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# Case Report: Expanding the ocular horizon: subperiosteal orbital hematoma and Roth spots in known case of sickle cell disease

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Sickle cell disease is a prevalent haemoglobinopathy that is hereditary. People with sickle cell disease frequently experience vaso-occlusive crises, which lead to substantial morbidity from end-organ ischaemia and infarction. Numerous organs, including the bones, brain, lungs, spleen, retina, and penis, may be affected. As a result, symptoms may include painful bone crises, cerebrovascular accidents, acute chest syndrome, sequestration crises, retinal haemorrhage, and priapism. In sickle cell disease, subperiosteal orbital haemorrhage is a rare vaso-occlusive crisis (VOC). Roth spots (white-centred, flame-shaped retinal haemorrhages), occurring commonly in subacute bacterial endocarditis, can also be seen in leukaemia, pre-eclampsia, and hypertensive retinopathy. A 19-year-old male patient with sickle cell disease (HbSS) presented to the hospital with ptosis and swelling in his right periorbital region 3 days after being brought to the hospital for a sickle cell crisis. It was later determined to be an acute subperiosteal orbital hematoma along with Roth spots, an uncommon clinical manifestation of sickle cell disease, only after a thorough clinical and radiological assessment.

## KEYWORDS

sickle cell disease, vaso-occlusive crisis, subperiosteal orbital hematoma, Roth spots, magnetic resonance imaging

## Introduction

Sickle cell disease is one of the most prevalent hereditary autosomal recessive monogenic haematological disorders that affect red blood cells (1–3). The gene-coding defect produces rigid, sticky, crescent-shaped red blood cells that cannot pass through the capillary bed, leading to ischaemia-reperfusion damage in organs (4, 5).

Vaso-occlusive crises are the predominant feature of sickle cell disease. All organ systems are impacted by vaso-occlusive crises, causing end-organ ischaemia and infarction,

leading to high morbidity and early mortality. A diagnostic conundrum arises because the clinical presentation of the end-organ vaso-occlusive crisis may resemble that of several other clinical diseases. Splenic infarction (6), pulmonary involvement, acute chest syndrome (7), and orbital compression syndrome (8, 9) are among the recent instances of end-organ ischaemia.

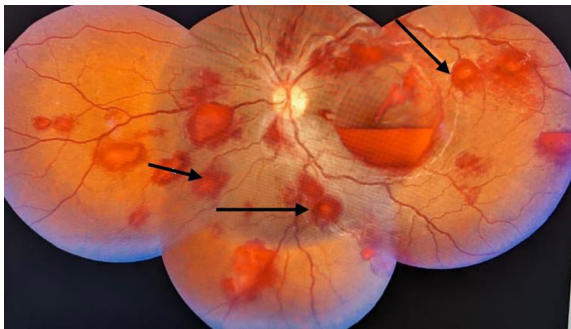
Roth spots, characterised by white-centred retinal haemorrhages, can be caused by several factors. Still, the most common theory links them to retinal capillary rupture brought on by endothelial cell dysfunction. After the vessel ruptures, red blood cells spill out, causing fibrin–platelet plugs to develop on the injured endothelial cell lining and starting the clotting chain reaction. Sick cell anaemia is a relatively rare but significant disorder among the many others linked to Roth spots (10, 11).

Orbital involvement in sickle cell disease is rare, but when it occurs, it can be made worse by orbital compression syndrome, which occurs when the bone marrow’s microvasculature is infarcted, leading to inflammation and necrosis, ultimately resulting in subperiosteal haemorrhage (12).

The rupture of diploic veins between the periosteum (periorbita) and the bony orbit results in subperiosteal orbital haemorrhage. A local haemorrhage from a rupture creates a haematoma between the bone and the periorbita. The periorbita peels away from the bone as a result of the pressure being raised by the blood, taking up space. Blood will not enter the orbit if the periorbita is intact, and imaging will show that the haematoma will have a bilenticular shape. If left untreated, a subperiosteal orbital haematoma can expand over time or resolve spontaneously. Generally, it is treated medically, but in cases of optic nerve dysfunction or extensive haematomas, it is treated surgically (13).

### Case presentation

A 19 year old male patient with a known case of sickle cell disease (HbSS) rushed to the hospitals emergency room with a high grade fever that had been present for 3 days. The fever was insidious in onset, progressed over time, and was not resolved by medications. He also complained of pain and swelling over his right eyelid associated with frontal and occipital headaches.



**FIGURE 1**  
A fundus image of a right eye with round flame-shaped haemorrhages and a white/pale centre suggestive of Roth spots.

On admission, the patient appeared pale, drowsy, and sick. The patient’s axillary temperature was 38.2°C, his heart rate was 115 beats per minute with regular rhythm and normal volume with no special character, and he had abdomino-thoracic breathing with a respiratory rate of 25 cycles per minute. His blood pressure was 100/60 mm Hg in the right brachial artery in the supine position, and his oxygen saturation was 96% on room air. The patient’s weight was 45 kg.

**TABLE 1** Table showing laboratory parameters of the patient and the normal laboratory values.

Laboratory parameters	Patient's value	Normal value
Haemoglobin (g%)	6	13–15
Mean Corpuscular Volume (MCV) (fL)	92.5	80–100
Platelets (lacks/cu mm)	1.63	1.5–4.0
White blood cells (cu mm)	10,600	4,000–10,000
Red blood cells (million/cu mm)	2.01	3.8–4.8
Erythrocyte Sedimentation Rate (ESR)	35	30
International Normalised Ratio (INR)	1.13	1.0–1.3
Serum urea (mg/dL)	20	7–17
Serum creatinine (mg/dL)	0.5	0.52–1.04
Serum sodium (mmol/L)	136	137–145
Serum potassium (mmol/L)	4.6	3.5–5.1
Serum magnesium (mg/dL)	1.7	1.6–2.3
Serum phosphorous (mg/dL)	3.5	2.5–4.5
Serum calcium (mg/dL)	8.8	8.4–10.2
Alkaline phosphatase (U/L)	123	38–126
Aspartate aminotransferase (U/L)	45	14–36
Alanine aminotransferase (U/L)	38	<35
Total bilirubin (mg/dL)	1.8	0.2–1.3
Conjugated bilirubin (mg/dL)	1.0	0.0–0.3
Unconjugated bilirubin (mg/dL)	0.8	0.0–1.1
Globulin (g/dL)	3.9	2.3–3.5
Albumin (g/dL)	3.3	3.5–5.0
Random Blood Sugar (RBS) (mg/dL)	124	110–140
Dengue, leptospirosis, Widal, malaria	Non-reactive	–
Peripheral smear	Normocytic, normochromic with anisopoikilocytosis showing pencil cells	–
Urine routine and microscopy	Normal	–

Upon general examination, the patient showed evident pallor without any icterus, cyanosis, clubbing, lymphadenopathy, or oedema. The ocular examination revealed peri-ocular soft tissue enlargement, right eyelid oedema, and restriction in the right eye's elevation.

Using the colour plates, colour vision was assessed and found to be normal. The pupils were equal in size and were reactive to light. Both pupils reacted normally to the swinging light reflex. Afferent pupillary light reflex deficit (APD) was absent. Both eyes' intraocular pressures were within normal limits. Fundus examination revealed round flame-shaped haemorrhages with a white/pale centre suggestive of Roth spots (Figure 1).

No apparent abnormalities were found after a thorough, comprehensive assessment of the cardiovascular, respiratory, central nervous, and gastrointestinal systems. The following are the results of routine and special laboratory investigations (Table 1). The blood culture results were negative, and the urine analysis was normal.

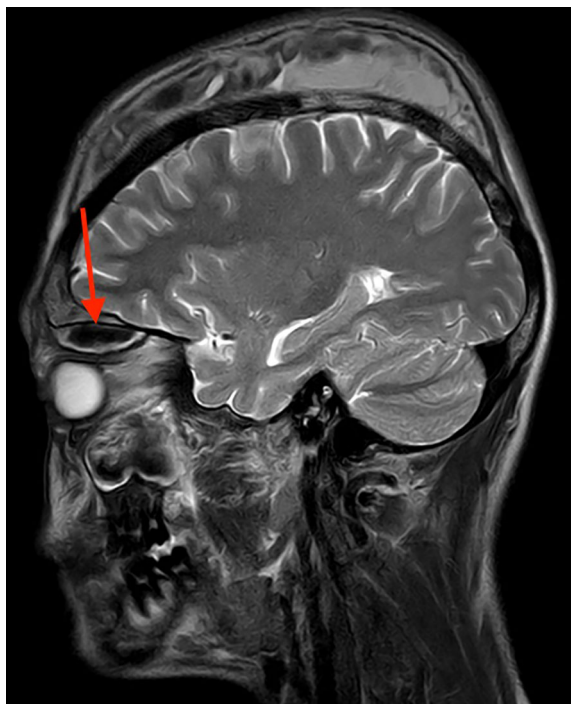
Plain MRI and contrast MRI of the right orbit were conducted, showing well-defined elliptical altered intensity lesions noted in the extraconal space of the superior quadrant and appearing heterogeneously hyperintense on T1 Weighted Image (T1WI) (Figure 2) and hyperintense on T2 Weighted Image (T2WI)/Fluid Attenuated Inversion Recovery (FLAIR), causing mass effect over the superior oblique and superior rectus muscles, suggestive of subperiosteal orbital hematoma. A contrast MRI of the brain was

conducted, revealing the heterogeneous signal intensity of the calvarium, suggestive of bone infarcts with subperiosteal collection involving the bilateral frontal and parietal bones, appearing hyperintense on T1WI and heterogeneously hyperintense on T2WI (Figure 3).

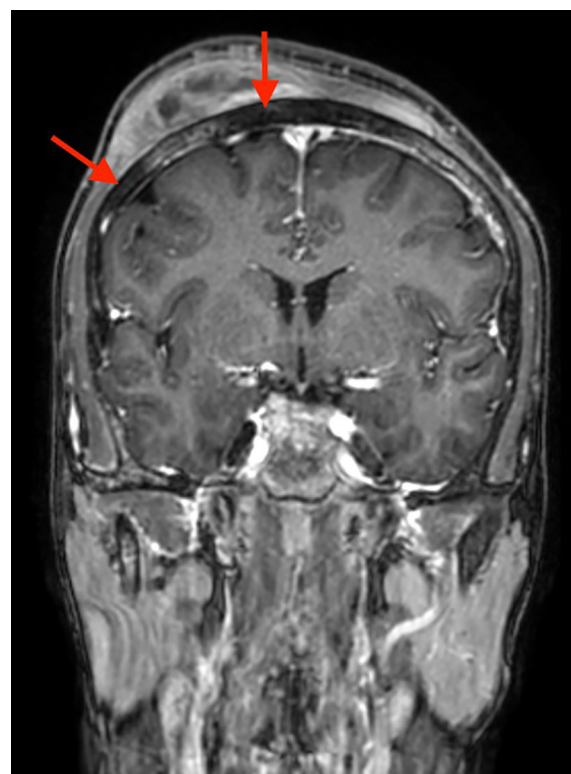
The patient was promptly given injectable methylprednisolone pulse therapy, intravenous fluids, analgesics, wide-spectrum antibiotics, hydroxyurea, and two units of packed red blood cell transfusions. Following a favourable response to medical treatment and full recovery, the patient was discharged once his condition was stabilised.

## Discussion

Ocular manifestations of sickle cell disease have been reported; when these manifestations are interconnected to the retina, they are called sickle cell retinopathy. Sickled red blood cells cause vaso-occlusion of the retinal microvasculature, which results in ischaemia and subsequent retinal tissue destruction. This is the pathogenesis of sickle cell retinopathy. Clinical manifestations of sickle cell retinopathies are often neovascularisation, cotton-wool spots, macular ischaemia, peripheral retinal vascular occlusion, and retinal haemorrhages (14).



**FIGURE 2**  
MRI orbit (plain) T2WI in sagittal view having well-defined peripherally enhancing elliptical altered signal intensity lesion measuring  $27 \times 10$  mm in extraconal space of superior quadrant of the right orbit, appearing hyperintense, causing mass effect over superior oblique and superior rectus muscles.



**FIGURE 3**  
MRI brain (contrast) T1WI in coronal view with the heterogeneous signal intensity of the calvarium, suggestive of bone infarcts with subperiosteal collection involving bilateral frontal and parietal bones measuring approximately  $7.8 \times 4.4 \times 1.4$  cm on the right side and  $4.4 \times 1.3 \times 6.4$  cm on the left side, appearing hyperintense.

Orbital bone infarction, followed by an inflammatory reaction that can quickly spread to the orbit and cause orbital pain and proptosis, is the primary mechanism for developing orbital compression syndrome (9), an acute illness. The hallmark features of orbital compression syndrome (OCS) include eyelid oedema, proptosis, periorbital pain, and restriction of extra-ocular movement, with or without reduced visual acuity. The development of haematomas, which can be intracranial (epidural) or orbital (sub-periosteal), is a distinctive characteristic of orbital wall infarction in SCD (15). Subperiosteal haemorrhage has been attributed to several processes, including mild trauma, underlying bleeding diathesis, and blood extravasation from necrotised vessel walls (16). In this instance, orbital infarction was thought to be the aetiology of the orbital haematoma, as evidenced by aberrant frontal bone and bone marrow heterogeneous signals adjacent to the hematoma formation.

Since children's orbital bones have greater marrow space than adults', orbital wall infarction usually affects young patients (16). The youngest recorded age was 2 years, while the average age of presentation in the instances that were documented was 13 years. Nearly all cases had a fever as a presenting symptom, and over two-thirds of the patients had associated pain crises elsewhere (17).

Intravenous corticosteroid administration may alleviate orbital pressure brought on by the inflammatory component of orbital bone infarction. Since osteomyelitis and bone infarction are sometimes difficult to distinguish clinically, concurrent antibiotic coverage is advised. If there are signs of optic nerve dysfunction, surgical exploration and haematoma evacuation are necessary to avoid vision loss and ensure prompt recovery. Approximately one-fifth of the patients reported needing surgical procedures, whereas the majority of patients recovered with medical therapy (15). Orbital infarction is a rare manifestation of sickle cell disease, which may also be considered a vaso-occlusive crisis. The management is the same as for other crises: potent analgesics, cautious yet vigorous hydration, and a comprehensive assessment of the underlying cause (18).

## Conclusion

Sickle cell disease is known to have ocular manifestations, known as sickle cell retinopathy, which includes neovascularisation, ischaemia, and occlusion of retinal vessels. However, Roth spots are an uncommon finding in sickle cell disease, which indicates complex ocular manifestations. Orbital infarction leading to subperiosteal orbital hematoma is another very rare vaso-occlusive crisis of sickle cell disease. Therefore, any patient with a known sickle cell disease presenting to the hospital with complaints of unilateral orbital swelling should be further evaluated with magnetic resonance imaging to rule out orbital hematoma and fundus examination to look for Roth spots. Careful assessment, diagnosis, and the timely initiation of the proper supportive treatment are highly recommended to avoid irreversible vision loss. The majority of ocular involvement cases do not need specialised treatment; instead, they respond well to conservative treatment, including adequate hydration, analgesics, and hydroxyurea.

In conclusion, even though they are uncommon, orbital infarction, hematoma, and retinal haemorrhage should be suspected in all sickle cell disease individuals who present with eyelid swelling.

## 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 studies involving humans were approved by the Datta Meghe Institute of Higher Education and Research Institutional Ethics Committee. 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

RP: Conceptualization, Data curation, Formal Analysis, Funding acquisition, Investigation, Methodology, Project administration, Resources, Software, Supervision, Validation, Visualization, Writing – original draft, Writing – review & editing. SA: Formal Analysis, Supervision, Writing – review & editing. VK: Formal Analysis, Writing – review & editing. RD: Formal Analysis, Writing – review & editing. SS: Formal Analysis, Writing – review & editing. MK: Formal Analysis, Writing – review & editing.

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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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