with aortic dissection experience hypertension (9), particularly in patients with highly fluctuating blood pressure.

It is not the entrapment itself that has a fatal effect on patients with aortic dissection, but the series of changes caused by the progression of the dissection, such as pericardial tamponade, aortic rupture and bleeding, and ischemic necrosis of internal organs and extremities. Therefore, the blood pressure of patients with aortic dissection should be strictly controlled. In this case, the patient had both pheochromocytoma and aortic dissection; therefore, we first chose to perform endoluminal repair of the aortic dissection to address the concerns of pheochromocytoma resection. The patient had a functional pheochromocytoma; adequate preoperative preparation can significantly reduce operative mortality (10). Preoperative hypotension, volume expansion, and arrhythmia correction are fundamental measures to reduce perioperative mortality (10). Therefore, we actively administered phentolamine to lower the blood pressure, metoprolol to control the heart rate, and lactated Ringer's solution to expand the volume before surgery.

This is a rare case of pheochromocytoma combined with TBAD. This case suggests that the possibility of pheochromocytoma should be considered in young patients with paroxysmal exacerbations of persistent hypertension. If the patient has a sudden onset of severe tearing pain in the chest and back, the patient should be alerted to aortic dissection, and CTA should be performed urgently to confirm the diagnosis. When young TBAD patients without a previous history of hypertension present with paroxysmal aggravation of persistent hypertension, the possibility of pheochromocytoma complicating TBAD should be considered. Treatment should first address the most life-threatening aortic dissection, followed by pheochromocytoma resection.

Data availability statement

The raw data supporting the conclusions of this article will be made available by the authors, without undue reservation.

Ethics statement

The studies involving humans were approved by The Ethics Committee of Shaoxing People's Hospital. The studies were conducted in accordance with the local legislation and institutional requirements. The participants provided their written informed consent to participate in this study. Written informed consent was obtained from the individual(s) for the publication of any potentially identifiable images or data included in this article.

Author contributions

LF: contributed to conception and design of the study. DY: organized the database and wrote the first draft of the manuscript. The manuscript was revised by XL. All authors contributed to the article and approved the submitted version.

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Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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Supplementary material

The Supplementary Material for this article can be found online at: https://www.frontiersin.org/articles/10.3389/fcvm.2023.1236896/full#supplementary-material

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Case Report: Successful endovascular treatment of acute type A aortic dissection

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Introduction: Open surgical repair remains the current gold standard for the treatment of acute type A aortic dissection. However, especially elderly patients with relevant comorbidities who are deemed unfit for open surgery may benefit from a minimally invasive endovascular approach.
Methods: We report a case of an 80-year-old male with retrograde acute type A aortic dissection and peripheral malperfusion after receiving thoracic endovascular aortic repair due to thoracic aortic aneurysm. Our individualized endovascular approach consisted of left carotid-subclavian bypass, proximal extension of thoracic endovascular aortic repair using a covered stent graft and a single covered stent graft for the ascending aorta in combination with an uncovered stent for the aortic arch.
Results: Postoperative computed tomographic angiography demonstrated excellent outcome with no signs of endoleak or patent false lumen. Follow-up after 3.5 years showed a stable result with no signs of stent failure or dissection progress. No aortic re-interventions were needed in the further course.
Discussion: An individualized endovascular approach may be justified for acute type A aortic dissection in elderly patients with high surgical risk if performed in specialized aortic centers. Additional short-length stent graft devices are needed to address the anatomical challenges of the ascending aorta. For enhanced remodeling of the dissected aorta, the use of an additional uncovered stent may be advisable.

KEYWORDS

aortic dissection, type A, TEVAR, endovascular, ascending aorta

1. Introduction

Acute type A aortic dissection (ATAAD) is a possibly lethal event that demands urgent surgical repair according to current guidelines (1, 2). Though surgical strategies have been developed continuously, mortality and morbidity are still high (3). Perioperative complications such as stroke are not uncommon and may influence the patients' outcome significantly (4, 5). Especially in elderly patients, malperfusion and advanced age may lead

Abbreviations
ATAAD, acute type A aortic dissection; CTA, computed tomographic angiography; NIRS, near infrared spectroscopy; TEVAR, thoracic endovascular aortic repair; PETTICOAT, provisional extension to induce complete attachment.

to significant increase in perioperative risk, possibly even reconsidering the decision for standardized open repair (6). Therefore, offering the possibility of endovascular instead of open surgical treatment for these patients to reduce surgical trauma may be of great interest. Compared to standardized interventional treatment of acute type B aortic dissection by thoracic endovascular aortic repair (TEVAR), an interventional approach for ATAAD comes along with major challenges (2). Due to the possible involvement of the aortic root and aortic valve as well as the coronary arteries and branch vessels of the aortic arch, a stent-based "TEVAR-like" approach is accompanied by substantial technical difficulties and may even be harmful. Various prostheses are available on the market to perform standardized TEVAR of the descending thoracic aorta, e.g., GORE C-TAG® or TAG® (GORE®, Flagstaff, AZ, USA), ZENITH TX2 and Alpha (Cook®, Bloomington, IN, USA), Valiant Captiva (Medtronic®, Dublin, Ireland), or Relay® (Bolton, Barcelona, Spain; now Terumo®, Inchinnan, UK) to name the most popular ones. Thereby, in case of TEVAR for thoracic aortic aneurysm, oversizing of the stent graft is an important key point and should at least exceed the diameter of the landing zones by 10%–15% of the reference aortic diameter (7). In case of type B aortic dissection, oversizing should almost be avoided and may even lead to a higher risk for retrograde dissection if the oversizing rate is greater than 5% (8). Transferring these experiences for the treatment of the ascending aorta, especially in case of ATAAD, should be performed with caution. However, we believe that under certain circumstances, the evaluation of interventional treatment options for ATAAD may be justified to avoid open central repair in elderly patients with several comorbidities and consecutive high perioperative risk. Therefore, we report a case of an 80-year-old male presenting with retrograde ATAAD and consecutive successful endovascular treatment.

2. Case description

2.1. Patient information

An 80-year-old male patient underwent elective computed tomographic angiography (CTA) scan for checkup of a known thoracoabdominal aneurysm. Nine years before, he received open surgical repair of juxtarenal abdominal aortic aneurysm with reimplantation of the left renal artery. Results of the recent CTA (SOMATOM Definition Flash, Siemens®, Erlangen, Germany) are depicted in **Figure 1** and showed a thoracic aortic aneurysm with 63 mm of the descending aorta. According to the current guidelines, TEVAR was recommended to prevent aortic dissection or rupture (2). Therefore, he was referred to a local vascular surgery department for TEVAR. Proximal landing zone diameter distal from the left subclavian artery was estimated to be 30 mm. At this point of time, the patient was free of symptoms. Due to the involvement and reimplantation of the left renal artery, the left kidney was already atrophic for several years, resulting in chronic renal failure stage 3b with a glomerular filtration rate of 45 ml/min. In addition, he suffered from long-term arterial hypertension with fourfold antihypertensive medication, non-insulin-dependent type 2 diabetes and dyslipidemia, both treated with oral medication, as well as an adequate substituted hypothyroidism. There was no anamnestic evidence of genetic or hereditary aortic diseases as well as similar familiar cases.

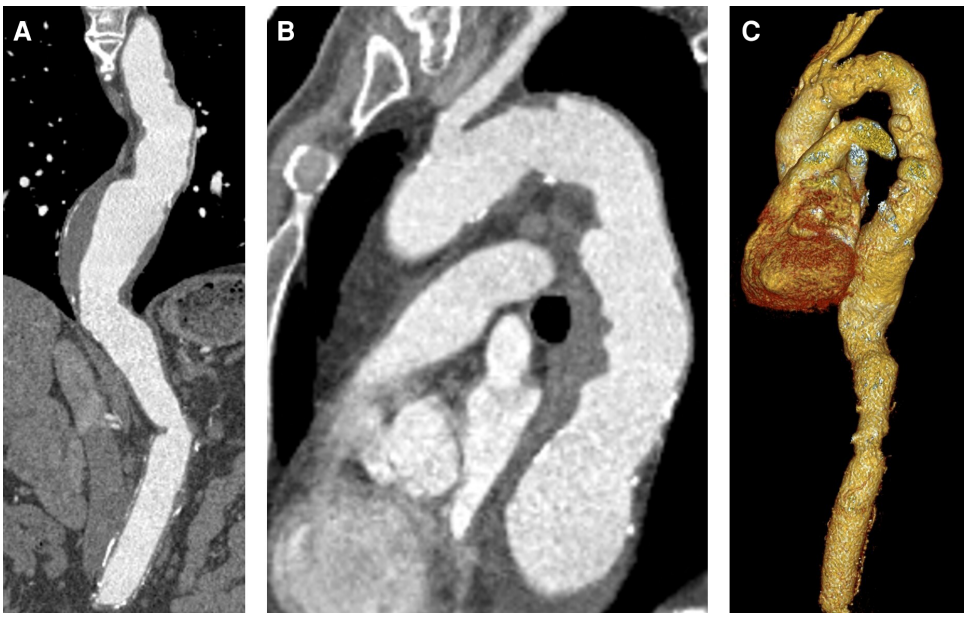

FIGURE 1
Preoperative CTA. (**A**) Descending aortic aneurysm with 63 mm diameter. (**B**) Proximal landing zone distal to the left subclavian artery (30 mm). (**C**) 3D reconstruction.

The patient was prepared for TEVAR in a standard fashion via right femoral artery under general anesthesia. After releasing the thoracic stent graft (Valiant Navion™ 37/37/255 mm, Medtronic®, Dublin, Ireland), which included >20% proximal oversizing, perioperative angiography showed retrograde acute aortic dissection at the proximal edge of the TEVAR prosthesis involving the aortic arch and the ascending aorta. The patient was quickly referred to CTA, which confirmed the diagnosis of retrograde ATAAD. He was transferred stable and uneventfully to our clinic for further treatment.

2.2. Clinical findings

On admission, the patient was intubated, mechanically ventilated, and sedated. Pupils were small, equally round, and reactive to light. He was hemodynamically stable without any inotropic medication in normal frequent sinus rhythm; the lungs were ventilated and clear to auscultation bilaterally. The abdomen was soft and normoactive bowel sounds were present in all four quadrants. Diuresis was sufficient. The extremities were warm and peripheral pulses were palpable except for the right leg, which was cold and livid, indicating the presence of peripheral malperfusion. Hemoglobin was 11 g/dl and lactate 7 mg/dl; pulmonary function was not impaired. At this point of time, no clinical evidence of further systemic or localized malperfusion except for the right leg was present, resulting in class Penn Ab according to the current Penn classification (9).

2.3. Diagnostic assessment

After analyzing the CTA, iatrogenic ATAAD with entry location at the proximal edge of the TEVAR prosthesis in the descending aorta was confirmed, most likely caused by the tip of the stent. Entries in the aortic arch or the ascending aorta could not be identified. According to the new TEM classification by Sievers et al., this resulted in "A, E3, M3+" (A = type A dissection according to Stanford, E3 = entry in the descending aorta, M3+ = peripheral malperfusion with clinically relevant malperfusion) (10). Landing zone length of the ascending aorta was 72 mm, proximal landing zone diameter was 33 mm, and distal landing zone diameter was 28 mm. Preoperative CTA (SOMATOM Definition Flash, Siemens®, Erlangen, Germany) is shown in **Figure 2**. Imaging of the intracranial vessels was not available.

Considering the advanced age as well as extensive comorbidities, we decided to perform complete endovascular treatment in combination with surgical left carotid-subclavian bypass. Thereby, left carotid-subclavian bypass was performed to reduce the potential risk of spinal cord injury or cerebrovascular accident.

2.4. Therapeutic intervention

Prior to surgery, a 7 Fr sheath via left internal jugular vein was inserted in addition to the pre-existing 5 lumen central venous line and arterial access line in left radial artery. Near infrared spectroscopy (NIRS) electrodes were placed bilaterally on the forehead. General anesthesia was continued using propofol and sufentanil. The patient was placed in a supine position prior to standard sterile draping. After application of 2 g cefazoline and team timeout, a skin incision was made parallel to the left clavicle on the neck as access for carotid-subclavian bypass. After cutting the platysma, careful exposition of the jugular vein, the common carotid artery, and the vagus nerve was performed. Then, the left subclavian artery was exposed. After heparin administration, the vessel was partially clamped and incised longitudinally. A Dacron graft (FlowNit® Bioseal, 8 mm, Artivion®, Hechingen, Germany) was anastomosed end-to-side with 5–0 prolene. The same procedure was carried out for proximal anastomosis to the left common carotid artery after careful placement of a distal and proximal clamp. NIRS showed stable and equal values during the whole procedure. After successful de-airing, the circulation was released. For further

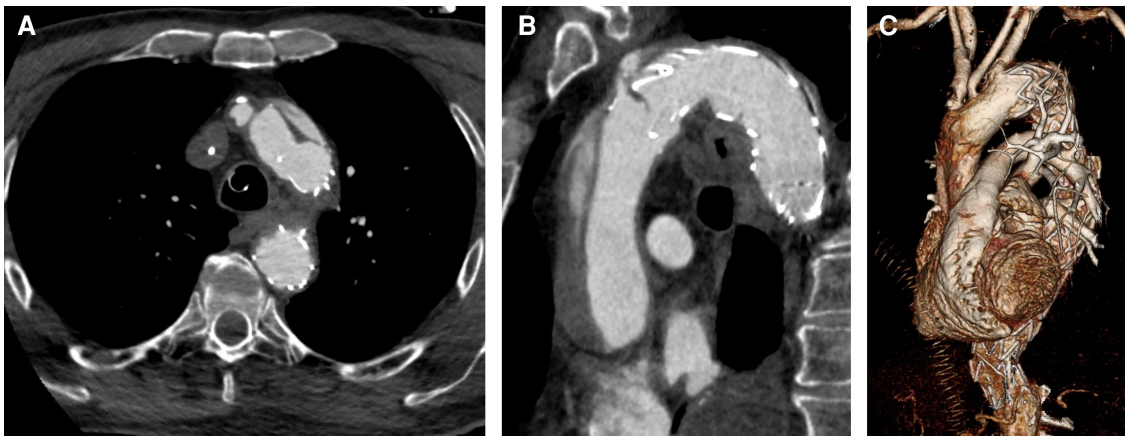

FIGURE 2
CTA after TEVAR and consecutive retrograde ATAAD. (**A**) Entry proximal of TEVAR stent graft. (**B**) Dissection involving the ascending aorta and the aortic arch. (**C**) 3D reconstruction.

angiography, a 5 Fr sheath was placed in the prosthesis after application of a purse-string suture followed by a diagnostic pigtail which was placed in the ascending aorta.

After successfully performing the carotid-subclavian bypass, a 2-cm skin incision was made in the right groin area and the right common femoral artery was punctured percutaneously. Two endovascular closure systems (ProGlide™, Abbott® Cardiovascular, Plymouth, MA, USA) were inserted and prepared for the end of the procedure. Then, a 16-Fr sheath was inserted followed by a flexible wire (Radifocus™, Terumo®, Inchinnan, UK) to pass the aortic valve for pigtail catheter-guided insertion of a pre-bent double curved wire (Safari™, Boston Scientific®, Marlborough, MA, USA), which was placed in the left ventricle. In addition, a temporary pacing electrode was placed in the right ventricle through the right femoral vein. After angiographic control of catheter position and visualization of true and false lumen, the 16-Fr sheath was removed and the stent graft delivery system (Relay® Plus NBS 34/34/109 mm, Terumo®, Inchinnan, UK) was inserted until reaching the aortic arch. After carefully placing the edge of the stent graft delivery system close to the branch of the left common carotid artery (Zone 2), rapid pacing was performed (180/min) and the stent was released. Angiographic control showed adequate expansion and stent position as well as sufficient perfusion of the left common carotid artery. As next step, another thoracic stent graft delivery system (Valiant Navion™ 37/37/55 mm, Medtronic®, Dublin, Ireland) was prepared and inserted until reaching the ascending aorta. Angiographic control was performed precisely to visualize both coronary ostia and the brachiocephalic trunk. Thereby, a 6-Fr pigtail (Boston Scientific®, Marlborough, MA, USA) was parked at the non-coronary cusp considering the radiographic angle to support the identification of the landing zone and avoid protrusion of the stent graft into the coronary arteries. After exact positioning between the coronary ostia and the brachiocephalic trunk, the stent graft was released during rapid pacing (180/min). For further stabilization and aortic remodeling, analogous to the PETTICOAT technique (provisional extension to induce complete attachment), an additional uncovered nitinol stent (E-XL™ 36/07 mm, formerly JOTEC® GmbH, now Artivion®, Hechingen, Germany) was inserted into the aortic arch to connect both stent grafts (11). Angiographic and perioperative transesophageal echocardiography showed satisfactory results with adequate positioning and successful elimination of false lumen flow. The stent graft delivery system including the sheath was removed, and endovascular vessel closure was performed by using the sutures prepared by ProGlide™. Then, the pigtail catheter and sheath were removed from the subclavian-carotid bypass graft. Finally, a redon drainage was placed and the wound was closed in layers finishing with intracutaneous skin suture.

Perioperative transesophageal echocardiography revealed preserved biventricular function, no relevant aortic insufficiency, or any wall motion abnormalities. No persistent flow could be detected in the false lumen. Intraoperative angiographic steps are shown in **Figure 3**.

2.5. Follow-up and outcomes

The patient was transferred to the intensive care unit under stable conditions. Due to slight anisocoria and still cold and pale right leg, CTA was performed. No signs of cerebral ischemia or vessel obliteration were evident. Right femoral and iliac artery was dissected, but distal perfusion was preserved and peripheral pulses were palpable. Symptoms diminished in the further course and the patient was extubated after the first postoperative day. However, he initially presented with paraparesis with only minimal preserved motoric function of the right leg. Fortunately, motoric function recovered significantly in the further

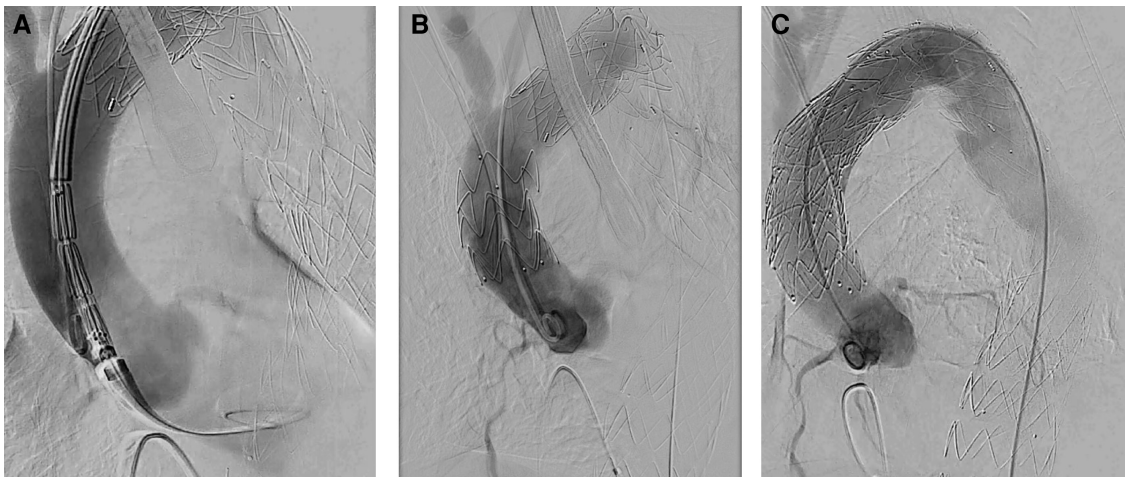

FIGURE 3
Intraoperative angiography. (A) Aortic dissection after releasing TEVAR stent for proximal expansion. (B) Aortic dissection after stenting of the ascending aorta and parking of a pigtail catheter in the non-coronary cusp. (C) Postoperative result after placing an uncovered stent in the aortic arch ("Reverse PETTICOAT").

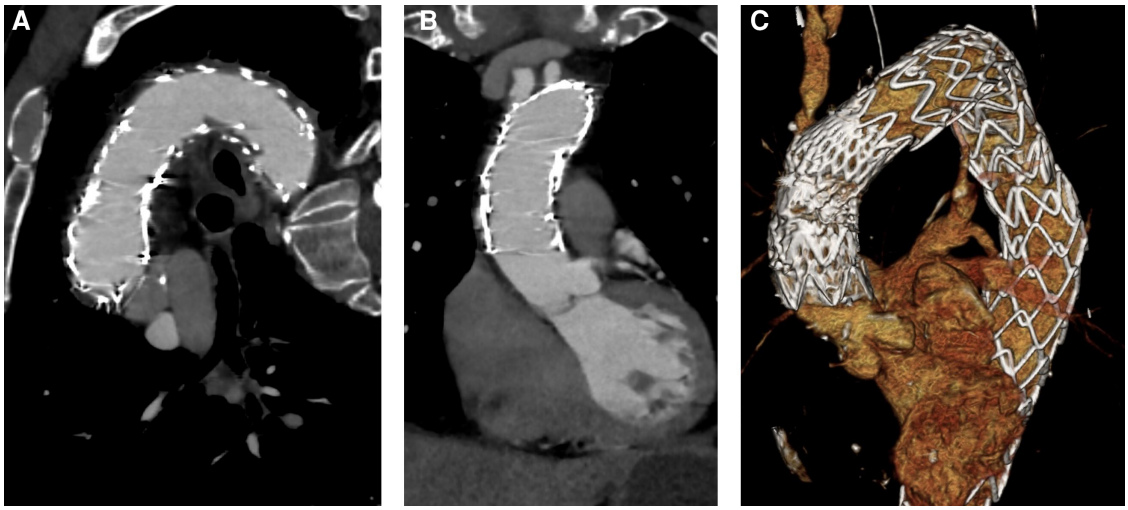

FIGURE 4
Postoperative CTA. (A) Stent grafts covering the ascending aorta and the aortic arch with no patent false lumen. (B) No residual dissection in the ascending aorta. (C) 3D reconstruction.

course. No signs of endoleak or false lumen patency could be detected by CTA. Results of postoperative CTA (SOMATOM Definition Flash, Siemens®, Erlangen, Germany) is shown in **Figure 4**. The patient was referred to the normal ward on the eighth postoperative day after slowly recovering from postoperative delirium.

No aortic re-interventions were necessary after discharge. After 3.5 years of follow-up, CTA showed no progress of the disease, including no evidence of stent failure, no signs of endoleak, and no aneurysm or dissection progress. CTA (SOMATOM Definition Flash, Siemens®, Erlangen, Germany) results are shown in **Appendix Figure A1**. A timeline that summarizes the patient's course including the relevant events is shown in **Appendix Figure A2**.

3. Discussion

In this case report, we demonstrate the efficacy and anatomical feasibility of an individualized endovascular approach for the treatment of ATAAD. Several other groups have reported similar cases, bearing in mind that these procedures were carried out in highly specialized aortic centers by an interdisciplinary team and require meticulous planning (12, 13). However, the rate of technical success is reported to be greater than 90%, and periprocedural mortality is extremely low (12–14). Thereby, the anatomical feasibility represents one of the key points and has been investigated in the past, proving that an endovascular approach may not only be restricted to rare, isolated cases (15). However, open surgical treatment still represents the gold standard of care and should be carried out, if possible (2). The overall aim to close/resect the entry tear to avoid aortic rupture and resolve organic malperfusion should be kept in mind, making no compromises (16). Especially due to the complex pathology of ATAAD including involvement of the aortic root, the coronary arteries, and additional branches of the aortic arch, endovascular approaches remain very controversial at this time. Most of the published data refer to cases with favorable anatomy and/or locally restricted disease processes, which significantly complicates the transfer of treatment concepts to more complex cases (17). Adding an uncovered nitinol stent for the aortic arch according to the PETTICOAT technique to minimize the risk of true lumen collapse and enhance the remodeling process of the dissected aorta is of upmost importance, but a rather individual approach according to this case (11). However, this "reverse PETTICOAT technique" may be a useful adjunct, especially considering recent results of the STABLE Trial that demonstrated favorable clinical and anatomical results as well as positive aortic remodeling when adding a distal bare metal stent (18). Of specific concern is the anchoring of the device in within the proximal landing zone (ascending aorta). Evolving concepts like the "Endo Bentall approach" published by the Freiburg group to address the challenges of aortic insufficiency and aortic root dissection are promising, but are still experimental (19). The choice of the stent graft as well as the degree of oversizing in the ascending aorta is one of the most difficult parts. In our case, proximal oversizing rate was 15%. Furthermore, available stent graft systems for the ascending aorta are scarce and restricted to off-label use, especially since Valiant Navion™ (Medtronic®, Dublin, Ireland) got removed from the market. Additional systems that can be used for the ascending aorta may be GORE® C-TAG® (GORE®, Flagstaff, AZ, USA), ZENITH TX2 (Cook, Bloomington, IN, USA), Relay NBS (Bolton, Barcelona, Spain; now Terumo®, Inchinnan, UK), and Lamed Ankura® (Lamed, Oberhachingen, Germany), wherein the latter may be used with caution due to the proximal extended stent tip (13).

Different suggestions have been made for the minimum size of the landing zone in the ascending aorta and the safe distance to the coronary ostia, which vary between 10 and 40 mm (14). However, there is general agreement that new short-length stent graft devices are needed to address the anatomical challenges of the ascending aorta. Though we are able to demonstrate a successful endovascular approach for the treatment of complex ATAAD in DeBakey type 1 dissection with preoperative malperfusion, the aortic root including the aortic valve was not involved. Furthermore, no pericardial tamponade was evident, which would have been a major influencing factor for our strategy. These findings are in common with results from other groups (13). Therefore, concepts like the "Endo Bentall approach" are of particular interest for the future and may have great potential to treat elderly high-risk patients who are deemed to unfit for open surgery (19). Nevertheless, we are able to show stable results without any disease progress after 3.5 years of follow-up. In a multicenter series with 12 patients, Nienaber et al. could also detect no case of endoleak occurring during the whole follow-up period (13). These results are promising and demonstrate the potential sustainability of this approach for highly selected cases.

Data availability statement

The original contributions presented in the study are included in the article/Supplementary Material, further inquiries can be directed to the corresponding author.

Ethics statement

Ethical approval was not required for the study involving humans in accordance with the local legislation and institutional requirements. Written informed consent to participate in this study was not required from the participants or the participants' legal guardians/next of kin in accordance with the national legislation and the institutional requirements. Written informed consent was obtained from the individual(s) for the publication of any potentially identifiable images or data included in this article. Written informed consent was obtained from the participant/patient(s) for the publication of this case report.

Author contributions

LP: Conceptualization, Data curation, Investigation, Methodology, Validation, Visualization, Writing – original draft, Writing – review & editing. RH: Data curation, Supervision, Writing – review & editing. MM: Supervision, Writing – review & editing. AP: Supervision, Writing – review & editing. MK: Supervision, Writing – review & editing. VF: Supervision, Writing – review & editing. JK: Supervision, Validation, Writing – review & editing. SB: Conceptualization, Data curation, Methodology, Supervision, Validation, Writing – review & editing.

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Conflict of interest

MM has received speaker fees and travel grants from Artivion® (Hechingen, Germany). VF has received educational grants, including travel support, fees for lectures and speeches, as well as for professional consultation and research, along with study funds from Medtronic® GmbH (Dublin, Ireland) and Artivion® (Hechingen, Germany). JK is a proctor for and has received travel support and speaker fees from Artivion® (Hechingen, Germany), Medtronic® GmbH (Dublin, Ireland), JOTEC GmbH (now Artivion®, Hechingen, Germany), and Vascutek/Terumo® (Inchinnan, UK).

The remaining authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

Publisher's note

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Appendix

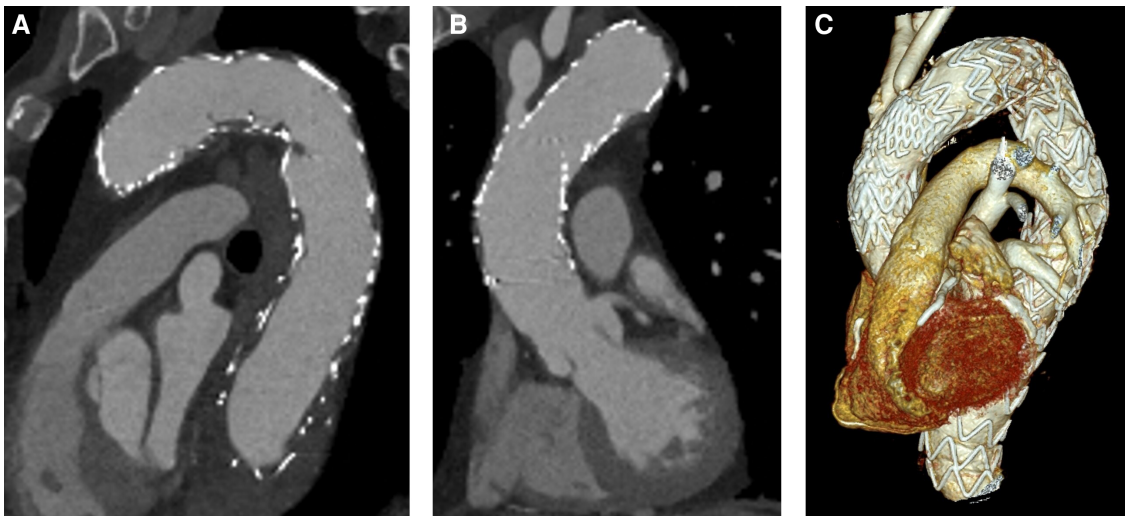

FIGURE A1
CTA follow-up 3.5 years later. (**A**) No endoleak in the aortic arch. (**B**) No disease progress in the ascending aorta. (**C**) 3D reconstruction.

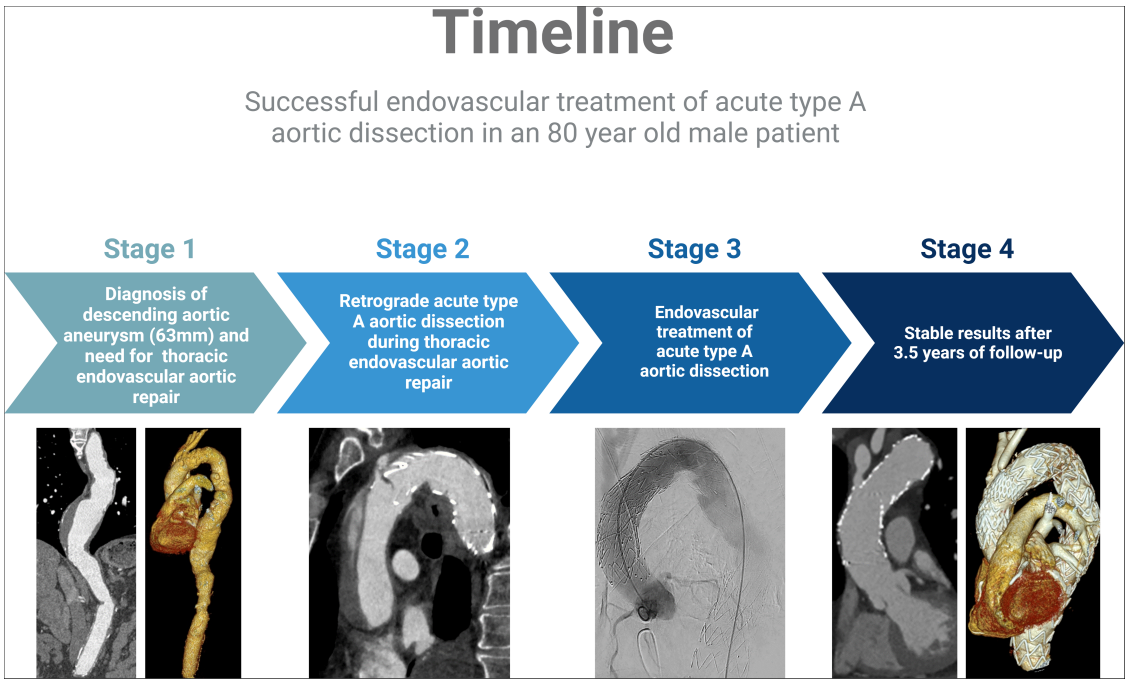

FIGURE A2
Timeline describing the patient's course including the relevant events.

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Case Report: Transcatheter treatment of aortic coarctation in a 58-year-old patient with LACHT syndrome and left lung agenesis

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LACHT (Lung Agenesis, Congenital Heart, and Thumb anomalies) syndrome is an extremely rare congenital anomaly and presents significant challenges in adults due to its poor survival rates. Herein, we report a case of late diagnosis and successful transcatheter treatment of aortic coarctation in a 58-year-old male patient with LACHT syndrome, medically resistant arterial hypertension, and left lung agenesis. Baseline CT angiography showed isthmic aortic coarctation and left lung agenesis, with compensatory right pulmonary artery and vein thickenings. The patient underwent balloon dilation and subsequent implantation of a covered NuMED 45 mm 8-ZIG CP stent with satisfactory outcomes. The pressure gradient decreased from 43 to 23 mmHg. The arterial pressures normalized during the follow-up with fewer medications. Genetic testing identified a heterozygous mutation (c.6583C > T) in the FBN2, supporting the diagnosis of variant Marfan syndrome.

KEYWORDS
LACHT syndrome, aortic coarctation, lung agenesis, aorta covered stent, hypertension

Introduction

LACHT (Lung Agenesis, Congenital Heart, and Thumb anomalies) syndrome, also known as the Mardini Nyhana association, is an exceedingly rare and medically serious congenital disorder (1). To date, only 13 documented cases of LACHT syndrome exist worldwide, with one instance leading to prenatal diagnosis and pregnancy termination at 36 weeks (2). Among these cases, only two have reported cardiac malformations, including aortic coarctation (2). Notably, no documented cases of lung agenesis associated with aortic coarctation have survived into adulthood. Furthermore, information on the genetic basis of LACHT syndrome remains limited, with no specific pathogenic gene identified to date. In this report, we present a case of late diagnosis and successful transcatheter treatment of aortic coarctation in a 58-year-old male with LACHT syndrome. Familial genetic testing revealed a heterozygous mutation (c.6583C > T) in FBN2, supporting the diagnosis of a variant of Marfan syndrome.

Case presentation

A 58-year-old male patient was transferred to our institution for rapidly worsening medically-resistant arterial hypertension and shortness of breath. The patient also complained of dizziness and weakness in both lower limbs, with occasional symptoms of blurred and darkening vision, lower limb edema, and syncope. The patient had a history of left

pulmonary agenesis that was diagnosed at the age of 8 years old without any further investigation. The patient has been taking antihypertensive drugs irregularly in the past until it was adjusted recently to include amlodipine 5 mg once daily, Irbesartan 150 mg once daily, Metoprolol 12.5 mg twice daily, hydrochlorothiazide 12.5 mg once daily, and Terazosin 2 mg once nightly. One month after adjusting the medication, the blood pressure remained uncontrolled. The patient had a body mass index of 20.5 kg/m^2, was not a smoker, and reported moderate alcohol consumption. Family history showed that his mother had mild arterial hypertension. Physical examination showed an arm-leg systolic pressure gradient of 40 mmHg and a grade 2/6 vascular murmur in the left scapular area. Physical abnormalities included funnel chest, mild collapse of the left chest wall, and the absence of breathing sounds in the left lung. Upon admission, serum creatinine, homocysteine, B-type natriuretic peptide, and low-density lipoprotein cholesterol were mildly elevated, while thyroid function and blood sugar were normal. Echocardiography showed a left atrium diameter of 43 mm, left ventricle diameter of 56 mm, interventricular septum thickness of 12 mm, left ventricular posterior wall thickness of 12 mm, and left ventricular ejection fraction of 61%. Computed tomographic thoracic imaging was

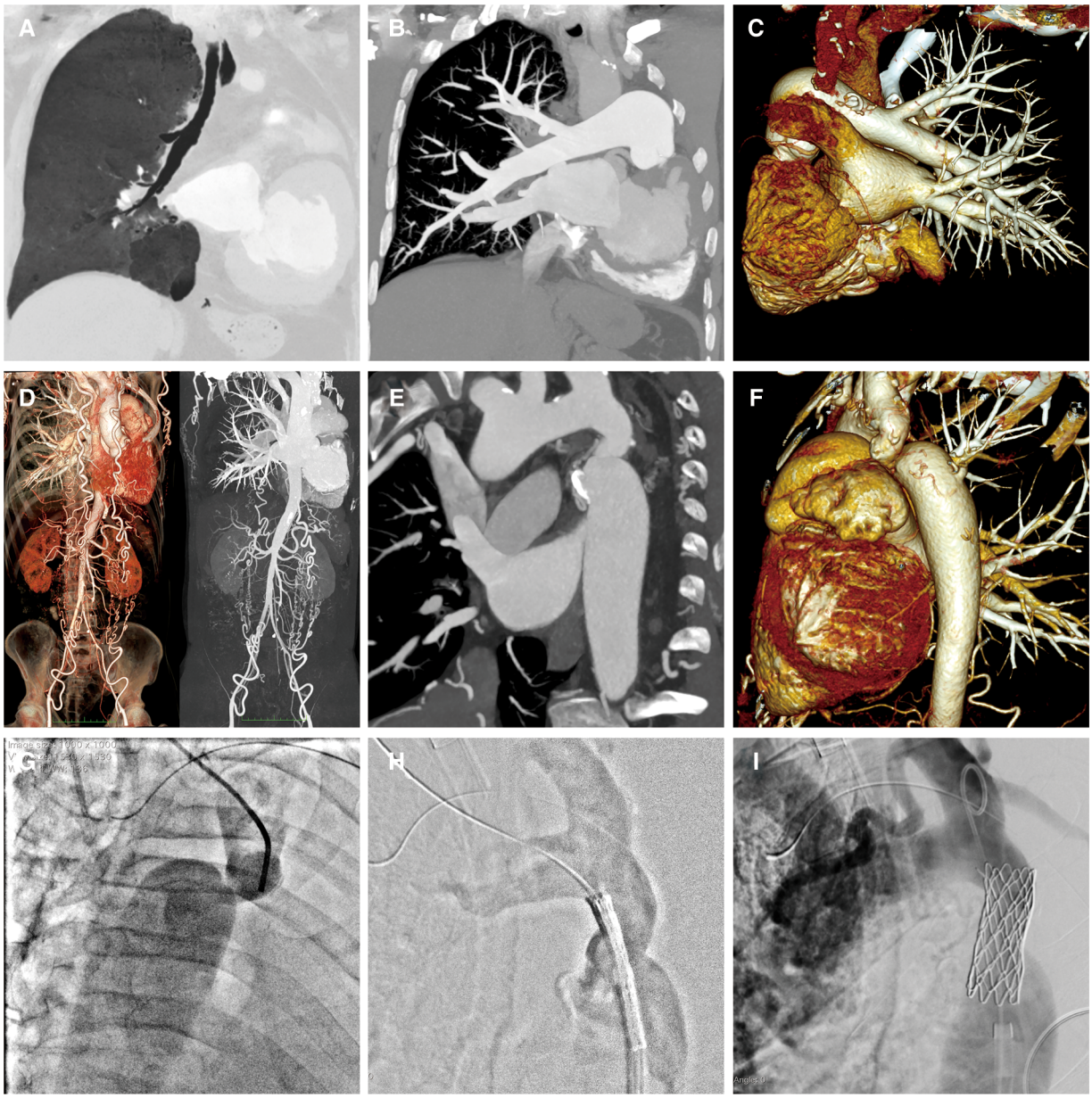

FIGURE 1
A-C: Pulmonary artery CTA reveals the absence of the left lung structure, with no display of the left pulmonary artery and its branches. The right lung structure and pulmonary artery show no filling defect, and the branches of each lobe segment artery are well displayed with compensatory dilatation. **D-F**: Aortic CTA shows localized narrowing at the beginning of the descending aorta and branches are formed at the distal ends of the subclavian artery, intercostal arteries, intrathoracic arteries, and both iliac arteries. **G-I**: Aortic angiography assesses the degree of narrowing, locates the position of the balloon-expandable covered stent, and evaluates the post-release effects.

performed and revealed left pulmonary agenesis (Figures 1A–C) along with a severe isthmic coarctation of the aorta just distal to the dilated left subclavian artery. There was abundant collateral circulation bypassing the coarctation in the suprascapular, internal thoracic, and superficial abdominal arteries (Figures 1D–F). 24 h ambulatory blood pressure monitoring showed non-dipper hypertension. Pulmonary function tests showed mixed ventilation dysfunction with predominant moderate to severe obstruction. Renin, angiotensin, and aldosterone levels were within the normal range at three-time points (0:00, 8:00, 16:00) in both standing and lying positions. The rheumatoid factor test showed no specific positive indicators. Carotid artery ultrasound, renal artery ultrasound, and adrenal CT scan showed no significant abnormalities. A coronary angiogram showed no significant stenosis in the left anterior descending artery or circumflex artery, and an aneurysmal-like dilation at the opening of the right coronary artery.

With the consent of the patient and family, and approval from the hospital ethics committee, a whole-exome sequencing family genetic screening was performed, and revealed a heterozygous mutation in FBN2, c.6583C > T supporting the diagnosis of variant Marfan syndrome, with the mother being wild-type and her son showing the heterozygous mutation (Figure 2).

A multidisciplinary decision was made to treat this patient with transcatheter stent implantation under general anesthesia. The left femoral vein was accessed to place a temporary cardiac pacemaker. The right radial arterial access was then obtained to measure the pressures and perform invasive angiography. A 14F Cook sheath was introduced through the right femoral artery, followed by aortic dilation with a Fortrex 6 × 40 mm balloon. Subsequently, a 45 mm covered 8-Zig CP-stent (NuMED, USA) was mounted on a 13 × 40 mm balloon and implanted in the initial segment of the descending aorta (Figures 1G–I). Angiography confirmed the safe adhesion and secure placement of the stent. There was a significant reduction in the pressure gradient from 43 to 23 mmHg.

Post-procedural monitoring did not reveal any abnormalities in renal function. A repeat aortic CT angiography was performed one-month post-procedure and showed stable results. Eighteen months post-procedure, the patient's blood pressure remained stable within the normal range under two antihypertensive drugs, and limb symmetry was restored. A 6 min walking test of 530 meters was conducted. There was no change in echocardiography findings compared to hospitalization, and the left ventricular ejection fraction was within the normal range.

Discussion

LACHT syndrome, or Mardini-Nyhan association, is an extremely rare disorder and is characterized by complex cardiac vascular malformations and skeletal abnormalities (2, 3).

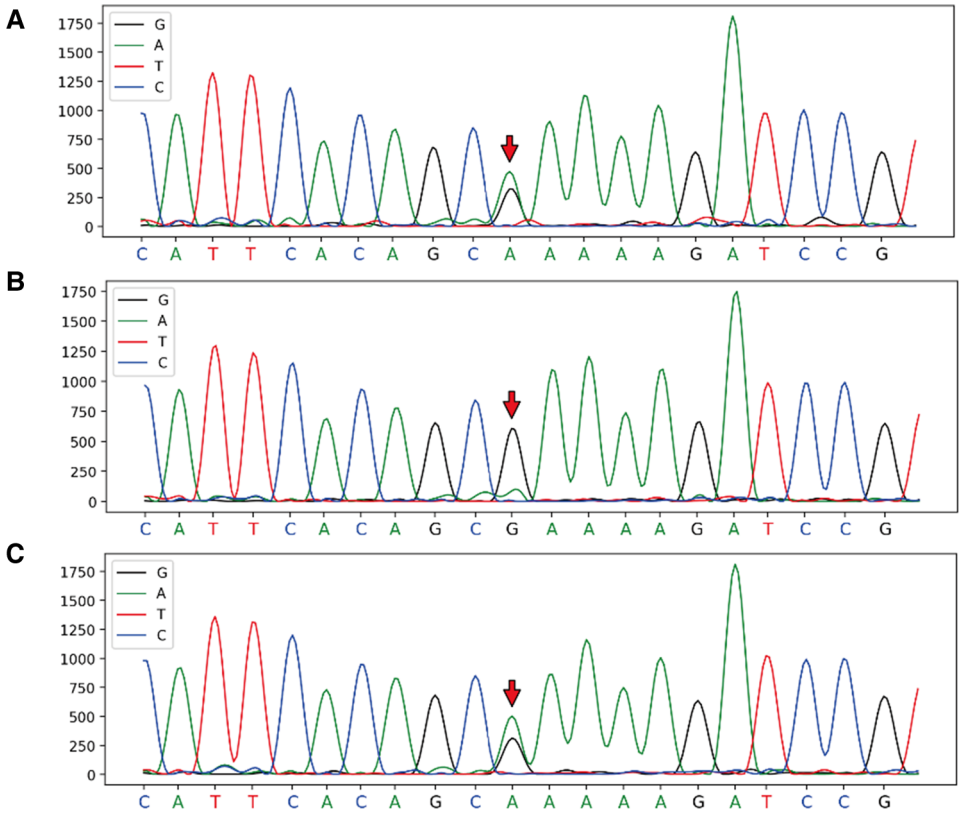

FIGURE 2
Full exon gene sequencing first generation sequencing family verification chart verification site: FBN2, c.6583C > T, chromosome position: chr5:127624873. (A) Proband: heterozygous mutation at chr5:127624873; (B) Mother: wild-type at chr5:127624873; (C) Son: heterozygous mutation at chr5:127624873.

Pulmonary agenesis is a rare congenital developmental defect and is classified into three types according to the complete or partial absence of the lung parenchyma, bronchus, and pulmonary artery (1, 4). Nearly half of the cases of pulmonary agenesis are accompanied by other congenital abnormalities affecting various systems (2). The clinical manifestations of pulmonary agenesis can vary from asymptomatic cases to respiratory conditions such as dyspnea and respiratory distress. Our patient presented with type 1 pulmonary agenesis (absence of lung parenchyma, bronchus, and pulmonary artery) and was diagnosed with chronic obstructive pulmonary disease.

Coarctation of the aorta is a congenital condition that accounts for 5% to 10% of all congenital vascular malformations (5). Marfan syndrome (MFS) is an autosomal dominant connective tissue disorder that affects the ocular, skeletal, and cardiovascular systems. Aortic tear and rupture are the leading causes of death in individuals with MFS. In our patient, family gene screening revealed a heterozygous mutation at the FBN2 gene, specifically c.6583C > T. This mutation is consistent with the inheritance pattern of a dominant disease (6) and has been associated with a variant Marfan syndrome (7).

Although the diagnosis of aortic coarctation is not challenging, it can be overlooked, particularly in elderly patients who may be diagnosed solely with essential hypertension without undergoing comprehensive and meticulous examinations. The patient's delayed diagnosis, survival, and absence of significant target organ damage can be attributed to the development of the aorta with marked collateral circulation (8). The patient suffered from congenital hypoplasia of the left pulmonary vein, resulting in compensatory dilation of the right pulmonary vein and blood draining into the left atrium. We hypothesize that the increase in left atrial volume can be compensated, resulting in a compensatory increase in left ventricular volume. This situation leads to the volume load of the left ventricle entering the aorta, which can also be compensated. Consequently, the aortic vessels experience vascular remodeling due to increased pressure load caused by aortic constriction. Simultaneously, compensatory collateral circulation forms at the distal end of the aortic constriction segment, mitigating vascular remodeling and ischemic tissue damage caused by volume and pressure load. Thus, the initial rise in blood pressure is not substantial, and organ damage remains within a compensatory range. Patients display mild ischemic symptoms and signs exclusively, and the progression of hypertension-related organ damage is relatively slow.

The treatment options for aortic coarctation include surgical and transcatheter repair (9, 10). The patient has one lung agenesis, and the risk of surgical anesthesia was high. We considered treating his coarctation percutaneously with a covered stent to be less invasive and aggressive. However, the patient presented a significant aortic dilation near the proximal end of the constriction with a substantial difference in diameter compared to the distal narrowed portion. Furthermore, the constriction was located at a bend in the vessel, making interventional treatment challenging. We considered the stent length and balloon diameter according to baseline anatomy using the standard approach (10). Following expansion, the stent adhered well to the vessel wall at the level of the narrowing segment. Despite some procedural and technical advantages of other new-generation endovascular stents (11), we have selected the CP stent because of the availability in the armamentarium and operators' experience with CP stents. Even though the residual pressure gradient was not optimal, significant control over blood pressure was achieved with medications. We refrained from further expansion or additional stent placement to avoid the risk of vessel tearing due to excessive expansion (11). Subsequent follow-up showed that the patient's blood pressure was normalized with medication, supporting the correctness of our comprehensive decision.

Conclusion

We present the case of a 58-year-old male patient with LACHT syndrome with a late diagnosis and treatment of aortic coarctation. The patient underwent successful aortic balloon angioplasty and stent implantation, resulting in better control of normal blood pressure and symptom relief. Genetic testing revealed a variation in the FBN2 gene 253 (c.6583C > T), supporting the diagnosis of variant Marfan syndrome.

Data availability statement

The original contributions presented in the study are included in the article/Supplementary Material, further inquiries can be directed to the corresponding author/s.

Ethics statement

The studies involving humans were approved by Ethics Committee of the First People's Hospital of Zunyi. The studies were conducted in accordance with the local legislation and institutional requirements. The participants provided their written informed consent to participate in this study. Written informed consent was obtained from the individual(s) for the publication of any potentially identifiable images or data included in this article.

Author contributions

QT completed literature search and article writing. NH, FG, ML, MX and JL assisted in the collection of materials and related data and pictures. NH and JL assisted in the collation of pictures and the modification of the article. ZY and QJ designed the ideas of the article and the modification of the article. All authors contributed to the article and approved the submitted version.

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Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

Publisher's note

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Case Report: Kommerell's diverticulum and left aberrant subclavian artery stenosis hybrid treatment with branched aortic stent-graft

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Kommerell's diverticulum in association with left or right aberrant subclavian arteries is a rare finding and is challenging to treat. Contemporary surgical and endovascular techniques provide a broad arsenal of possible treatments. Imaging techniques and modeling technology allow a more personalized strategy for each patient. In this case, we present a symptomatic patient with a Kommerell's diverticulum and a left aberrant subclavian artery complicated by proximal stenosis and poststenotic aneurysm. A hybrid technique using a single-branched thoracic stent-graft (Castor, MicroPort Medical, Shanghai, China) in combination with a surgical left subclavian-carotid bypass and endovascular occlusion of the poststenotic aneurysm using a vascular plug device (Amplatzer Vascular Plug, Abbott, Chicago, United States) was performed. This approach was planned and facilitated by the use of a 3D model. Alternative treatment options and the strengths of this approach are briefly reviewed and discussed.

KEYWORDS

Kommerell's diverticulum, aberrant subclavian artery, subclavian steal syndrome, branched aortic stent-graft, right aortic arch, hybrid endovascular treatment

1. Introduction

Kommerell's diverticulum is a rare vascular anomaly of the aortic branches, a persistent remnant of the fourth primitive dorsal arch, often associated with an aberrant subclavian artery. It can be present in both the left or right aortic arches, with aberrant right or left subclavian arteries, respectively (1). The prevalence of Kommerell's diverticulum and aberrant subclavian arteries is 0.4%–2.3% (2). Kommerell's diverticulum can be classified into three types according to the relationship with the subclavian artery; Kommerell's diverticulum in the left aortic arch with right aberrant subclavian artery (ASCA), Kommerell's diverticulum in the right aortic arch with left ASCA, and aortic diverticulum without ASCA (3).

The mean size of Kommerell's diverticulum at diagnosis ranges from 20 to 30 mm, with a mean growth rate of 1.5 mm/year in different series, and dissection or rupture have been widely described (3, 4). The clinical spectrum associated with this anomaly is wide and mainly depends on the trajectory and permeability of the aberrant subclavian artery; it can produce compressive symptoms including dysphagia, dyspnea, recurrent laryngeal nerve palsy, claudication, or even left subclavian steal syndrome if significant left subclavian artery stenosis is present. Other complications such as thrombosis and lower extremity emboli have also been described (3, 4).

Treatment indication has been suggested when the subclavian artery is aneurysmatic (>30 mm at the level of the diverticulum's orifice) and/or when the Kommerell's diverticulum measures more than 50 mm, measuring the cross-sectional area of the confluence with the descending aorta (1, 5). Treatment is also recommended in symptomatic patients regardless of size (2). Surgical, endovascular, and hybrid treatment approaches have been reported (5–9).

In this case, we present a symptomatic patient with a Kommerell's diverticulum and a left ASCA complicated by proximal stenosis and poststenotic aneurysm, treated using a hybrid technique. A novel single-branched thoracic stent-graft (Castor, MicroPort Medical, Shanghai, China) was used in combination with surgical left subclavian-carotid bypass and endovascular occlusion of the poststenotic aneurysm using a vascular plug device (Amplatzer Vascular Plug, Abbott, Chicago, United States). This approach allows less invasive treatment based on hybrid techniques and uses a commercial, non-customized graft that can be easily reproduced in other centers. Planning and monitoring were facilitated by using a 3D model obtained from an ECG-gated CT angiography.

2. Case presentation

A 41-year-old male patient with a past medical history of dyslipidemia and without other cardiovascular risk factors presented with a ten-year history of left arm positional claudication, without vertebrobasilar associated symptoms. There was no history of familiar cardiovascular disease.

Physical examination found symmetrical radial pulses, a lower blood pressure in the left arm (120/90 mmHg) compared to the right arm (140/95 mmHg), no heart or vascular murmurs, and a lack of neurological semiology.

A supra-aortic vessel Doppler ultrasound was performed. There was no atheromatosis and flow velocities were normal in the right vessels and left carotid artery. However, the left subclavian artery flow was blunted, with a proximal aliasing area up to 170 cm/s and a posterior blunted flow of 50 cm/s. In addition, the left vertebral artery flow was inverted, with a velocity of 48 cm/s, and was not modified by left arm hyperemia.

An ECG-gated CT angiography showed a right-sided aortic arch with normal diameters and four supra-aortic vessels, in the following order of origin: left common carotid artery, right common carotid artery, independent right subclavian artery, and aberrant left subclavian artery. The left subclavian artery origin was located in the descending thoracic aorta in relation to an aneurysmatic diverticulum in the left aortic wall, suggestive of a Kommerell's diverticulum with a maximum diameter of 30 mm. Immediately after the origin, the left subclavian artery had critical stenosis and a post-stenosis saccular aneurysm of 18 mm. The remaining left subclavian artery had a smaller diameter compared to the right subclavian artery, and the left vertebral artery was normal.

After these findings, a diagnosis of Kommerell's diverticulum in the right aortic arch with left aberrant subclavian artery and symptomatic critical stenosis and post-stenosis saccular aneurysm was made.

The treatment proposed by a multidisciplinary cardiovascular team was exclusion of the Kommerell's aneurysm and the post-stenotic saccular aneurysm to reduce the risk of rupture and treatment of the left subclavian artery critical stenosis to reduce the claudication symptoms. A 3D model of the aorta was obtained to better study the case (**Figure 1**). Normal ventricular and valvular function was found on preoperative transthoracic echocardiogram. Open surgery was evaluated as the first treatment option; however, after carefully studying the case, a surgical approach via left or right thoracotomy could guarantee proximal and distal control of the descending aorta. Considering the anatomical limitations and, therefore, the high surgical risk, a hybrid approach was preferred.

A left carotid-subclavian bypass followed by exclusion of the Kommerell's diverticulum using a one-branch thoracic endograft (Castor, MicroPort®) and exclusion of the post-stenotic saccular aneurysm using an Amplatzer Vascular Plug II was planned. The approach was explained to the patient and informed consent to treatment was obtained.

Under general anesthesia, the left supraclavicular approach was used to perform a left carotid-subclavian bypass with a polytetrafluoroethylene (ePTFE) heparin-coated 6 mm ringed vascular prosthetic graft. The patient woke up without complications. One day after, the endovascular procedure was performed under general anesthesia. Open surgical access was performed for the right axillary artery and left humeral artery, and percutaneous access was performed for both femoral arteries. The through and through technique was used from the right femoral access to the right axillary access to facilitate the placement of the endograft and the deployment of the branch in the right subclavian artery. During deployment maneuvers, the endograft advanced slightly forward, partially occluding the ostium of the right common carotid artery. This was solved by performing distal traction of the endograft with a Reliant® balloon, managing to withdraw it a few millimeters. A lengthening of the coverage of the right subclavian artery was performed to avoid an excessive angulation using a 10 × 27 mm covered stent (iCover, iVascular, Barcelona, Spain). Finally, from the left humeral access, a 14 mm Amplatzer Vascular Plug II was placed, excluding the Kommerell's diverticulum and the saccular aneurysm of the left subclavian artery (**Figure 2**, intraoperative images provided in **Supplementary Image S1–S4**).

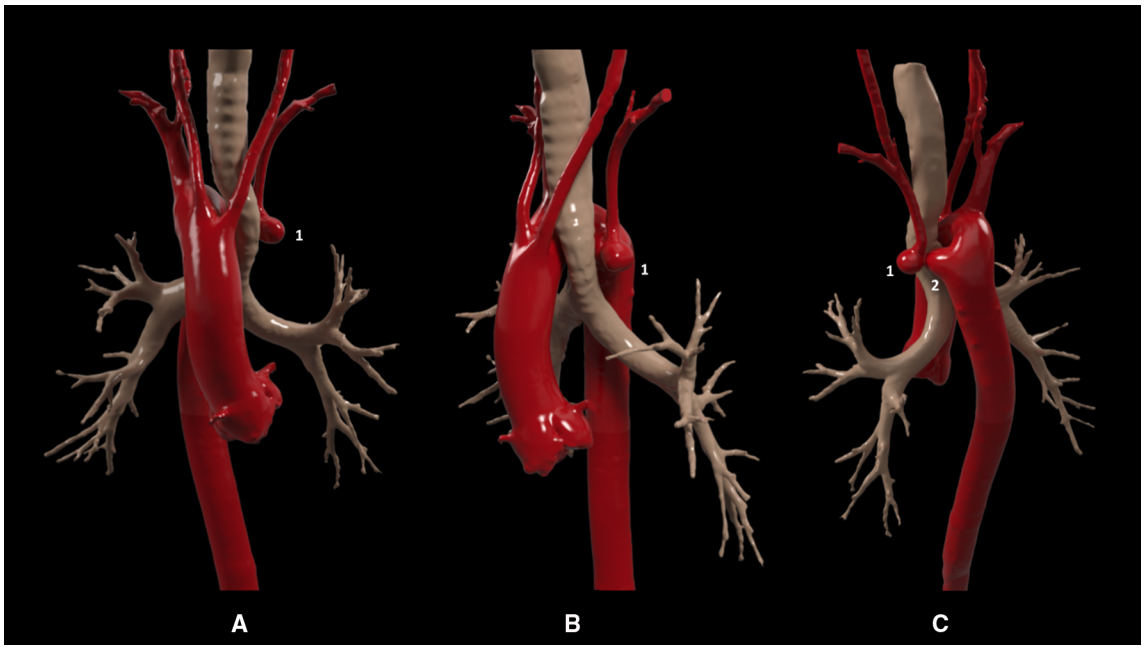

FIGURE 1
Pre-interventional 3D model. Anterior (**A**), lateral (**B**), and posterior (**C**) views showing a right-sided aortic arch and supra-aortic vessels, in the following order of origin: the left common carotid artery, right common carotid artery, independent right subclavian artery, and finally an aberrant left subclavian artery with critical proximal stenosis and post-stenosis saccular aneurysm (1) in relation to a Kommerell's diverticulum (2).

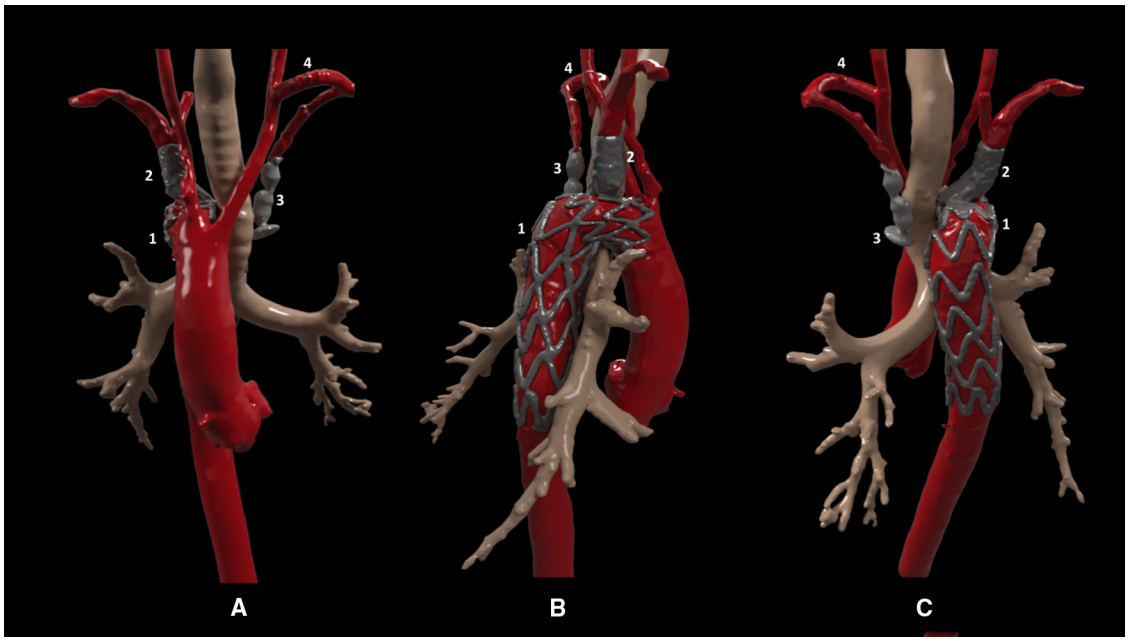

FIGURE 2
Post-interventional 3D model. Anterior (**A**), lateral (**B**), and posterior (**C**) views showing (1) the one-branch thoracic endograft (Castor, MicroPort®) in aorta with the branch placed in the right subclavian artery, extended with a covered stent, (2) Amplatzer Vascular Plug II, (3) in the left subclavian artery excluding Kommerell's diverticulum the poststenotic saccular aneurysm (therefore not visible in the model), and (4) left carotid-subclavian extra anatomical bypass.

There were no immediate postoperative complications. Dual antiplatelet treatment was started with aspirin and clopidogrel and the patient was discharged on the fourth day of hospitalization.

The left arm claudication disappeared and a control ECG-gated CT angiography was performed 1 month after the intervention, which showed the complete thrombosis of the Kommerell's diverticulum and the post-stenotic aneurysm,

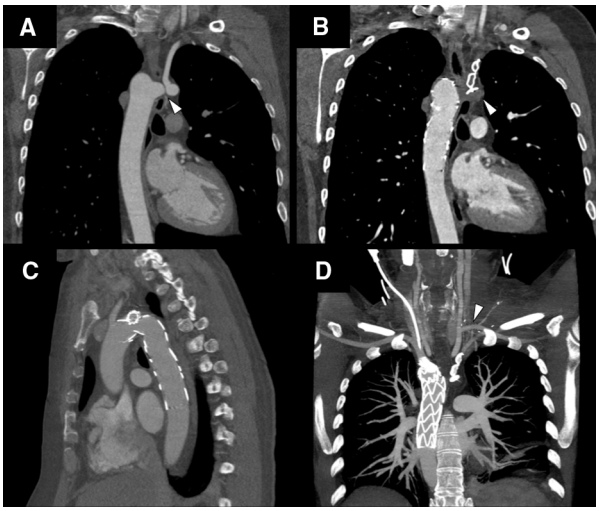

FIGURE 3
ECG-gated CT angiography panel. (**A**) Pre-interventional image, Kommerell's diverticulum and aberrant left subclavian with critical stenosis and poststenotic saccular aneurysm (white arrowhead). (**B**) Post-interventional image, complete exclusion and thrombosis of Kommerell's diverticulum and poststenotic aneurysm (white arrowhead). (**C**) Branched thoracic endograft (Castor, MicroPort®), correct alignment without endoleaks or thrombotic complications. (**D**) Maximum intensity projection (MIP) image showing supra-aortic vessel permeability. From left to right as seen in the image; right subclavian artery, right carotid artery, left carotid artery, extra anatomical bypass from the left carotid to left subclavian artery (white arrowhead), and native left subclavian artery excluded with an Amplatzer Vascular Plug II device.

the perfusion of all the supra-aortic branches was correct, and the endograft was well positioned without endoleaks or thrombotic complications (**Figure 3**). After 6 months of follow-up, the patient is still under dual antiplatelet therapy and free of left arm claudication symptoms.

3. Discussion

While Kommerell's diverticulum is a rare entity nowadays, incidental diagnosis of vascular anomalies is increasing due to the extensive use of a variety of cardiovascular imaging techniques. Treatment indication therefore requires a careful clinical and anatomical evaluation to determine if symptoms are caused by the anomaly and to determine the risk of complications. In this case, the correlation between claudication symptoms, the physical examination, and the ultrasound evaluation, along with the ECG-gated CT angiography information was clear and led to treatment indication besides the diverticulum's diameter.

As shown before, treatment indication for ASCA and Kommerell's diverticulum has been suggested in the American Heart Association (AHA) 2022 guidelines in the presence of symptoms related to the ASCA or a diameter of 30 mm at the level of the orifice of the diverticulum (1). According to the guidelines of the European Association for Cardio-Thoracic surgery (EACTS) and the European Society for Vascular Surgery (ESVS), the surgical approach for an ASCA and Kommerell's diverticulum should include the removal of the aberrant artery, with further subclavian-carotid transposition or bypass, the excision of the diverticulum, and proper aortic reconstruction with graft replacement of the descending aorta or total arch replacement (2, 6). Multiple surgical approaches have been described depending on the patient's anatomy and the presence of concomitant anomalies (7); however, there are still cases where the anatomy of the aortic arch and the descending aorta limits the surgical approach. In these cases, hybrid techniques combining surgical and endovascular approaches have been reported; the frozen elephant trunk technique in association with a TEVAR placement and coiling of the diverticulum with subclavian-carotid transposition or bypass and endovascular techniques with custom-designed endovascular grafts to exclude the ASCA and diverticulum have also been reported (8, 9). In reviewing the literature, less data is available regarding endovascular techniques with commercial branched endovascular grafts.

In this case, the hybrid approach of left carotid-subclavian bypass with exclusion of the diverticulum using a commercial one-branch thoracic endograft (Castor, MicroPort®) associated with exclusion of the post-stenotic saccular aneurysm using a vascular plug is an alternative exportable and efficient approach to treat these patients.

The approach reproducibility is limited, in any case, to each patient's anatomy, and requires a detailed evaluation and planification of every therapeutic alternative. In the case we describe, 3D modeling permitted a better understanding of the relations between structures and the *in silico* planification of the hybrid procedure, along with traditional CT reconstructions.

This situation is, therefore, a good example of the trend toward a more personalized medicine that we are experiencing nowadays, which requires modern technology and a high level of coordination and cooperation between the different areas involved in the treatment of these patients, such as cardiovascular imaging professionals and vascular and cardiac surgeons.

Data availability statement

The original contributions presented in the study are included in the article/**Supplementary Material**, further inquiries can be directed to the corresponding author.

Ethics statement

Written informed consent was obtained from the individual(s) for the publication of any potentially identifiable images or data included in this article.

Author contributions

ÁR: Conceptualization, Writing – original draft. CT: Writing – review & editing. AV: Writing – review & editing. CF: Writing – review & editing. AH: Writing – review & editing. AG: Writing – review & editing. AB: Writing – review & editing. JD: Writing – review & editing.

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Publisher's note

Supplementary material

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SUPPLEMENTARY IMAGE S1
Intraoperative image showing a sharp angulated aortic arch with the folded branched thoracic endograft and the right subclavian branch deployed.

SUPPLEMENTARY IMAGE S2
Intraoperative image comparing the endograft position before (A) and after (B) performing a corrective distal traction to avoid partial occlusion of the right common carotid artery. The endotracheal tube (red line) is used as a reference.

SUPPLEMENTARY IMAGE S3
Intraoperative image after complete deployment of the endograft.

SUPPLEMENTARY IMAGE S4
Intraoperative images showing the extension of the endograft branch with a covered stent to avoid excessive angulation in the right subclavian artery. Initial incorrect apposition of the thoracic endograft branch in the right subclavian artery with excessive angulation (A). Coverage with a stent to the origin of the dominant right vertebral artery (B). Final image of the realigned endograft branch after covered stent implantation (C).

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Case Report: The application of amplatzer vascular plug to repair aortic dissection intimal tears and false lumen

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In recent years, significant advancements have been made in endovascular therapy for aortic dissection, resulting in the development of various treatment methods. Nevertheless, there is a contentious discussion regarding the suitability of different treatment methods for addressing the unique features of individual lesions. Specifically, the matter of whether the presence of small intimal tears or multiple distal tears requires extensive aortic coverage with graft stents, which may block the blood supply to important organs, intercostal arteries, or lumbar arteries, remains unresolved. Further research is necessary to determine the need for complex fenestrated or branched endovascular aortic repair (F/B-EVAR) in the management of chronic post-dissection aneurysms involving visceral arteries. Our report highlights the successful endovascular repair of three patients with aortic dissection, encompassing one Stanford A type and two Stanford B types. Utilizing the Amplatzer Vascular Plug (AVP) alone or in combination with the coil embolization technique, we achieved favorable clinical outcomes. This article aims to provide valuable insights and new perspectives on the tailored management of aortic dissection by conducting a comprehensive examination of the subtle differences in the treatment processes and techniques applied to these three patients.

KEYWORDS

endovascular treatment, embolization technique, amplatzer vascular plug, coils, aortic dissection

Introduction

Aortic dissection is a severe cardiovascular emergency (1), occurring at a rate of 2–3 cases per 100,000 individuals in China, leading to a mortality rate of 7.3% (2). Guidelines recommend various aortic replacement surgical techniques for Stanford Type A, while Type B is treated with endovascular aortic repair (EVAR) (3, 4). Advancements in technology prompt exploration of endovascular intervention for Type A. However, both open and endovascular surgeries involve intricate procedures with significant mortality rates (5). Hence, embolization is a technique that medical professionals carefully consider for treating commonly occurring intimal tears or stable chronic aortic dissection.

The Amplatzer Vascular Plug (AVP), a vascular occlusion device, is designed to optimize embolization in various endovascular surgery applications (6, 7). Available in four models, each tailored to different vascular anatomy and clinical scenarios, AVP's application for the direct occlusion of aortic dissection entry tears has limited study, and evidence supporting its safety and efficacy is insufficient. Our center selectively treated three patients with Stanford A and Stanford B type aortic dissection, using

TABLE 1 Demographics of 3 patients with chronic dissection treated with amplatzer vascular plug II.

		Patient 1	Patient 2	Patient 3
Age (years)		51	62	38
Sex		Female	Male	Male
Stanford type (A/B)		A	B	B
Previous aortic surgery		TEVAR for aortic dissection 2 years ago	F/B-EVAR for aortic arch and thoracic aorta 9 months ago	F/B-EVAR for thoracic and abdominal aorta 2 years ago
Tears position		ascending aorta; infrarenal abdominal aorta; right renal artery	abdominal aorta near the celiac artery; both iliac arteries	infrarenal abdominal aorta; right iliac artery
Surgical procedure	Step 1	Under general or general anesthesia, femoral artery access was performed.		
	Step 2	Intraoperative angiography is used to identify the number, location, and size of tears, determine the extent of involvement of the false lumen, and evaluate the blood supply to visceral branches.		
	Step 3	4 mm × 7 mm AVP II blocked ascending aorta tear	Coils embolize the narrowed false lumen	Procedure 1: 16 mm, 20 mm, and 22 mm AVP II were used to embolize the false lumen
	Step 4	7 × 25 mm Viabahn stent blocked right renal artery tear	14 mm, 18 mm, 20 mm, 22 mm AVP II were used to embolize the false lumen	Procedure 2: 8 mm × 6 mm AVP II blocked abdominal aorta tear
	Step 5	10 × 7 mm AVP Ⅱ blocked infrarenal abdominal aorta tear	Coils embolize the distal false lumen	Procedure 2: 20 × 16 mm AVP II and coils were used to embolize the false lumen
	Step 6			Procedure 2: 13 mm × 50 mm Viabahn stent and 10 mm × 80 mm Fluency stent in right iliac artery TL
Follow-up (months)		6	3	6
Outcome		Successful	Successful	Successful

TEVAR, thoracic endovascular aortic repair; F/B-EVAR, fenestrated or branched endovascular aortic repair; AVP II, amplatzer vascular plug II; TL, true lumen.

AVP II (Abbott, Minnesota, USA) to close intimal tears or combining it with coils for false lumen embolization (Table 1). Our initial idea was to use embolization techniques to reconstruct the aneurysm neck, addressing post-dissection aortic aneurysms involving visceral arteries. This approach aimed to avoid complex surgeries for preserving visceral arteries and to create conditions for standard EVAR procedures (Figure 1). To our delight, the results of the first surgery exceeded our expectations, with both patients showing complete thrombosis of the false lumen, eliminating the need for subsequent procedures in the true lumen. In this paper, we delve into the analysis of the treatment process and technical intricacies, aiming to provide valuable experiences and fresh perspectives for the surgical management of aortic dissection.

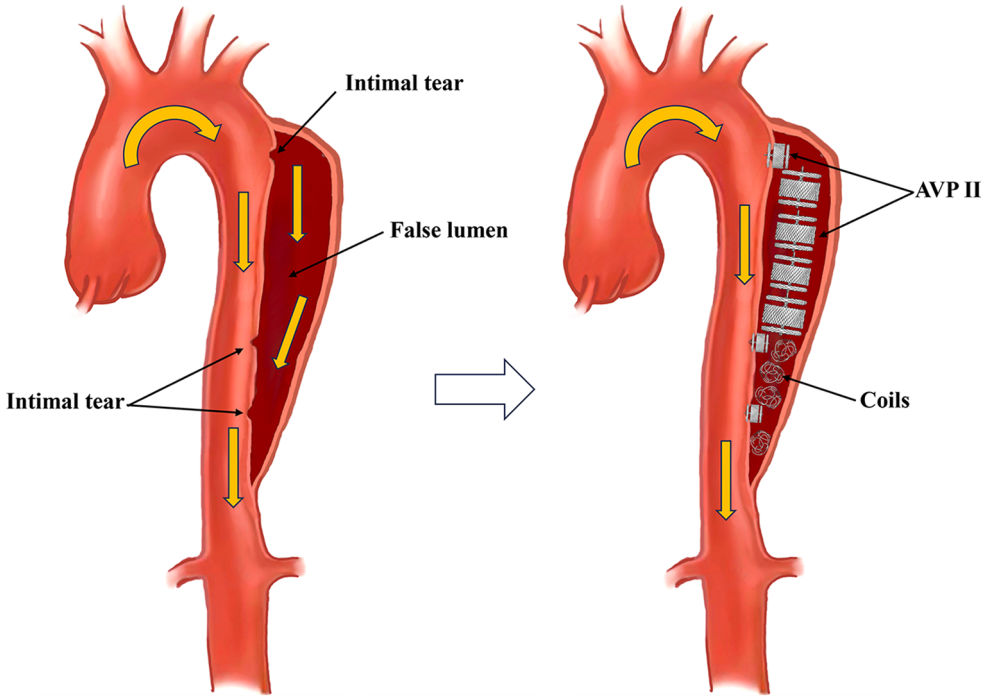

FIGURE 1
Aortic dissection treated using AVPs and coils.

Case report

Case 1: A 51-year-old female underwent thoracic endovascular aortic repair (TEVAR) for aortic dissection 2 years ago. The patient was readmitted because of persistent chest discomfort. Computed tomography angiography (CTA) revealed multiple tears at the origin of the ascending aorta and around the renal artery, with post-dissection aneurysm formation (Supplementary Figures S1A,C).

Under general anesthesia, a 6 F catheter was inserted through the right femoral artery to perform false lumen angiography in the ascending aorta (Figure 2A). A 4 mm × 7 mm AVP II was placed in the tear, with two parts in the false lumen and one in the true lumen. Aortography confirmed proper placement of AVP and absence of false lumen opacification (Figure 2B). Abdominal aortography used to find tear near renal artery (Figure 2C). A 7 × 25 mm Viabahn stent (W. L. Gore & Associates, Flagstaff, AZ, USA) was placed in the right renal artery to block the tear at the opening of the renal artery. Then, using a 7F guide catheter, we inserted an AVP II measuring 10 × 7 mm into the tear in the abdominal aorta. Aortography confirmed proper AVP placement with smooth blood flow in the right renal artery stent, and no false lumen opacification (Figure 2D). After 6 months, the follow-up CTA showed complete thrombosis in the false lumen of both the ascending and abdominal aorta without endoleaks. The AVP position remained stable. The abdominal aorta diameter decreased from 51 to 38 mm, while the true lumen diameter increased from 20 to 27 mm (Supplementary Figures S1B,D).

Case 2: A 62-year-old male underwent fenestrated or branched endovascular aortic repair (F/B-EVAR) for aortic arch and thoracic aorta 9 months ago for type B aortic dissection. Post-surgery CTA revealed a 57 mm Post-dissection abdominal aortic aneurysm extending to both iliac arteries. Multiple tears were located at the level of the celiac trunk and bilateral iliac arteries (Figure 3A).

To avoid complex endovascular techniques, the goal is to reconstruct the aneurysm's neck as a healthy anchoring site before implementing standard EVAR in the second phase. Under local anesthesia, the false lumen is accessed through an intimal tear in the left iliac artery (Figure 3B). Due to limited space, several Interlock mechanically detachable coils (Boston Scientific, Natick, MA, USA) were inserted at the level of the celiac trunk artery. After that, AVP II with diameters of 14 mm, 18 mm, 20 mm, and 22 mm were released sequentially in the false lumen. Finally, multiple coils (COOK, Bloomington, IN, USA) were placed at the

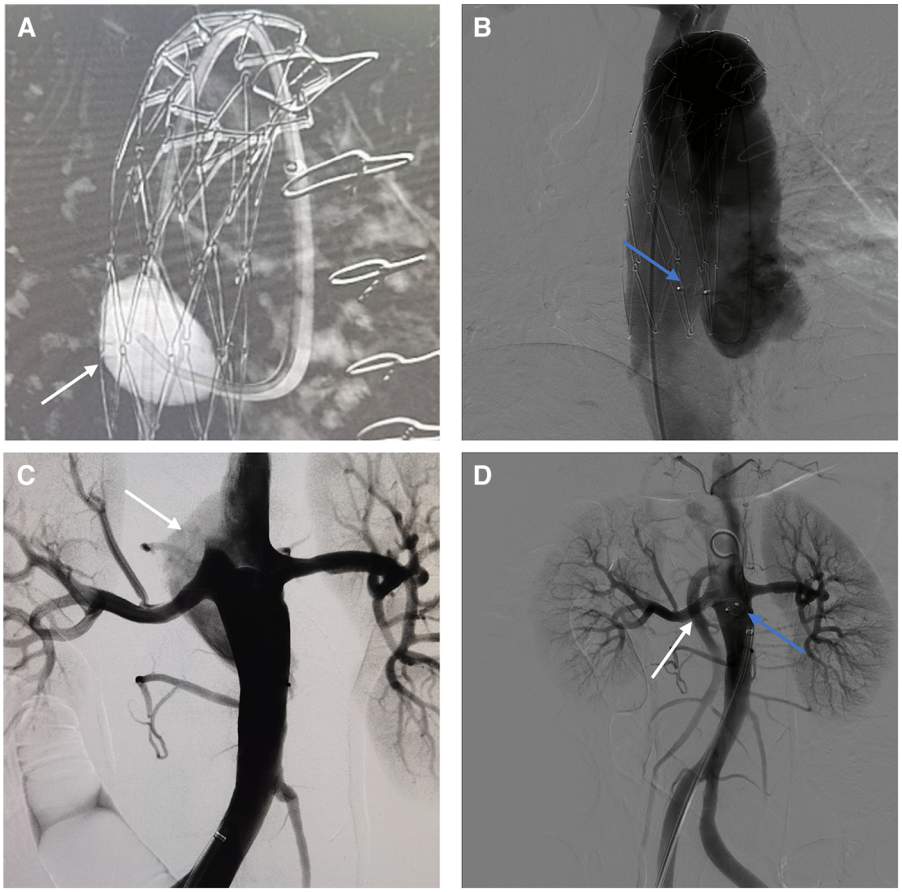

FIGURE 2
(A) Intraoperative imaging of aneurysm cavity. (white arrow) (B) Use AVP to embolize the tear of ascending aortic. (blue arrow) (C) The false lumen of the abdominal aorta. (white arrow) (D) Final intraoperative angiography showed the AVP (blue arrow) and the stent (white arrow).

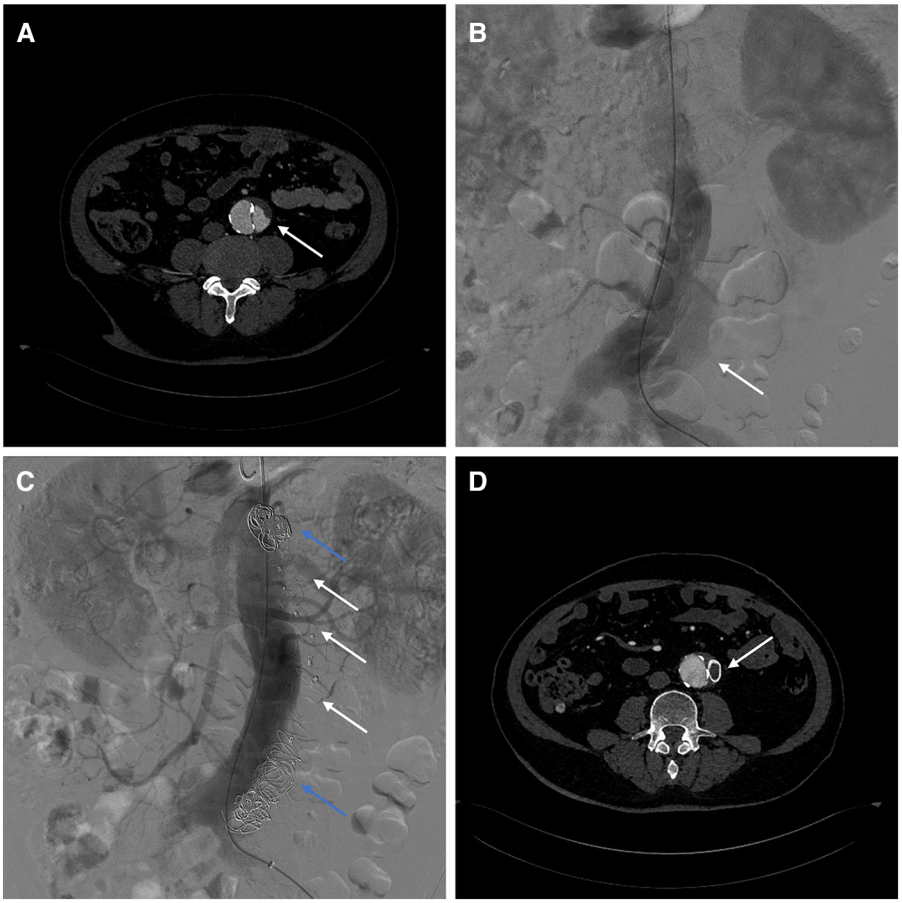

FIGURE 3
(A) Preoperative CTA showed blood flow in false lumen of aorta. (B) Intraoperative angiography showed the false lumen opacification in abdominal aorta. (C) Angiography showed no opacification of the false lumen after AVPs (white arrow) and coils (blue arrow) placement. (D) 3-month post-surgery CTA shows AVP in abdominal aorta.

distal false lumen to aid with embolization. Angiography confirmed no false lumen opacification (Figure 3C). Postoperative CTA examination after 3 months showed complete thrombosis of the false lumen in the abdominal aorta. The diameter of the abdominal aorta decreased from 57 mm to 47 mm, while the diameter of the true lumen increased from 22 mm to 37 mm (Figure 3D). Therefore, a second-stage EVAR surgery is not necessary.

Case 3: A 38-year-old male underwent F/B-EVAR for type B aortic dissection 2 years ago. Postoperative CTA follow-up: The patient has a post-dissection abdominal aortic aneurysm, with a maximum diameter of approximately 52 mm. The intimal tears are located at the level of the infrarenal abdominal aorta and the right common iliac artery respectively. All visceral arteries originate from the true lumen (Supplementary Figure S2A).

Procedure 1: Under local anesthesia, the false lumen was accessed via a tear in the right iliac artery (Figure 4A). Subsequently, AVP II with diameters of 16 mm, 20 mm, and 22 mm were placed to embolize the false lumen (Figure 4B). Intraoperatively, angiography revealed that several lumbar arteries arose from the false lumen. Since the therapeutic goal had been attained, the procedure was terminated to prevent acute spinal cord ischemia. The second phase of the surgery is scheduled to take place in a few months. Follow-up CTA revealed persistent blood flow in the mid to distal false lumen of the abdominal aorta, with distal narrowing, indicating the possibility of re-embolization therapy (Supplementary Figures S2B,C). Procedure 2: Under local anesthesia, right femoral artery access. Angiography showed post-dissection aortic aneurysm formation, and the tear was located in the infrarenal abdominal aorta and right iliac artery (Figure 4C). The 8F guiding catheter passed through iliac artery tear into false lumen, then entered true lumen via abdominal aorta tear. Placed 8 mm × 6 mm AVP II at tear site. A 20 × 16 mm AVP II was inserted into the stenosed false lumen, and Interlock mechanically detachable coils (Boston Scientific, Natick, MA, USA) was used to embolize the right internal iliac artery. Subsequently, a 13 mm × 50 mm Viabahn stent (W. L. Gore & Associates, Flagstaff, AZ, USA) and a 10 mm × 80 mm Fluency stent (Bard, Temple, AZ, USA) were placed in the right iliac artery to block the tear. Intraoperative angiography reveals patent visceral arteries and no false lumen opacification (Figure 4D). The 6-month postoperative CTA examination showed stable positions of the AVP and stent graft, with complete thrombosis in the false lumen. The abdominal aorta diameter decreased from 52 mm to 47 mm, while the true lumen increased from 24 mm to 28 mm (Supplementary Figure S2D).

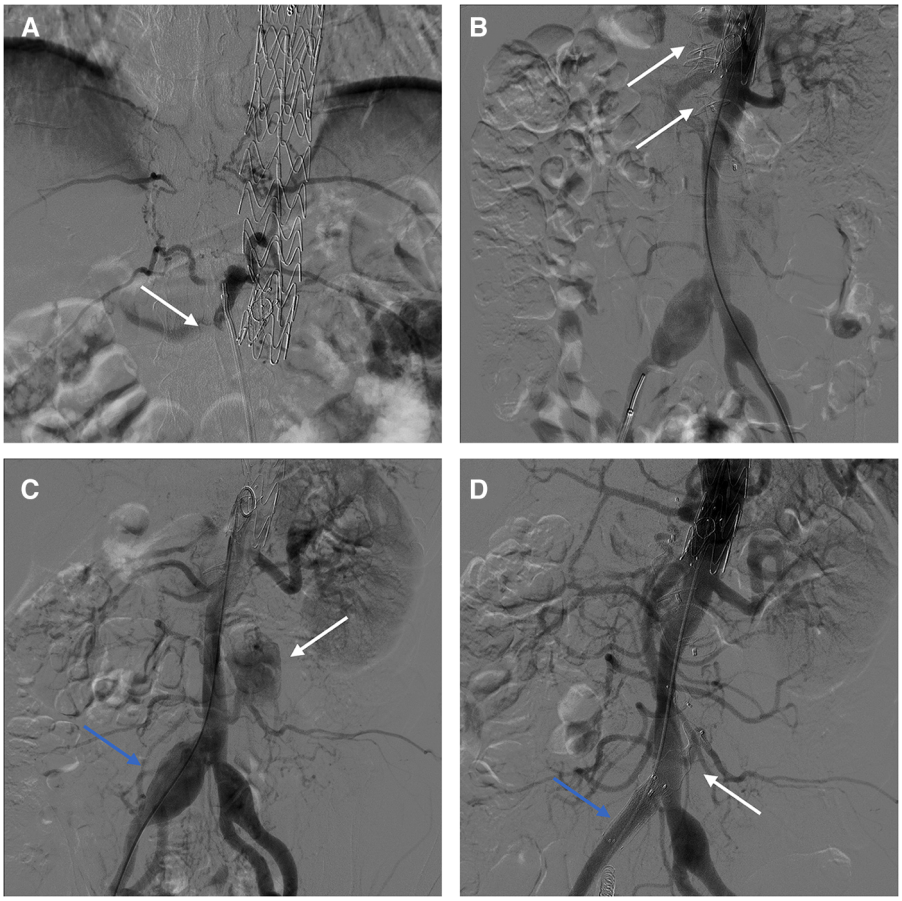

FIGURE 4
(A) Intraoperative angiography showed abdominal aortic false lumen opacification. (B) AVPs were inserted into the false lumen. (C) Opacification of false lumen in infrarenal aorta (white arrow); Aneurysmal dilatation of right iliac artery (blue arrow). (D) Final intraoperative angiography showed the AVP (white arrow) and the stent (blue arrow).

Discussion

After TEVAR, the persistence of distal tears impedes the complete closure of the false lumen, leading to inadequate thrombus formation and facilitating the advancement of distal dissection. This becomes a primary reason for secondary surgical treatment and sparks debates on whether to simultaneously seal the distal tear during TEVAR (8, 9). Currently, various techniques exist in the field, such as F/B-EVAR, chimney technique, embedded branch stent, provisional extension to induce complete attachment (PETTICOAT) technique, stent-assisted balloon-induced intimal disruption and relamination in aortic dissection repair (STABILISE) technique, Knickerbocker, etc (10, 11).

Although significant progress has been made in endovascular treatment of aortic dissection, there remain numerous unresolved issues including etiology, pathophysiology, anatomical changes, prognosis, as well as treatment strategies, techniques, and various materials and tools. Currently, clinical research is primarily focused on the improvement of treatment modalities and materials. Due to factors such as aortic compliance and compatibility, the use of covered stents in treatment leads to a relatively high rate of long-term reintervention. For anatomically complex aortic dissections, there has been an emergence of numerous surgical approaches in recent years. The selection of appropriate individualized treatment should be based on a deep understanding of the disease, extensive clinical experience, and advanced technical skills. Currently, commonly chosen techniques for selective patients include chimney technique and F/B-EVAR technique (12, 13). While these methods can achieve the desired therapeutic effects, they also come with a high rate of re-intervention (14, 15) and significant economic costs. Therefore, it is crucial to simplify treatment strategies and procedures based on patient benefit.

At present, there have been only a few case reports that have described the application of AVP for selectively sealing tears and filling false lumens. Compared to other surgical methods, the use of AVP closure for tears has advantages such as less trauma and lower economic costs. Moreover, the most important aspect is that this technique is relatively simple in terms of operation. In 2012, Yeom et al. (16) published the first documented case of employing AVP Ⅱ for the occlusion of a distal tear in chronic type B aortic dissection. Subsequent studies have demonstrated a high success rate of AVP occlusion in sealing the tear, as well as favorable long-term aortic remodeling (17–20). AVP is a nickel-

titanium mesh occlusion device with multiple layers. It is designed to be precise in positioning, controllable in release, and retrievable. AVP selectively blocks the tears, thereby preventing extensive ineffective coverage of the graft within the true lumen. This significantly lowers the risk of spinal cord ischemia and eliminates the possibility of complications related to the stent.

Using AVP to occlude the tears in combination with false lumen embolization effectively solves the problem of incomplete tears occlusion and small hidden endoleaks. These two methods can also be applied alone. It's worth exploring the use of false lumen embolization to treat post-dissection aortic aneurysm that involve the visceral arteries. This technique can effectively rebuild a healthy anchor zone, which means we can avoid complicated surgeries and even obviating the need for further EVAR.

Based on the concept of simplifying surgery, we employed embolization techniques for selective treatment in three patients with aortic dissection, including one case of Stanford type A, achieving better-than-expected outcomes. The technique procedure is relatively simple, and there's no need for excessive placement of stents within the true lumen.

The AVP occlusion of the intimal tears is applicable for the treatment of select patients with type A and type B aortic dissection. However, strict patient selection is required due to limitations in materials and intimal stability. Firstly, the diameter of the AVP should correspond to the size of the intimal tear, so it's important to choose a tear that is both more regular in shape and smaller than the diameter of the AVP. Furthermore, in the acute phase, the endarterium is fragile and morphologically unstable, with the size of the tears prone to change. There is a potential risk of device displacement and endarterium tearing when AVP is used, making thorough preoperative assessment extremely important. The chronic phase of aortic dissection exhibits a stable intimal flap, which makes it suitable for embolization technique treatment. However, there are also issues such as a relatively large diameter of the entry tears. Therefore, precise measurements should be taken before surgery to select an appropriate AVP. Thirdly, in the area of visceral arteries with true and false lumens blood supply, embolization of the false lumen should be combined with the treatment of graft stents for the visceral arteries to avoid organ ischemia. In the occlusion of AVP with intimal tears located at the opening of cervical artery and visceral artery, it is crucial to exercise caution in order to prevent stenosis or unintentional closure of the arterial opening. Additionally, the use of embolization techniques alone is not suitable for ruptured or impending rupture of aortic dissection.

Conclusion

Simplification of endovascular treatment is one of the important goals in the design of procedures for complicated aortic dissections. The selective application of embolization techniques can yield significant benefits in cases of aortic dissection or post-dissection aortic aneurysms that involve visceral arteries. Although embolization technology is not a recent technique, with continuous improvements and innovations in materials, its application will become more and more widespread. In the future, the development of specialized embolic devices for complex morphologies of aortic dissection will have significant practical value.

Data availability statement

The raw data supporting the conclusions of this article will be made available by the authors, without undue reservation.

Ethics statement

The studies involving humans were approved by Scientific Research Ethics Committee of Qilu Hospital of Shandong University. The studies were conducted in accordance with the local legislation and institutional requirements. Written informed consent for participation was not required from the participants or the participants' legal guardians/next of kin because this study was a retrospective study, and informed consent of patients was not required. Written informed consent was obtained from the participants for the publication of our study.

Author contributions

RL: Writing – original draft, Writing – review & editing. YL: Conceptualization, Funding acquisition, Supervision, Writing – review & editing. JJ: Conceptualization, Data curation, Funding acquisition, Methodology, Project administration, Resources, Supervision, Validation, Writing – review & editing.

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Conflict of interest

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Supplementary material

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Case Report: Combined transcatheter arterial embolization and aortic stent-graft have better efficacy for bronchial artery aneurysms

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Bronchial artery aneurysm (BAA) is a rare and fatal condition that requires immediate treatment. However, conventional surgical and transcatheter arterial embolization treatments are less effective. In the present case, a 76-year-old hypertensive woman was admitted with dizziness and diagnosed with an unruptured bronchial artery aneurysm, which was treated by transcatheter arterial embolization and aortic stent-graft. The patient's clinical status was stable during the 4-year follow-up. Simultaneously, we reviewed 79 research papers, analyzing past BAA cases for their etiology, symptoms, and treatment outcomes. We found that catheter arterial embolization and aortic stent-graft implantation, especially for BAA of short-necked and arterial tortuosity, demonstrate superior efficacy compared to other methods. Therefore, we consider this approach to be the preferred choice in clinical BAA treatment.

KEYWORDS

bronchial artery aneurysm, endovascular treatment, transcatheter arterial embolization, aortic stent-graft, cardiovascular surgery

1 Introduction

Bronchial artery aneurysm (BAA) has a prevalence of less than 1% in patients examined by bronchial arteriography, but it can be fatal (1). Given the possible asymptomatic nature of BAA, timely intervention after diagnosis is imperative to mitigate the risk of rupture and subsequent massive hemoptysis (2, 3). As interventional techniques advance, transcatheter arterial embolization (TAE) is progressively replacing traditional surgery as the primary treatment method for BAA (4). However, TAE carries a risk of complications, including transverse myelitis, bronchial infarction, oesophageal-bronchial fistula, and even spinal cord ischemia (5). Furthermore, TAE poses the risk of inadequate bronchial artery embolization, increasing the probability of hemoptysis recurrence, particularly in cases involving persistent pulmonary tuberculosis (6). Consequently, in response to the limitations associated with TAE, the utilization of a combined strategy involving TAE and aortic stent-graft has emerged as a novel therapeutic option (7).

In this report, we successfully treated a 76-year-old female patient with a 25-mm diameter BAA using TAE and aortic stent-graft and the patient had a favorable

prognosis without complications during a 4-year follow-up period. TAE and aortic stent-graft not only proved to be effective in the treatment of BAA in past reports but also demonstrated positive therapeutic effects and a good short-term prognosis in this case (8, 9).

In addition, the article reviews previous reports of BAA and provides a discussion of the etiology, symptoms, and treatment of a total of 85 patients with BAA, including the present case.

2 Case report

2.1 Case description

A 76-year-old woman presented to the neurology department with persistent dizziness and concerns regarding a possible brain lesion. The patient had a history of hypertension for over 20 years and a family history of cardiovascular disease. However, the patient claimed to have achieved long-term control of hypertension with amlodipine besylate tablets and had no personal or family history of psychiatric disorders or neurological diseases. Furthermore, a comprehensive physical examination was conducted on the patient. The results showed that the general appearance, head, neck, chest, abdomen, extremities, neurological system, and skin were all normal. However, the vital signs, namely, blood pressure and heart rate, were higher than normal at 175/62 mmHg and 103 beats per min, respectively. Therefore, due to the inability to rule out cardiovascular disease, vascular surgery staff participated in the consultation for this patient.

2.2 Diagnostic assessment

The suspicion of an aneurysm was first raised by an abnormal enhancement of a mediastinal lesion on a contrast-enhanced computed tomography (CT) scan of the chest (Figure 1A). The subsequent three-dimensional reconstruction of the CT angiogram showed a BAA with a 25-mm diameter and a dilated and tortuous inflow vessel from the right bronchial artery trunk (Figure 1B), which together with an aortic arch angiogram confirmed this diagnosis (Figure 1C).

2.3 Treatment

Following the puncture of the right femoral artery using the modified Seldinger technique, a 5 Fr arterial sheath and a 0.035-inch supersmooth guidewire were carefully inserted. Subsequently, aortography was conducted using a 5F pigtail catheter (Terumo, Tokyo, Japan) to assess the position of the opening of the aneurysmal inflow vessel of the bronchial artery. Super-selective angiography of the bronchial artery, performed with a 5F Cobra catheter (Terumo, Tokyo, Japan), confirmed the absence of spinal artery branch involvement and revealed a bronchial artery aneurysm with a diameter of approximately 25 mm.

To achieve comprehensive embolization, a sandwich therapy plan was devised to target both the inflow and outflow vessels, as well as the aneurysm itself. However, due to the tortuosity of the inflow vessel, it was challenging to advance into the outflow vessel using a microcatheter (MC-PE28131, Terumo, Tokyo, Japan) and a 0.018-inch micro-guidewire (Figure 2). Consequently, a cautious and gradual injection of polyvinyl alcohol (PVA) foam embolic particles (500–700 µm, Cook Medical, Bloomington, USA) was performed at the aneurysm site until distal blood vessels were no longer visible in angiography. Subsequently, occlusion of the inflow vessel was achieved using multiple embolic coils (IMWCE-35-8-10/15, COOK Medical, Bloomington, USA). In a strategic move to prevent potential revascularization by collateral arteries from the thoracic aorta, an aortic stent-graft (Ankura^{TM} TAA Stent Graft System, TAA2626B080, Life Technology Sciences, Shenzhen, China) was deployed in the thoracic aorta segment at the BAA opening, with a 20% oversizing. Subsequent angiography demonstrated the disappearance of the BAA, confirming the successful occlusion of the aneurysm.

2.4 Follow-up

The patient was discharged from the hospital without any complications. Follow-up after 1 year, utilizing CT angiography and three-dimensional aorta reconstruction, revealed an absence of contrast filling in the BAA, indicating a positive prognosis during the 1-year follow-up (Figure 3). Despite the patient's reluctance to undergo further hospital-based examinations due to financial constraints, an ongoing assessment was sustained through telephone interviews conducted every year to evaluate the patient's clinical status until the fourth year. Throughout the series of telephone follow-ups, the patient consistently expressed satisfaction with the treatment outcomes and claimed no discomfort.

3 Timeline

The figure below illustrates the timeline for the diagnosis, treatment, and follow-up of this case (Figure 4).

4 Discussion

As a rare and primarily asymptomatic illness, BAA has an extremely high death rate if it ruptures and hence requires rapid treatment (1–3). In this case, a 76-year-old patient with hypertension presented with persistent headaches, prompting the performance of a contrast-enhanced CT examination that unveiled the presence of BAA with a diameter reaching 25 mm. Given the patient's advanced age, our therapeutic decision-making aimed to minimize potential complications. Consequently, a strategic choice was made to employ a less invasive intervention, opting for TAE over conventional surgical

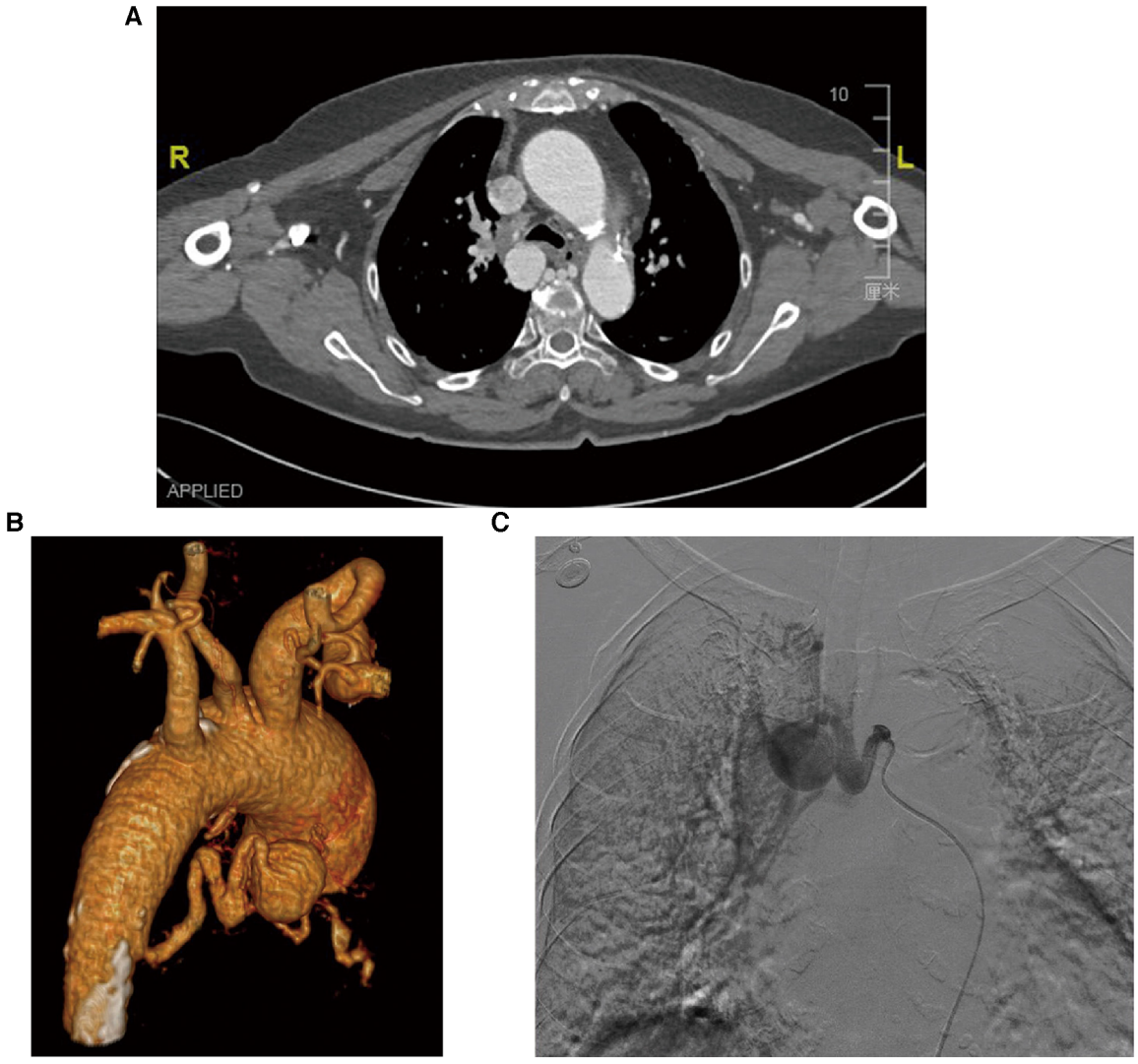

FIGURE 1
Diagnostic images of the patient with BAA. (**A**) The CT scan image of the chest shows an abnormally enhanced lesion in the mediastinum and dilated bronchial arteries near the descending aorta, but no entrapment or hemopneumothorax. (**B**) Three-dimensional reconstruction of the CT angiographic model shows a 25-mm-diameter BAA from the descending aorta with a dilated and tortuous inflow vessel. (**C**) Selective bronchial arteriography shows that the aneurysm was fed by a bronchial artery dilated by the descending aorta.

approaches (4). In the application of TAE, the sandwich therapy method emerges as the preferred approach due to its efficacy in averting bypass reflux by embolizing both inflow and outflow vessels (10). However, the catheter could not enter the outflow artery due to arterial tortuosity. Therefore, we employed a catheter to enter the BAA and induce embolization of the outflow artery by slowly injecting polyvinyl alcohol (PVA) particles to prevent blood reflux. Notably, coils are a safer option, but they present challenges when navigating through tortuous arteries. Concurrently, to mitigate the potential risks of coil dislocation and the washout of PVA particles by blood flow, we implemented an aortic stent-graft (8, 9). Notably, the application of an aortic stent-graft has demonstrated efficacy in isolating the thoracic aorta, preventing the formation of new collateral arteries and thereby impeding aneurysm refilling (9).

Furthermore, to obtain more clinical data and enhance our understanding of BAA, we searched for the keyword "bronchial artery aneurysm" using Pubmed and Google Scholar. Through the search, we selected a total of 85 cases from 79 articles for analysis (Supplementary Table 1). The sample consists of 41 women and 44 men, with a mean age of 57.35 years. The criteria for classifying the effectiveness and success of patient treatment were based on the Society of Interventional Radiology (SIR) guidelines (11).

The etiology of BAA is complex and yet not fully understood. However, current research suggests that its formation is caused by increased bronchial arterial blood flow and structural weakening of the vessel wall (8). According to reports, the etiology of BAA may be associated with hereditary factors, such as pulmonary sequestration and pulmonary artery agenesis. Additionally,

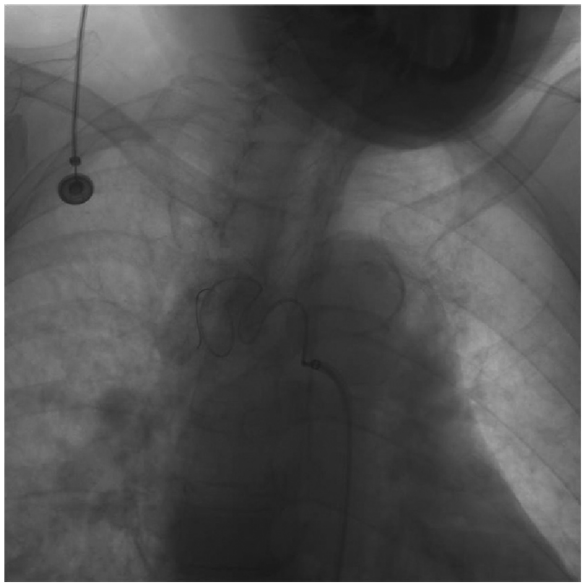

FIGURE 2
Angiogram during catheter intervention. Intraoperative angiography of the BAA shows that the catheter failed to enter the aneurysm due to tortuous arteries.

acquired factors such as atherosclerosis, bronchiectasis, tuberculosis, and sepsis have been suggested as etiology (12). In the review, the most common cause for patients was bronchiectasis (19/85), followed by tuberculosis (7/85) and hypertension (6/85), which corroborates the findings of previous studies. Interestingly, the number of patients with mycotic aneurysm (5/85) was also significant in our review. However, mycotic aneurysms have rarely been noticed and reported before. By definition, the mycotic aneurysm is a dilatation of the arterial wall caused by bacterial, fungal, and viral infections (13). Pathogens tend to invade damaged arteries, leading to infection of the intima. Then, it leads to the rapid degradation of the deeper arteries, ultimately causing the formation of an aneurysm (14). Therefore, despite the complex and challenging etiology of BAA, patients with a history of cardiovascular disease, bronchiectasis, and tuberculosis are at an increased risk of developing BAA and require additional clinical attention. It is noteworthy that over the past two decades, the incidence of mycotic aneurysms as the etiology of BAA has been increasing each year. Thus, we should remain vigilant about this emerging trend in BAA etiology (13).

BAA not only has a complex etiology but also has a diverse clinical presentation (15). In our review, we observed that hemoptysis (28.24%, 24/85) was the most common clinical symptom in patients with BAA, followed by chest pain (18.82%, 16/85) and dysphagia (8.24%, 7/85). However, the main symptoms of BAA depend on its location (8). Intrapulmonary BAA commonly presents with hemoptysis, but there is a small proportion of asymptomatic cases. On the other hand, intrapulmonary BAA has been reported to cause dysphagia as the main symptom, although it is asymptomatic in most cases

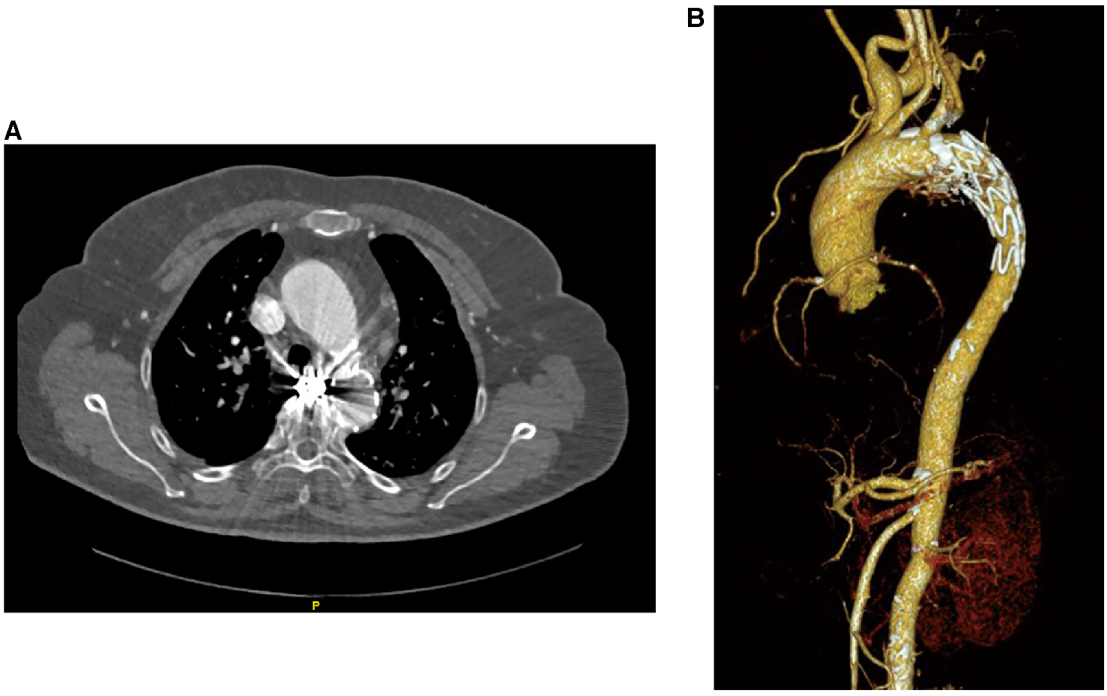

FIGURE 3
Patient's follow-up examination pictures after 1 year of treatment. (**A**) CT angiography was performed on the patient 1 year after treatment, and the scan suggests the presence of metallic artifacts from coils within the BAA. (**B**) Three-dimensional reconstruction of the vessel based on the CT angiography data shows no rupture of the BAA and no new collateral arteries supplying the BAA.

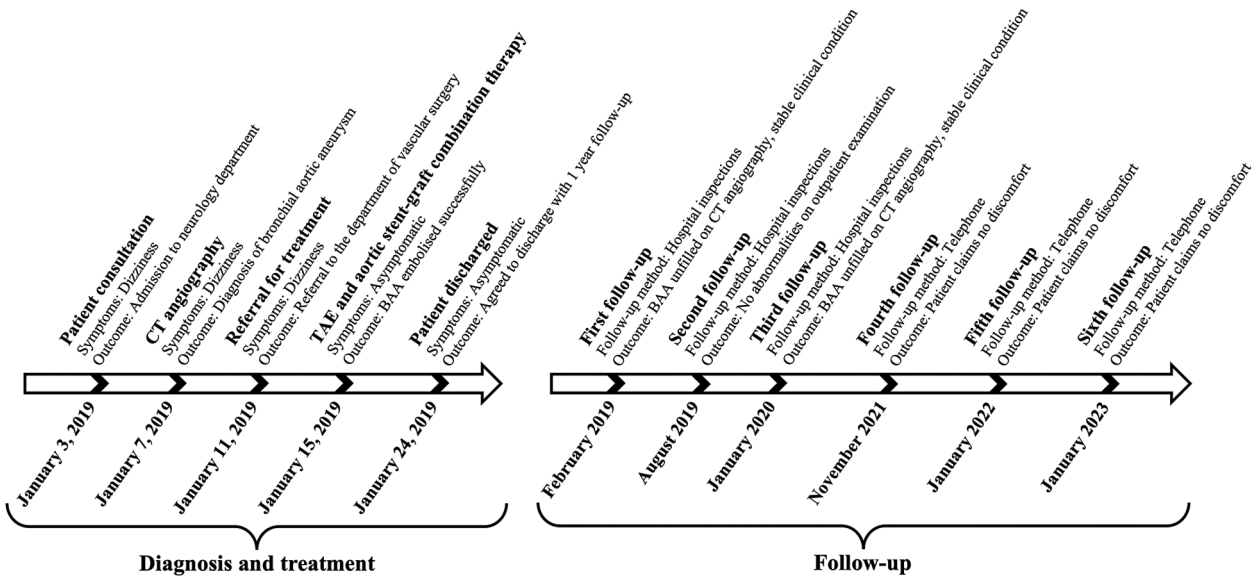

FIGURE 4
Timeline of treatment and follow-up.

(16). It is worth noting that there are asymptomatic patients in all types of BAA, accounting for a certain percentage of patients in our review (14.12%, 12/85). These patients are usually detected incidentally on chest CT scans, so it is easy to delay treatment (17). Additionally, the presence or absence of BAA rupture can also affect the symptoms. Among patients with ruptured BAA, a significant proportion (40.00%, 10/25) experienced chest pain symptoms. In contrast, patients without ruptured BAA had a lower proportion of chest pain symptoms (6.98%, 6/86), with the most frequent symptom in these cases being haemoptysis, occurring in 17.44% (15/86) of patients. Notably, in the case review, the average diameter of BAA was 25.65 mm. Unruptured BAAs had an average diameter of 29.09 mm, while ruptured BAAs had a significantly smaller average diameter of 20.81 mm. The risk of BAA rupture appears to be inversely proportional to the diameter of the BAA. Therefore, if patients with BAA exhibit a smaller aneurysm diameter along with symptoms of chest pain, the likelihood of BAA rupture is significant. This necessitates urgent intervention for these patients.

So far, there are no standardized diagnostic and therapeutic guidelines for BAA, resulting in the use of a variety of diagnostic and therapeutic approaches (12). CT angiography stands out as the most frequently employed imaging modality for BAA diagnosis, with previous studies showing promise in MRI techniques (1, 18). TAE has emerged as the preferred treatment for BAA due to its safety, minimally invasive nature, and effectiveness (4). However, recent studies have demonstrated that combining aortic stent-graft with TAE yields better outcomes compared to TAE alone, particularly in patients with tortuous arteries and short necks (7). This combined approach not only mitigates the movement of TAE embolic coils but also effectively isolates the artery supplying the aneurysm, presenting a valuable advancement in BAA treatment strategies (19). In our case, the combination therapy yielded favorable outcomes and a positive short-term prognosis for BAA with a severely tortuous inflow artery, consistent with previous research. Furthermore, we conducted a comprehensive review of the success rates of different treatment methods. Among the 19 patients subjected to surgical intervention, 2 fatalities occurred, resulting in a mortality rate of 10.53%. In the cohort of 49 patients undergoing exclusive TAE, 9 experienced treatment failure, yielding a success rate of 81.63%. Notably, within the 13-patient group receiving combined therapy, the treatment success rate reached an impressive 100%. Therefore, combined therapy has a huge potential for the treatment of BAA. In contrast, certain studies suggest that the use of aortic stent-grafts may increase the risks of spinal cord ischemia and pseudoaneurysm rupture, warranting a cautious approach to their utilization (10, 20). However, due to the short follow-up period in this case, further monitoring is necessary to determine whether its long-term prognosis is favourable.

5 Conclusion

BAA has a low incidence but can be fatal. Based on the literature review and this case, we consider TAE and arterial stent implantation to be effective for BAA, particularly in cases with short necks and tortuous arteries. Given the complexity of BAA etiology and symptom diversity, our brief analysis of past literature provides insights into the causes and symptoms of BAA. This contributes to a better understanding of BAA and highlights the efficacy of TAE and aortic stent-graft.

Data availability statement

The original contributions presented in the study are included in the article/Supplementary Material, further inquiries can be directed to the corresponding authors.

Ethics statement

The studies involving humans were approved by Ethics Committee of the Second Affiliated Hospital of Nanchang University. The studies were conducted in accordance with the local legislation and institutional requirements. The participants provided their written informed consent to participate in this study. The manuscript presents research on animals that do not require ethical approval for their study. Written informed consent was obtained from the individual(s) for the publication of any potentially identifiable images or data included in this article.

Author contributions

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Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

Publisher's note

Supplementary material

The Supplementary Material for this article can be found online at: https://www.frontiersin.org/articles/10.3389/fcvm.2023.1328674/full#supplementary-material

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Case Report: Coil embolization of ascending aortic pseudoaneurysm in patient with Behcet's disease

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Background: Behcet's disease (BD) is a systematic vasculitis that affects vessels with various sizes, presenting as venous thrombosis and arterial pseudoaneurysms. The most severe manifestation in BD is ascending aortic pseudoaneurysm, which is associated with high risks of rupture and mortality.
Case presentation: We present a case of ascending aortic pseudoaneurysm in a 50-year-old patient with BD. After preoperative evaluation, coil embolization was successfully performed to treat the pseudoaneurysm, resulting in a satisfactory outcome at the 1-year follow-up.
Conclusion: Coil embolization serves as an effective treatment option for ascending aortic pseudoaneurysm in BD when open surgical repair and stent graft placement are unsuitable.

KEYWORDS

Behcet's disease, ascending aortic pseudoaneurysm, coil embolization, thoracic endovascular repair, computed tomography angiography

1 Introduction

Pseudoaneurysm of the ascending aorta represents a rare vascular complication in patients with Behcet's disease (BD) and carries a potentially fatal risk (1). Given the heightened threat of rupture and mortality, these individuals often necessitate either open surgical or endovascular repair. While traditional open surgical repair remains a viable option for select patients, the presence of anastomotic complications contributes to an unfavorable outcome, including issues such as leakage, occlusion, and pseudoaneurysm formation (2). Recently, mounting evidence has underscored the efficacy of endovascular repair (3, 4).

In this report, we present a case of ascending aortic pseudoaneurysm in a patient with BD, successfully managed through coil embolization, leading to a favorable outcome at the one-year follow-up. Consent was obtained from the patient for the publication of this data.

2 Case presentation

The patient, a 50-year-old man with a confirmed 16-year diagnosis of BD, was referred to our hospital because a recent computed tomography angiogram (CTA) conducted at another facility had shown a pseudoaneurysm of the ascending aorta. He had reported left-sided chest pain since 2 weeks prior. The patient was known with visual impairment, which was attributed to recurrent episodes of ocular inflammation, and over the last several years, he had experienced approximately 10–20 oral ulcers per year.

Prior treatment with colchicine proved ineffective. Consequently, rheumatologists had adjusted the treatment regimen to incorporate glucocorticoids, cyclosporine, and adalimumab. However, during the tapering of adalimumab, the patient had experienced recurrent erythema nodosum in the extremities. The patient had discontinued adalimumab for 10 days upon admission to our center.

Additionally, the patient had comorbidities including hypertension and type II diabetes mellitus, for which long-term telmisartan 40 mg once daily and metformin 500 mg twice daily were prescribed. Upon admission, the patient reported mild left-sided chest pain, without accompanying compressive symptoms such as shortness of breath, dysphagia, stridor, or hoarseness. This subtle clinical presentation may be attributed to the pseudoaneurysm of the ascending aorta associated with BD, which can exhibit slow growth or even remain stable for a prolonged period, resulting in a lack of overt symptoms during chronic progression. The patient presented with a blood pressure of 117/53 mmHg and a heart rate of 65 beats/minute. A comprehensive physical examination revealed normal heart rhythm and apical position, absence of abnormal pulsations in the precordial region, and no pathological murmurs in the valve area. Electrocardiogram findings and biomarkers for cardiac injury were within normal ranges, excluding acute coronary syndrome.

The patient had a hemoglobin level of 124 g/L (reference range 130–175 g/L), a high-sensitive C-reactive protein level of 8.08 mg/L (reference range 0–3 mg/L), and an erythrocyte sedimentation rate of 28 mm/h (reference range 0–15 mm/h). CTA revealed an ascending aortic pseudoaneurysm with a patchy hypodense opacity along the irregular wall of the aortic arch, indicative of an inflammatory condition. Preoperative CTA was used to evaluate morphology and size of the pseudoaneurysm. The aortic pseudoaneurysm was situated in the anterior wall of the ascending aorta, approximately 39 mm from the aortic sinus. The diameter of the neck of the pseudoaneurysm measured 6 mm, whereas its maximum sac diameter reached up to 18 mm, with an adhered thrombus with a maximum thickness of 23 mm observed on its external surface (Figure 1).

After multidisciplinary consultation, decision was made to perform coil embolization. Under local anesthesia, a pigtail catheter was introduced into the proximal end of the ascending aorta via the right femoral artery. Angiography revealed the presence of the pseudoaneurysm, located at the anterior wall of the aorta, with a maximum sac diameter of 20 mm, and a neck diameter of 6.7 mm. Subsequently, the pigtail catheter was replaced with a 5F multifunctional catheter (Terumo, Tokyo, Japan) enclosed in a 6F long sheath (COOK, Bloomington, IN, USA). Five Interlock coils of varying sizes were introduced into the pseudoaneurysm cavity, including one measuring 20–400 mm, two measuring 15–400 mm, and two measuring 10–250 mm. Intraoperative angiography confirmed complete occlusion of the pseudoaneurysm sac with coils, effectively excluding it from arterial blood flow (Figure 2). The procedure duration was 40 min. The patient remained admitted for 6 days and was subsequently discharged. Successful treatment was confirmed by a follow-up CTA scan 1-month postoperatively, which revealed no evidence of endoleak. Subsequent assessment by CTA 1-year postoperatively demonstrated complete thrombosis and reduction in the size of the pseudoaneurysm sac (Figure 3). Meanwhile, the erythrocyte sedimentation rate and high-sensitivity C-reactive protein levels remained within normal

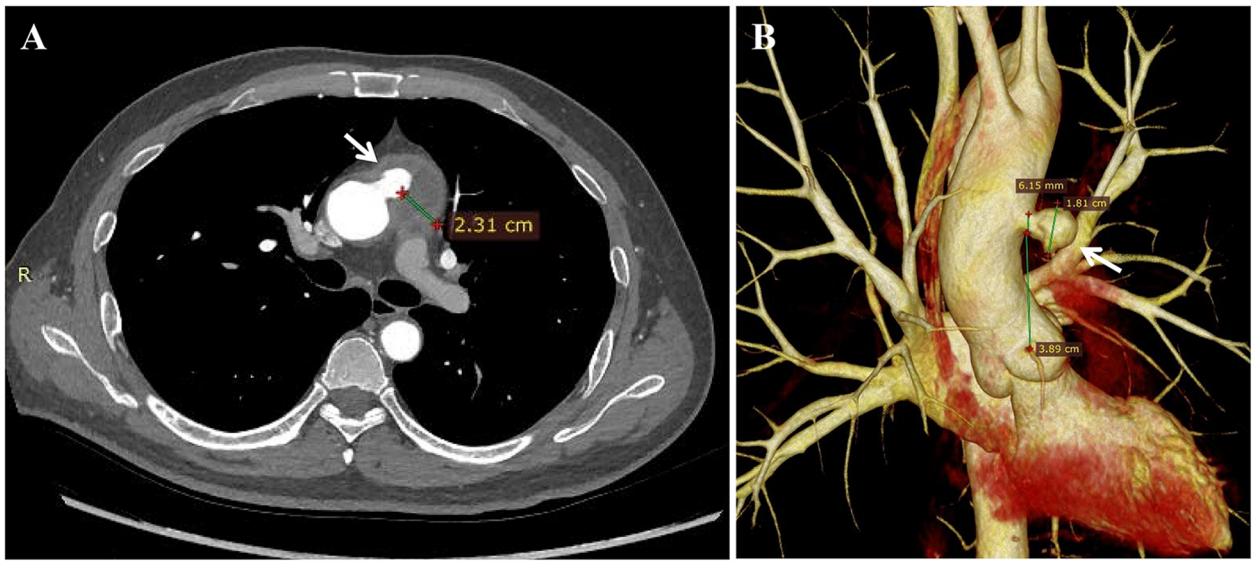

FIGURE 1
Preoperative CT angiography revealed (A) the pseudoaneurysm located at the anterior wall of the ascending aorta (indicated by the white arrow), with an adhered thrombus of maximum thickness 23 mm observed on its external surface. (B) Morphological measurements of the lesion were obtained from 3D reconstructed images, showing a maximum pseudoaneurysm neck diameter of 6 mm, maximum pseudoaneurysm sac diameter of 18 mm, and a distance of 39 mm from the aortic sinus to the pseudoaneurysm.

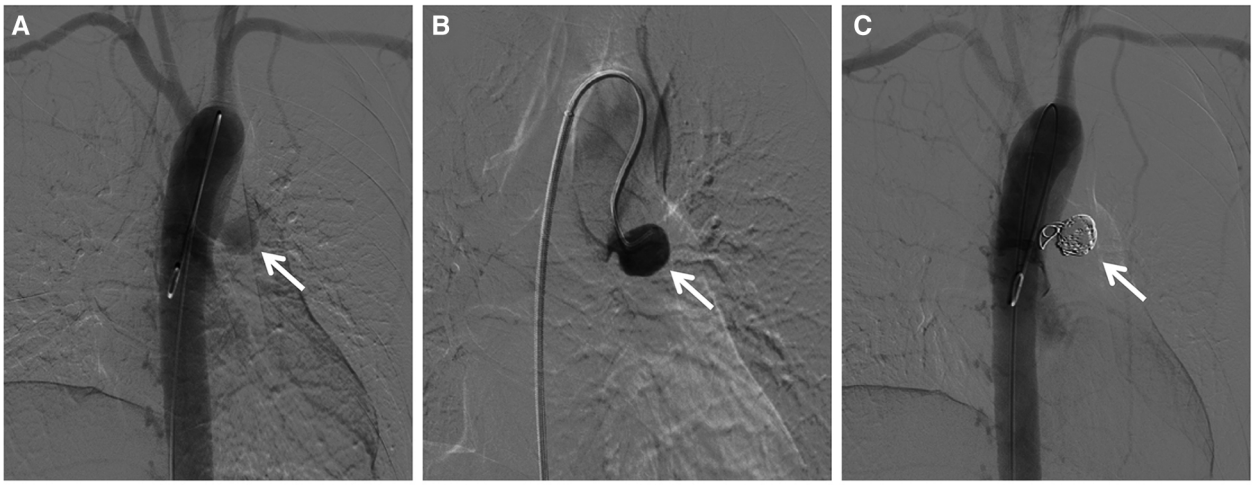

FIGURE 2
Intraoperative angiogram displayed (**A**) the anterior wall location of the aortic pseudoaneurysm and (**B**) clear visualization upon contrast administration. Following coil embolization, the angiogram demonstrated (**C**) complete occlusion of the pseudoaneurysm sac with coils, leading to exclusion from arterial blood flow (indicated by the white arrow).

ranges with drug treatment. Currently, the patient is being followed up at regular intervals. Cardiac surgery will only be considered in case of occurrence serious, late postoperative complications (The timeline with relevant data from the episode of care is presented in Figure 4).

3 Discussion

In addition to oral and genital ulcerations, and relapsing ocular inflammation, BD also affects vessels, including venous thrombosis and arterial pseudoaneurysm (5). Ascending aortic involvement represents one of the most serious complications of BD, with aortic rupture emerging as the leading cause of mortality in patients with vascular involvement (1, 6). The pathogenesis of pseudoaneurysm formation in BD is associated with the occlusion of nutrient vessels within the arterial wall due to pan vasculitis. Chronic inflammation leads to proliferation of fibroblast and smooth muscle cells within the intimal layer, along with deposition in the arterial wall, resulting in destruction of the medial and outer layers. Pseudoaneurysms in patients with BD are highly prone to rupture and necessitate emergency open surgical or endovascular repair.

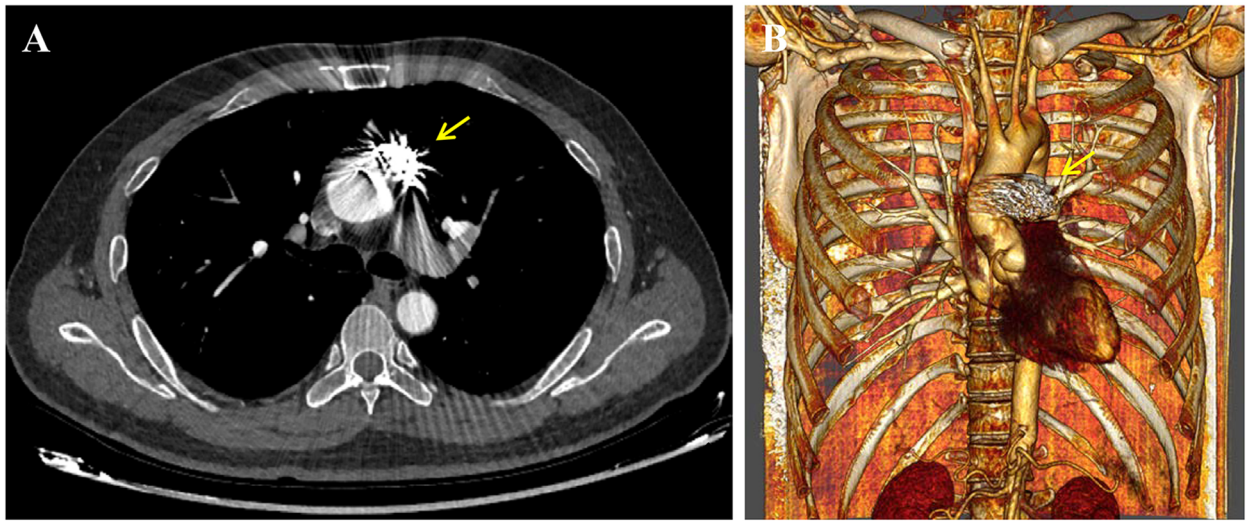

FIGURE 3
Postoperative CT angiography at the 1-year follow-up depicted (**A**) complete thrombosis and reduction in the size of the pseudoaneurysm sac. (**B**) 3D reconstructed image showed secure positioning of the coils without any evidence of migration (indicated by the yellow arrow).

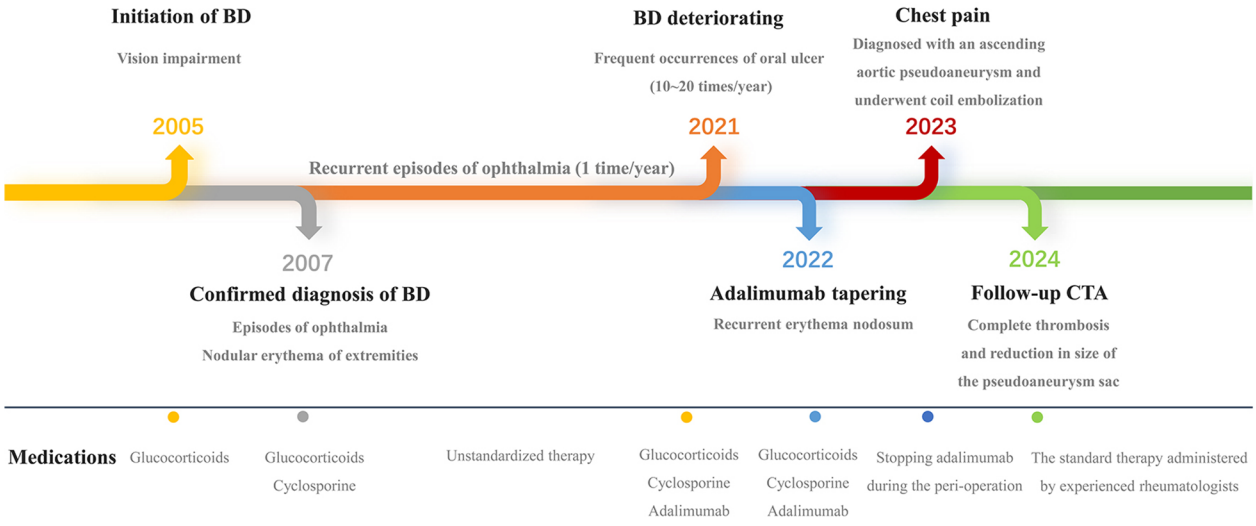

FIGURE 4
Timeline illustrating relevant data from the episode of care.

Open surgical repair has traditionally been considered the gold standard for treating ascending aortic pseudoaneurysms in patients with BD. However, this procedure is associated with high mortality rates and recurrent anastomotic pseudoaneurysms, leading to unfavorable outcomes. A meta-analysis demonstrated that the mortality rate following open surgical repair ranged from 10% to 30%, and 10% to 50% of patients with BD experienced recurrent pseudoaneurysms at the site of anastomosis (7). Moreover, because of the extensive trauma induced by sternotomy, and the deep hypothermic circulatory arrest, patients with major-comorbidities are often poor candidates for surgery.

Recently, endovascular repair has emerged as a novel option for treating pseudoaneurysms of the ascending aorta (3). The three most prevalent procedures include stent graft placement (8), vascular occlusion, and coil embolization (9), all of which have demonstrated reduced invasiveness and considerable efficiency. However, several unresolved questions hinder the widespread adoption of these interventions. Regarding stent graft placement, several challenges exist. Firstly, there is currently no stent graft specifically designed for ascending aortic lesions, limiting the options to off-label use of stent grafts designed for thoracic or abdominal aortic lesions. Secondly, the commercially available delivery systems used in our medical center are primarily designed for descending aortic lesions and may not provide sufficient length to effectively guide the stent graft from the femoral artery to the ascending aorta. Although using either the ventricular apex or axillary artery can reduce the delivery distance, it is unsuitable for patients with BD due to its high invasiveness (10). Lastly, mechanical stimulation and foreign body reactions caused by the stent graft may induce inflammation in the aortic wall (11), potentially leading to the recurrence of pseudoaneurysms at the edge of the stent graft. Several studies have also reported the successful use of endovascular occluders. Tarantini et al. (12) employed two Amplatzer septal occluder devices for percutaneous repair of ascending aortic pseudoaneurysm and aortopulmonary fistula. Komanapalli et al. (13) performed percutaneous repair of an ascending aortic pseudoaneurysm with a septal occluder device. When anatomically suitable, a less invasive approach using a vascular plug or septal occluder is an attractive option (14). However, the precise implantation of occluder devices at the neck of the pseudoaneurysm may pose technical challenges due to the variable location and size of these lesions. Currently, there is a lack of commercially available endovascular occluder devices in many medical centers. While coil embolization has become the preferred treatment for aneurysms in specific locations (15), its efficacy in the ascending aorta, particularly in patients with BD, remains uncertain. D'Ortenzio et al. (16) employed coil embolization to successfully address an aortic root pseudoaneurysm in a patient diagnosed with Loeys-Dietz syndrome. Dziekiewicz et al. (17) utilized coil embolization to treat a patient with post-traumatic pseudoaneurysm of the ascending aorta, demonstrating its efficacy as a safe and viable alternative for managing cases at high-risk for hemorrhage, where heparinization is unfeasible, both as a bridge procedure and as a long-term solution. Kim et al. (9) used coil embolization to address the pseudoaneurysm at the aortic sinotubular junction following multiple aortic operations and achieved a good outcome. Moreover, coil embolization can be used in combination with other endovascular techniques for the management of these patients. Wu et al. (3) combined bare stents with coil embolization to treat patients with vascular BD, resulting in an unsatisfactory outcome. Lyen et al. (18) utilized endovascular treatment to manage thoracic aortic pseudoaneurysms and proposed a novel treatment strategy that combined the use of occluder devices and coils as an effective treatment strategy for these patients. However, the efficacy of using coil embolization technology exclusively as an endovascular

treatment option for ascending aortic pseudoaneurysm in patients with BD remains unclear.

To our knowledge, our procedure represents the pioneering use of coil embolization for treating ascending aortic pseudoaneurysms in patients with BD. The decision to employ coil embolization for managing this patient was based on several factors: firstly, the anatomical characteristics of an anterior wall location of the ascending aortic pseudoaneurysm with a narrow neck provided favorable endovascular conditions for coil embolization; secondly, considering the presence of BD in this patient, which renders the aortic wall more vulnerable to damage, we aimed to minimize iatrogenic injury caused by devices such as catheters and wires during the endovascular procedure. However, the utilization of coil embolization in ascending aortic pseudoaneurysms entails potential risks, including intraoperative aortic rupture and incomplete pseudoaneurysm filling, as well as postoperative endoleak and coil migration. To mitigate these adverse events, several key considerations were taken into account during the procedure: (i) gradual release of coils is essential to minimize the risk of pseudoaneurysm rupture; (ii) the combination of different sizes of coils can be employed to achieve complete filling and appropriate oversizing; (iii) precise positioning of coils is crucial to prevent distal migration, therefore we employed controlled-release coils that could be retracted multiple times, adjusted for correct orientation, and subsequently redeployed within the pseudoaneurysm cavity. Pseudoaneurysm at the puncture site is a commonly encountered complication following endovascular repair in patients with BD and vascular involvement. In this case, meticulous attention to the management of the puncture site led to the absence of post-operative pseudoaneurysm at the right femoral artery.

In strict accordance with the European League Against Rheumatism (EULAR) recommendations for the management of BD (19), the patient received regular immunosuppressant and glucocorticoid treatment pre-operatively, with coil embolization scheduled 2 weeks after adalimumab administration in this case, in order to mitigate the risk of postoperative complications such as enlargement or recurrence of pseudoaneurysm. Additionally, experienced rheumatologists adjusted perioperative and postoperative drug dosages to effectively control disease activity. The ESR and hsCRP levels remained within normal ranges during the 1-year postoperative period, indicating that this satisfactory outcome of coil embolization may also be attributed to the sustained administration of immunosuppressants. Therefore, we suggest that standardized continuation of immunosuppressive therapy is strongly recommended following endovascular treatment in patients with vascular BD. Many questions remain unaddressed regarding the long-term durability and safety of transcatheter coil embolization. Nevertheless, this technique seems to serve as a viable alternative for ascending aortic pseudoaneurysm in patients with BD, when open surgical repair or stent graft placement entails a significant risk of postoperative complications.

4 Conclusion

Ascending aortic pseudoaneurysm represents a rare and potentially fatal complication of BD. Coil embolization may present a viable alternative to sternotomy in patients with suitable anatomical characteristics.

Data availability statement

The original contributions presented in the study are included in the article/Supplementary Material, further inquiries can be directed to the corresponding author.

Ethics statement

Written informed consent was obtained from the individual(s) for the publication of any potentially identifiable images or data included in this article.

Author contributions

MK: Writing – original draft, Conceptualization, Writing – review & editing. BZ: Writing – review & editing, Resources. HP: Writing – review & editing, Resources. HH: Writing – review & editing, Resources. JZ: Writing – review & editing, Conceptualization, Resources, Supervision.

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Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

Publisher's note

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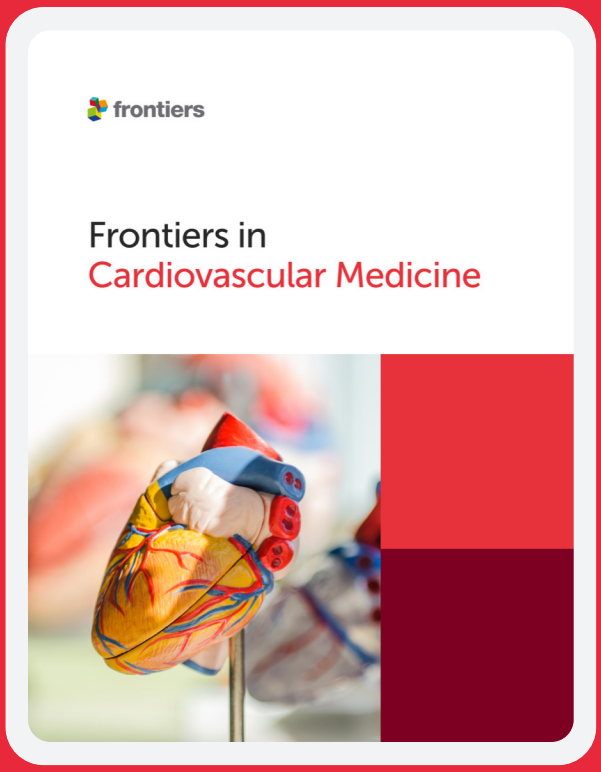

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