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Atrophic dermatofibrosarcoma protuberans: Two case reports and literature review

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Dermatofibrosarcoma protuberans is a rare, locally aggressive, slowly growing cutaneous fibroblastic sarcoma with a high recurrence rate and low metastatic potential. Atrophic dermatofibrosarcoma protuberans is a rare variant usually presents as atrophic plaques, easily neglected and misdiagnosed as benign lesions by patients and dermatologists. Here we report two cases of atrophic dermatofibrosarcoma protuberans, one of which was accompanied by pigment, and review other cases have been reported in the literature. Understanding the most up-to-date literature and early identification of these dermatofibrosarcoma protuberans variants can help clinicians avoid delayed diagnosis and improve prognosis.

KEYWORDS

dermatofibrosarcoma protuberans, atrophic, molecular diagnosis, case report, literature review

1 Introduction

Dermatofibrosarcoma protuberans (DFSP) is a slowly growing, aggressive dermal soft tissue tumor with a high recurrence rate and low metastatic potential (1). Classic DFSP often presents as a typical protuberant nodule and is histologically characterized by monomorphic spindle cells arranged in a storiform pattern (2). Although DFSP is a rare sarcoma with an incidence of 0.8 to 4.2 cases per million persons per year, it is nevertheless one of the most common cutaneous sarcomas (3). Atrophic dermatofibrosarcoma protuberans is a histopathological variant of DFSP, which presents as an asymptomatic depressed plaque. Atypical clinical presentation and indolent behavior are easily neglected by patients and clinicians, leading to delayed diagnosis. In this report, we describe two cases of atrophic DFSP, one of which also with hyperpigmentation, and review the literature to improve our understanding of the clinical and histopathological features of this unusual variant.

2 Case reports

2.1 Case 1

A 31-year-old woman attended our dermatology department to evaluate an erythematous plaque on her right chest that had been slowly progressing for four years. She was diagnosed as atrophic scar previously. Physical examination showed a 16×10 mm asymptomatic, smooth, atrophic plaque (Figure 1). Histopathologic examination revealed a monomorphic fibrohistiocytic spindle cell tumor arranged in a storiform pattern infiltrating the deep dermis and subcutaneous tissue forming honeycomb-like pattern (Figures 2A, B). Immunohistochemical analysis showed that cells were prominently positive for CD34 (Figure 2C) and negative for protein S-100. Next-generation sequencing (NGS) detected a fusion between exon 5 of COL1A1 and exon 2 of PDGFB. Based on these findings, the lesion was diagnosed as atrophic DFSP. The patient was treated by Mohs micrographic surgery and no recurrence was observed at the 1-year follow-up.

2.2 Case 2

A 45-year-old man presented to our dermatology department with a 5-year history of a depressed plaque on his left shoulder. The lesion was asymptomatic and slowly enlarged. He was previously diagnosed as blue nevus and anetoderma in other clinics. Physical examination showed a 30×20 mm round erythematous-to-bluish plaque with atrophy (Figure 3). Histopathologic analysis revealed infiltration of spindle cells with slender wavy nuclei into the deep dermis, arranged in parallel or horizontally oriented fascicles. Cells containing brown pigment were scattered in the dermis (Figures 4A, B). Immunohistochemical staining showed that the spindle cells were diffusely positive for CD34 but negative for S-100 while melanin-laden dendritic cells were positive for S-100 (Figures 4C, D). NGS detected a fusion between exon 46 of COL1A1 and exon 2 of PDGFB. Based on the clinicopathologic correlation, a diagnosis of atrophic pigmented DFSP was made. Mohs micrographic surgery was performed to excise the lesion.

3 Discussion

Atrophic dermatofibrosarcoma protuberans is a rare variant accounting for 1.7% of all DFSP cases (4). It was first described by Lambert in 1985 (5). Unlike the classical DFSP, atrophic DFSP usually presents as a slow-growing, atrophic or sclerotic plaque, which is more easily misdiagnosed as morphea, sclerodermiform basal cell carcinoma, atrophic scar, scleroderma, anetoderma, atrophic dermatofibroma, resolving panniculitis and lipoatrophy (6). Histologically, in addition to the storiform pattern noted in classical DFSP, neoplastic spindle cells arranged in parallel or horizontally oriented fascicles are also seen in atrophic DFSP (4). On immunohistochemistry, all tumors showed diffuse and strong positivity of CD34. Due to the overlap of morphology and

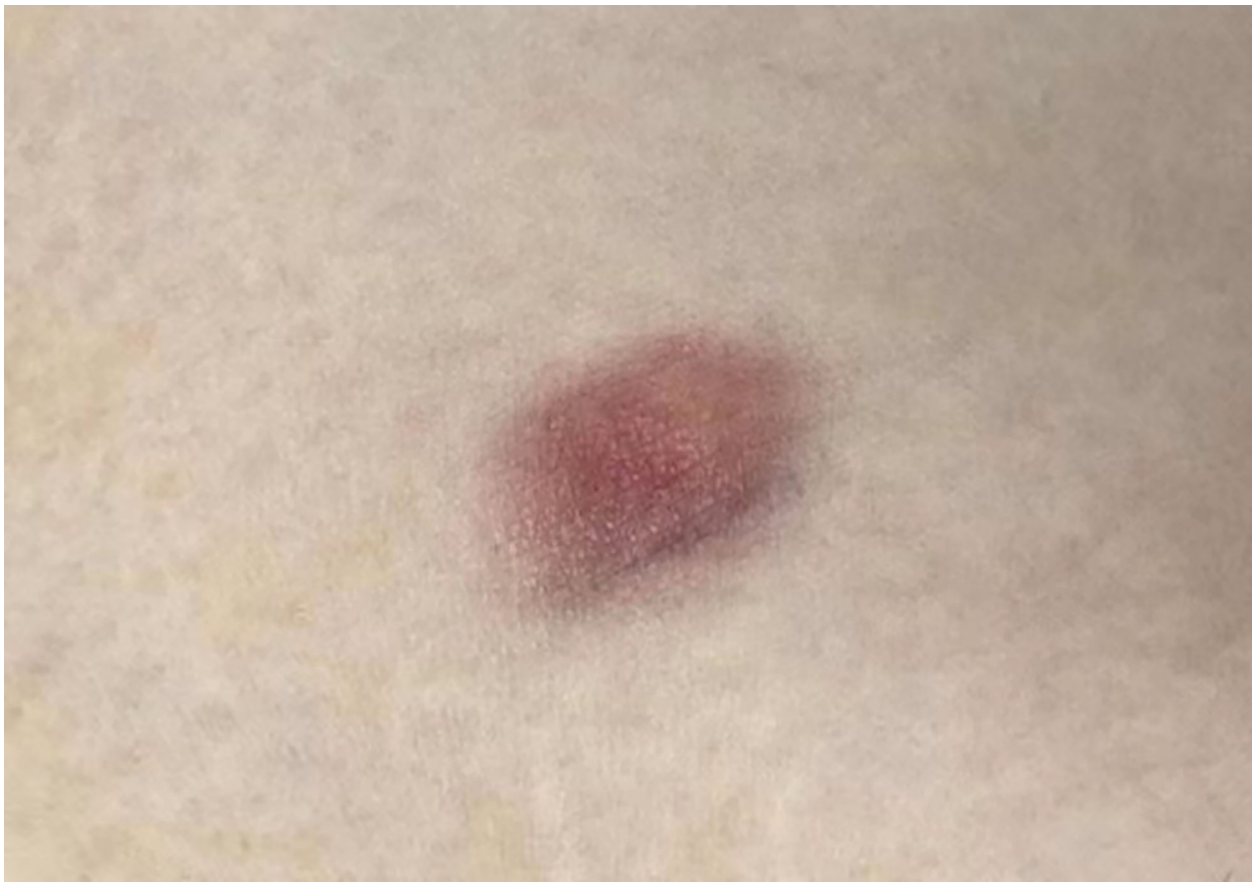

FIGURE 1
A 16×10mm atrophic erythematous plaque on her right chest.

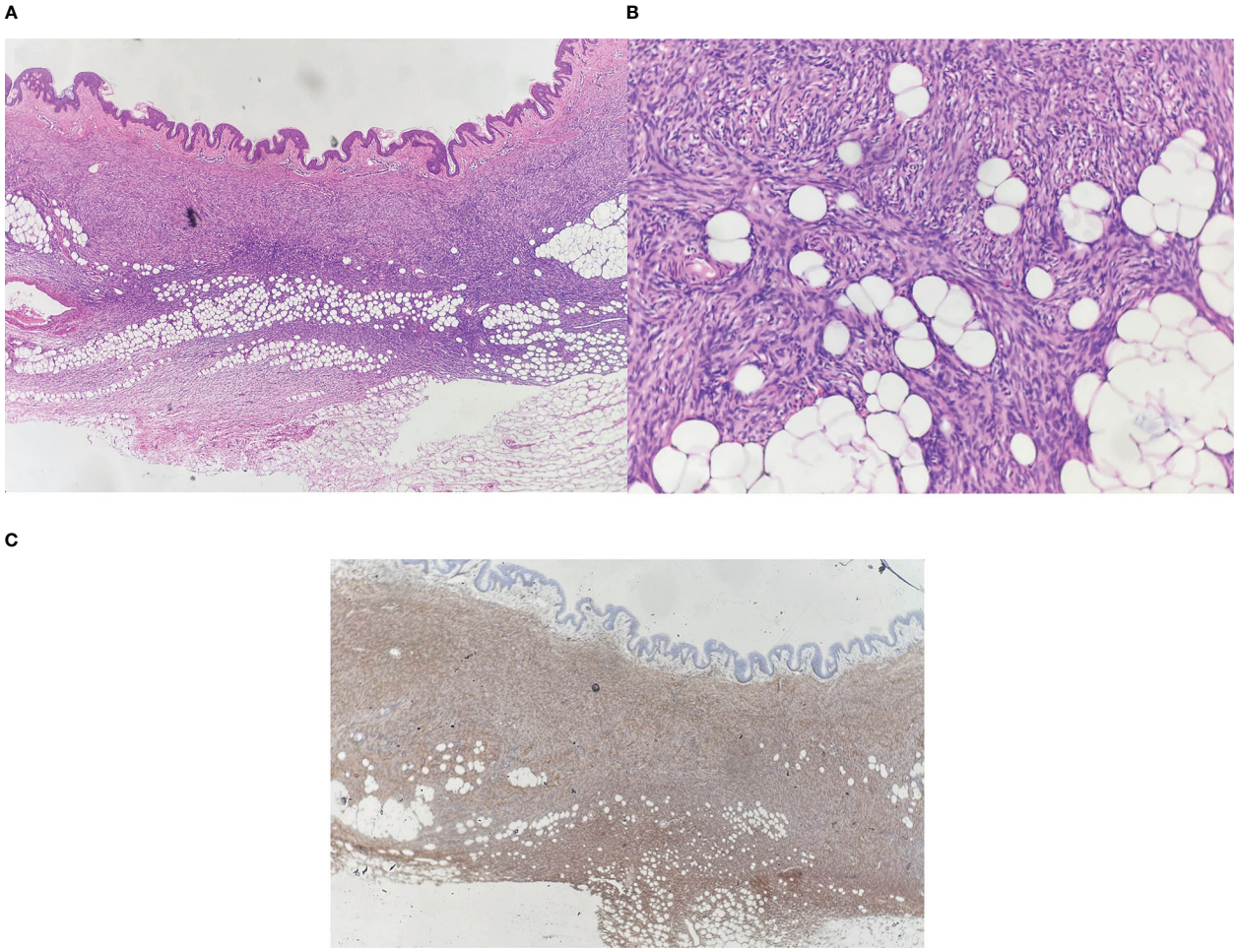

FIGURE 2
(A) Spindle cells infiltrate into the deep dermis and subcutaneous tissue, Hematoxylin-eosin (HE) ×40; **(B)** Dense proliferation of spindle cells in a storiform pattern, HE ×100; **(C)** Spindle cells are strongly positive for CD34, ×40.

immunoprofile, atrophic DFSP should be differentiated from other spindle cell tumors, including dermatofibroma, solitary fibrous tumor, spindle cell/pleomorphic lipomas, neurofibroma, superficial CD34-positive fibroblastic tumor, and medallion-like dermal dendrocyte hamartoma (7).

Molecular analysis is used to detect the COL1A1-PDGFB fusion gene, which is specifically expressed in DFSP and not related to pathological subtypes. FISH and NGS are two main molecular techniques for detecting the fusion gene. FISH is a straightforward method performed with either fusion probes or break-apart probes, which has good sensitivity and specificity. However, it doesn't provide detailed breakpoint information on both translocation partners. NGS is another technique for detecting fusion transcripts. Previous studies have shown that NGS was more sensitive than FISH for the COL1A1-PDGFB fusion detection, especially in borderline cases (8). Additionally, NGS can effectively detect all fusion genes and fusion partners, which can make up for the shortcomings of conventional detection methods, such as missed detection, false negatives, and the inability to distinguish fusion partners. However, there are also some challenges with data analysis and interpretation, analytical validation, high-quality sample tissues, and high testing costs (9).

Surgical excision is the gold standard for the primary treatment of DFSP with wide excision and margin-controlled removal (i.e. Mohs and slow Mohs) as viable options (10, 11). For cases with recurrent, unresectable, and metastatic tumors, radiation therapy and targeted immunotherapy such as tyrosine kinase inhibitors have been shown to be effective (12). Imatinib mesylate can competitively inhibit ATP binding to the PDGF-β receptor, which slows down kinase activity, limits the growth of the tumor, and promotes apoptosis (13).

Till now, 91 cases with atrophic DFSP, including ours, have been reported (Table 1). The age ranges from 7 months to 72 years with an average of 26.7. It seems like the atrophic variant most frequently occurs during the second to fifth decades of life (n=65), which is younger compared with classic DFSP. Children and congenital cases account for 34% of all cases (n=31). In contrast to classic DFSP has no sex bias (10), atrophic DFSP shows a preference for females with a F: M ratio of 3:1. Trunk and extremities are the most frequently involved sites (n=87), which are consistent with classic DFSP (1). The tumor size ranges from 0.5 to 13cm in greatest diameter. Clinical diagnosis includes morphea, morpheaform basal cell carcinoma, anetoderma, lymphocytoma, lipoatrophy, atrophic scar, and neurofibroma. Delayed diagnosis and misdiagnosis are common. 5/8 cases of atrophic pigmented DFSP were misdiagnosed as benign lesions, including nevus of Ota, postinflammatory hyperpigmentation, hemangioma, and lipoatrophy (20, 55, 61). A high index of

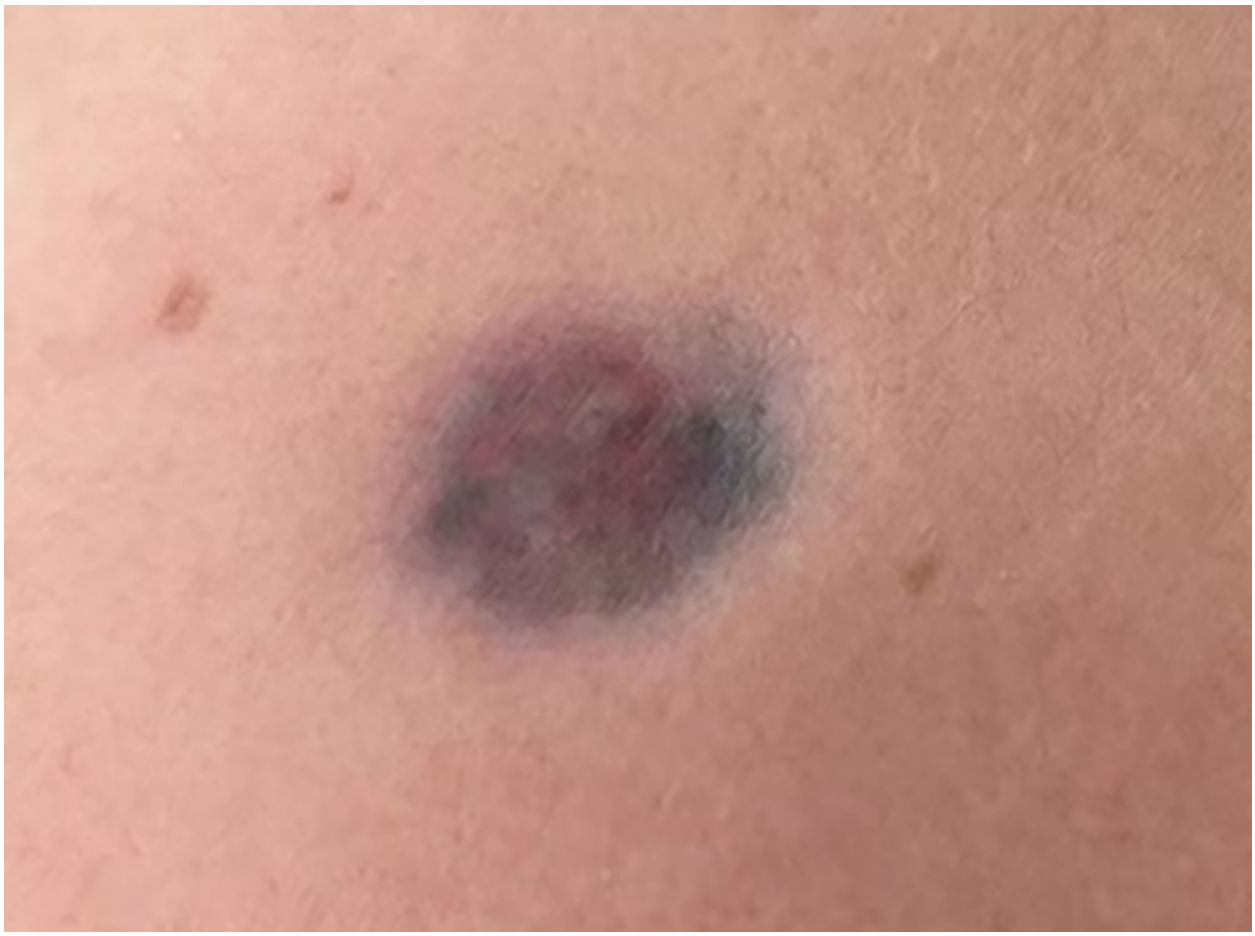

FIGURE 3
A round erythematous-to-bluish atrophic plaque.

TABLE 1 Reported cases of atrophic dermatofibrosarcoma protuberans.

Case (No.)	Age(y)	Sex	Site	Size(cm)	Clinical diagnosis	Reference
1	15	M	Supraclavicular	5×7	Morphea, morpheaform BCC	(5)
2	50	F	Inferior aspect of abdomen	3×6	DFSP	(5)
3	37	F	Chest	5×8	Morphea, morpheaform BCC	(5)
4	15	M	Posterior aspect of shoulder	3×5	Morphea	(5)
5	28	M	Inferior aspect back	2.5	N/S	(5)
6	25	F	Subclavicular	2×3	BCC	(14)
7	21	F	Upper aspect of back	1×1.5	Anetoderma	(15)
8	27	F	Upper aspect of back	2×3	Sclerosing BCC	(15)
9	7	F	Upper Arm	N/S	DFSP	(16)
10	1	M	Lower Back	N/S	DFSP	(16)
11	12	F	Back	N/S	DFSP	(16)
12	9	F	Shoulder	N/S	DFSP	(16)
13	6	M	Right shoulder	N/S	DFSP	(16)
14	5	M	Back	N/S	DFSP	(16)

(Continued)

TABLE 1 Continued

Case (No.)	Age(y)	Sex	Site	Size(cm)	Clinical diagnosis	Reference
15	10	F	Buttock	N/S	DFSP	(16)
16	10	F	Forefoot	N/S	DFSP	(16)
17	25	F	Shoulder	2.5×1.5	N/S	(17)
18	16	F	Left periumbilical region	3×4	Congenital atrophic DFSP	(18)
19	40	F	Periumbilical	N/S	Lymphocytoma	(19)
20	55	F	Shoulder	3×6	DFSP, lymphocytoma	(19)
21	42	F	Groin	N/S	BCC, scar	(19)
22	24	F	Infraorbital	N/S	Nevus of Ota	(20)
23	1.5	M	Left ankle	3.5×6.5	DFSP	(21)
24	42	M	Shoulder	2×3	N/S	(22)
25	21	F	Subclavicular	2×3	Anetoderma	(23)
26	53	M	Left clavicle	7×5	N/S	(24)
27	13	M	Left calf	4	N/S	(25)
28	3	F	Right thigh	0.5	Morphea	(25)
29	16	F	Left calf	3	Atrophic plaque	(25)
30	1.5	M	Left ankle	N/S	Atrophoderma, lipoatrophy	(25)
31	3	F	Periumbilical	5×4	Morphea	(25)
32	21	M	Epigastric	N/S	DFSP	(25)
33	72	F	Midback	3×1.5	N/S	(26)
34	16	F	Right leg	6×8	N/S	(27)
35	40	M	Chest	6	DFSP	(28)
36	26	F	Right breast	1×2	Atrophic DFSP	(29)
37	65	M	Lower aspect of back	2.5×3	Anetoderma, atrophic scar	(30)
38	41	F	Right Chest	4×5	Atrophic DFSP	(31)
39	23	M	Left anterior shoulder	3.3 × 2.8	Atrophic DFSP	(32)
40	16	F	Right thigh	N/S	DFSP	(33)
41	48	F	Left upper abdomen	2×0.8	Atrophic DFSP	(34)
42	55	F	Epigastric region	2×1.5	Atrophic DFSP	(35)
43	29	F	Right thigh	10	Atrophic DFSP	(36)
44	36	M	Left cheek	4×4	DFSP	(37)
45	30	M	Mid-back	5×3	Atrophic DFSP	(38)
46	7mo	M	Left upper groin	1.6×0.7	Atrophic DFSP	(39)
47	14	F	Right leg	4×3	Atrophic DFSP	(40)
48	18	F	Left supraclavicular	2	Atrophic DFSP	(41)
49	14	M	Hand	2.5	Congenital atrophic DFSP	(41)
50	8	F	Right thigh	2.5×2	Congenital atrophic DFSP	(41)
51	52	F	Left buttock	6 x 5	Atrophic DFSP	(42)
52	40	F	Right pars lumbalis	N/S	Atrophic DFSP	(43)
53	30	F	Upper abdomen	6×5	Atrophic DFSP	(44)
54	5	M	Right buttock	N/S	Congenital DFSP	(45)

(Continued)

TABLE 1 Continued

Case (No.)	Age(y)	Sex	Site	Size(cm)	Clinical diagnosis	Reference
55	36	M	Left chest	8×10	Morphea	(46)
56	30	F	Left Waist	1.3×0.8	Atrophic DFSP	(47)
57	37	F	Left shoulder	1×1.5	Atrophic DFSP	(48)
58	23	F	Back	2	Atrophic DFSP	(49)
59	24	F	Chest	N/S	Atrophic DFSP	(50)
60	19	F	Right precordium	2.5×3	Congenital atrophic DFSP	(51)
61	34	F	Left buttock	1.1×1.2	Atrophic pigmented DFSP	(52)
62	6	F	Right lower abdomen	4×2.5	Atrophic DFSP	(53)
63	15	M	Back	N/S	Atrophic DFSP	(54)
64	7	F	Left wrist	2×4	Lipoatrophy	(55)
65	8	F	Left forearm	1×1	Hemangioma	(55)
66	48	F	Anterior chest wall	13×12	Atrophic DFSP	(56)
67	31	F	Left lower abdominal wall	1.5	DFSP	(4)
68	14	M	Right upper abdominal wall	2	Spindle cell neoplasm, suspicious of DFSP	(4)
69	7	M	Left forearm	0.5	Atrophic pigmented DFSP	(4)
70	45	F	Infraclavicular region	N/S	DFSP	(4)
71	23	F	Left chest wall	1.5	Neurofibroma	(4)
72	44	M	Right back	2.5	Neurofibroma	(4)
73	27	F	Shoulder	2	Possible DFSP	(4)
74	19	M	Right chest wall	2.5	DFSP	(4)
75	28	F	Infraclavicular region	1	Neurofibroma	(4)
76	28	F	Chest wall	1.5	DFSP	(4)
77	24	F	Left lower neck	2	Spindle cell neoplasm	(4)
78	48	F	Left cheek	1.4	Spindle cell neoplasm	(4)
79	47	F	Left chest wall	1	Spindle cell neoplasm	(4)
80	34	M	Back	1.2	Spindle cell neoplasm, suspicious of DFSP	(4)
81	36	F	Left groin	2	Spindle cell neoplasm	(4)
82	63	F	Chest wall	1	Cutaneous diffuse neurofibroma	(4)
83	30	F	Chest	4×2	Atrophic DFSP	(57)
84	N/S	F	Right inferior breast	4×3.5	Atrophic DFSP	(58)
85	19	F	Right chest	12×5	Congenital atrophic DFSP	(59)
86	26	M	Left back	3×2.5	Atrophic pigmented DFSP	(60)
87	33	F	Left upper back	1.6 × 1.3	Postinflammatory hyperpigmentation	(61)
88	39	F	Right lower leg	N/S	Dermatofibroma	(62)
89	39	F	Abdomen	N/S	Morphea	(63)
90	31	F	Right chest	1.6×1	Atrophic DFSP	Our case
91	45	M	Left shoulder	N/S	Atrophic pigmented DFSP	Our case

BCC, Basal cell carcinoma; DFSP, dermatofibrosarcoma protuberans; F, female; M, male; N/S, not stated.

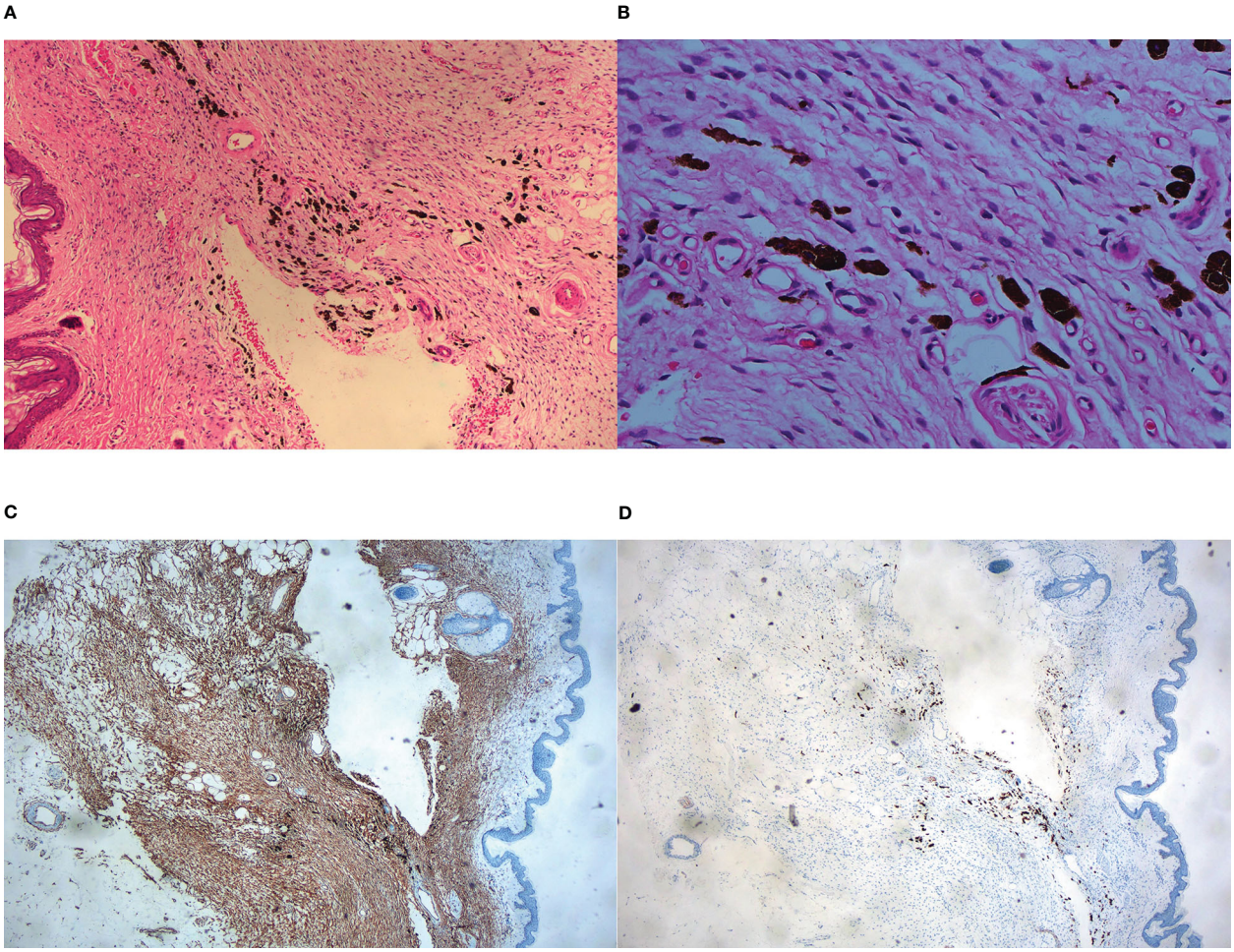

FIGURE 4
Histopathological examination. **(A)** Fascicles of spindle cells with reduced dermal thickness by about half on low-power view, HE ×40; **(B)** Monomorphic spindle cells with bland cytoplasm and scattered dendritic cells containing abundant melanin, HE ×200; **(C)** Immunohistochemistry showed $CD34^+$ cells infiltrating into subcutis and forming honeycomb-like pattern, ×40; **(D)** Immunohistochemistry showed spindle cells were negative for S-100, in contrast, melanin-bearing dendritic cells were positive for S-100, ×100.

suspicion and a broad differential diagnosis by dermatologists is necessary.

In summary, atrophic DFSP is a diagnostic challenge to both clinicians and pathologists due to its atypical clinical presentation. Because of the initial benign and indolent behavior, many patients only seek medical care years after its onset. Here, we present a case series to further characterize its clinical and pathological features and enhance recognition of atrophic DFSP. As with these cases, atrophic lesions with no apparent cause and no symptoms should be aware of atrophic DFSP in the early stage. Histopathologic, immunohistochemical, and molecular examinations are necessary to help reduce misdiagnosis and improve prognosis.

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.

Author contributions

YL analyzed the clinical data and drafted the manuscript. ZC collected the data and reviewed literature. SN managed the patient. ZW designed 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.

Publisher's note

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