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**Case Report**

# Subperiosteal Orbital Hematoma: A Rare Clinical Manifestation of Sickle Cell Disease - A Case Report

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**Keywords**
Sickle cell disease · Subperiosteal hematoma · Orbit · Sphenoid bone · Osteomyelitis

**Abstract**
Sickle cell disease (SCD), an inherited vaso-occlusive disorder, results in recurrent painful episodes and a variety of serious systemic complications that can lead to severe disabilities and even death. Here, we report a case of a 19-year-old African American patient with homozygous sickle cell trait who presented with right upper lid edema and ptosis, 3 days after his admission to the hospital following a sickle cell crisis. Initially, mistaken as a superinfection in the context of his disease, a diagnosis of orbital abscess was made. Intravenous antibiotics and a proper treatment plan were set accordingly. Only after extensive clinical and radiological examinations, it turned out to be an acute subperiosteal orbital hematoma, a rare clinical manifestation of SCD. The aim of our case report was to highlight the difference in orbital presentation between osteomyelitis and subperiosteal hematoma, as well as spreading awareness among medical professionals and especially ophthalmologists for this rare presentation of orbital wall infarction, as the initial differential diagnosis of SCD patients with ocular involvement.



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## Introduction

Sickle cell disease (SCD) is an autosomal-recessive inherited disorder with high morbidity and mortality. SCD is caused by a mutation of the seventh codon of the $\beta$-globin gene, GAG, which is substituted by GTG resulting in the synthesis of a mutated hemoglobin named "hemoglobin S" (HbS) found in erythrocytes [1]. When deoxygenated, HbS polymerizes, impairing and changing the shape of the erythrocyte into an elongated sickle, commonly referred as a sickle shape [1, 2]. The clinical manifestations of SCD are mainly marked by vaso-occlusive phenomena and hemolytic anemia which can lead to acute and chronic pain and tissue ischemia or infarction. SCD can affect all organs and can ultimately lead to various systemic complications such as acute thoracic syndrome, renal dysfunction, pulmonary arterial hypertension, and stroke [2]. These clinical events occur much more frequently while others are more rare in presentation [1, 2]. Here, we present the case of a young patient with an unusual clinical manifestation of SCD who was admitted to our hospital following a sickle cell crisis. The CARE Checklist has been completed by the authors for this case report, attached as online supplementary material (for all online suppl. material, see https://doi.org/10.1159/000532016).

## Case Presentation

A 19-year-old African American man with homozygous sickle cell trait was admitted to our hospital following a sickle cell crisis. Three days after his admission, the patient developed a painless subacute right upper eyelid swelling and mild ptosis (shown in Fig. 1). On the physical examination, no pain, ophthalmoplegia, or afferent pupillary defect was noted. Extraocular muscles were within normal limits. No diplopia or dystopia and Ishihara were unremarkable. Margin Reflex Distance-MRD1: 1/4 (OD: right eye/OS: left eye). Mild right upper lid swelling with comparable eyelid crease height was seen bilaterally and symmetric Hertel exophthalmometer findings were noted. Vision OD/OS 20/20 and no change or blurriness were noted. The rest of the intraocular examination was completely normal. A computerized tomography (CT)-scan was already done earlier that day showing a lenticular-shaped extraconal mass along the right superolateral orbit of 2 × 0.7 × 1.2 cm with no surrounding inflammation (shown in Fig. 2). Given the patient's clinical context, this emerging clinical manifestation was initially considered by infectious disease specialists as an orbital abscess with post-septal cellulitis and was therefore managed by intravenous antibiotic treatment. Given the poor clinical improvement 24 h later and after extensive clinical and radiological correlations, we decided to obtain additional imaging to better understand the situation. A magnetic resonance imaging (MRI) (shown in Fig. 3) revealed a rim enhancement-like of the extraconal mass which is typical of bony infarction and an increased T2 signal with associated diffusion restriction in the adjacent calvarium and right lateral orbital bones. Multiple other sites of heterogeneous marrow were also observed throughout the calvarium. These results are more likely to indicate bone infarction than an intra-orbital abscess or osteomyelitis, which according to these outputs were less likely. Given the patient medical history, clinical manifestations, and imaging findings, a subperiosteal orbital hematoma (SOH) was therefore diagnosed. A conservative management with supportive corticosteroids was initiated with a drastic improvement of the patient's condition in a week time.

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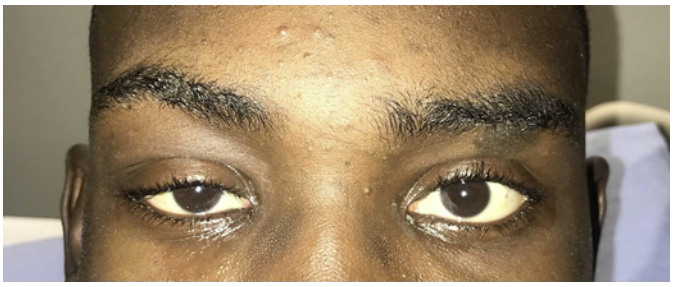

**Fig. 1.** Right upper eyelid swelling and ptosis.

## Discussion and Conclusion

SCD has a high prevalence across the regions of sub-Saharan Africa, the Mediterranean basin, Middle East, and India [2]. Patients with SCD are more likely to develop recurrent episodes of vaso-occlusion due to HbS polymerization, leading to erythrocyte rigidity and aggregation in blood circulation [1, 2]. This repetitive event is damaging to the vascular system and leads to tissue ischemia and injury resulting in an acute systemic painful vaso-occlusive crisis that requires urgent medical care [1, 2].

SCD's clinical manifestations are protean and their severity can vary considerably among the major genotypes and even among patients with the same genotype [2, 3]. One of the common clinical manifestations of SCD is bone infarction and all bones with active marrow can be affected. In general, long bones and vertebrae are the most involved and, less frequently, bones with little marrow space such as the orbital wall [4, 5]. However, the latter can still be affected, as per our case, particularly in children and adolescents, as they exhibit more marrow space in the orbital bones especially the orbital roof [4, 6].

Ophthalmic manifestations in SCD usually include anterior segment ischemia, glaucoma, angioid streaks, retinopathy, and retinal artery occlusions [6–8]. In this case report, our patient presented with an acute SOH, which, based on the existing data, is an uncommon manifestation of SCD. The etiology of SOH is currently still misunderstood. SOH has been suggested to be related to the extravasation of blood from necrotized vessel walls, underlying bleeding diathesis, and minor trauma [6, 7, 9, 10]. In the literature, SOH has been reported in several other cases of patients with SCD [6, 7, 9]. In these cases, orbital wall infarction with subperiosteal hemorrhage was often accompanied with eyelid swelling, with or without visual impairment. Then, subsequent to the bone infarction, the inflammatory response triggered in the orbit can alter important structures, resulting in an orbital compression syndrome. Finally, isolated cases of SOH have also been reported within other clinical context, following sinusitis [10, 11] in laboring patients [12, 13] and in diseased patient [14, 15].

The distinction between bone infarction and osteomyelitis is definitely challenging. Indeed, patients with SCD are immunocompromised and are more susceptible to infections, especially osteomyelitis, which shares many clinical features with bone infarction [5, 6, 9]. In order to differentiate between bone infarction and osteomyelitis, a combination of clinical features, laboratory workups, and imaging investigations must be assessed by the physician to their discretion [16]. The use of contrast-enhanced MRI, the current modality of choice for diagnosing osteomyelitis, would further aid in differentiating infarction over osteomyelitis [5, 16]. In our case, the results of the first ophthalmic examination and computerized tomography scan revealed no signs of inflammation of the extraocular muscles and fatty tissues of the orbit and no ophthalmic manifestations such as ophthalmoplegia, pain with eye movements, thus questioning the initial diagnosis of post septal cellulitis. Despite there being eyelid swelling in

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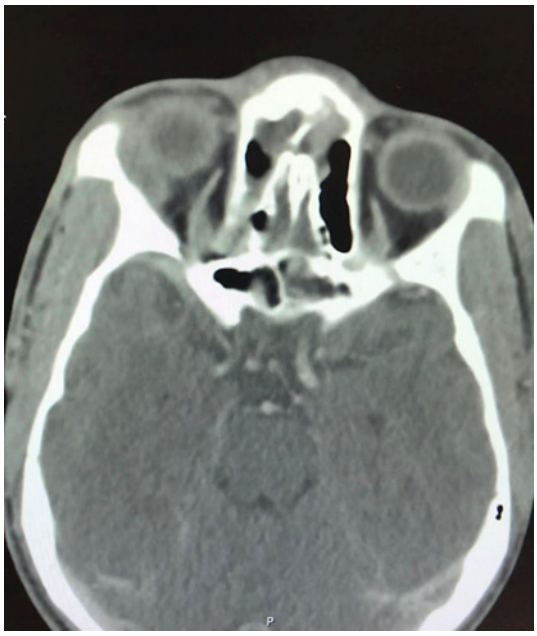

**Fig. 2.** Axial computed tomography (CT)-scan: lenticular shaped extraconal mass along the right superolateral orbit of 2 × 0.7 × 1.2 cm.

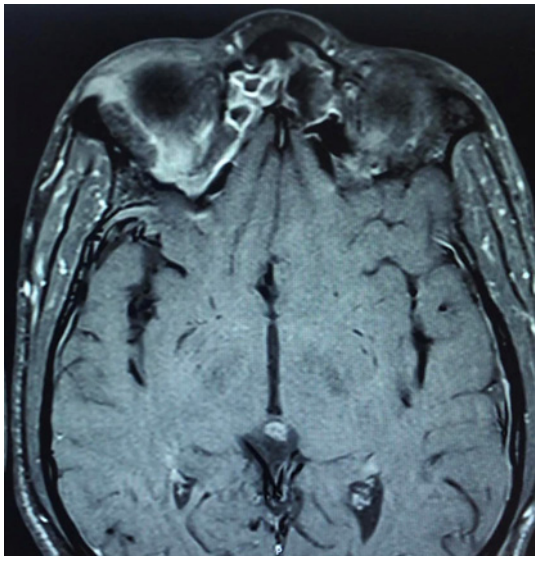

**Fig. 3.** Axial magnetic resonance imaging (MRI): rim enhancement-like of the hypointense extraconal mass on T1 weighted.

the presentation of both SOH and orbital cellulitis, they can be distinguished based on their clinical and radiological features. Clinically, if conservative corticosteroid treatment yielded no improvement of symptoms after 2 weeks, it would lead the diagnosis toward infection, or even osteomyelitis, over infarction [5]. Here, MRI played a pivotal role in the final diagnosis by detecting the hemorrhagic signal in the bone marrow and the subperiosteal collections which are specific features for differentiating infarction from other inflammatory conditions.

In this case report, our patient had a very mild form of SOH which did not endanger his vision. We therefore decided to adopt a nonsurgical management, which is the preferred method of treatment according to the literature, as SCD patients often recover spontaneously

with added benefit through supportive measures [6]. A conservative treatment with supportive corticosteroids associated with regular clinical follow-up is indeed sufficient to manage SOH. However, when the SOH becomes vision-threatening, a surgical orbital decompression is ultimately indicated. Ophthalmologists caring for patients with SCD should be aware and always consider the possibility of a SOH and differentiate it from an orbital abscess or osteomyelitis when confronted to a similar case.

## Statement of Ethics

The case report was conducted ethically in accordance with the World Medical Association Declaration of Helsinki. Written informed consent was obtained from the patient for publication of this case report and any accompanying images. This study protocol was reviewed and the need for approval was waived by the Ethics Committee of Carrot Health Eye Surgery Clinic.

## Conflict of Interest Statement

No conflict of interest in applicable (e.g., employment, consultancies, honoraria, stock ownership and options, expert testimony, grants or patents received or pending, royalties) which took place in the previous 3 years. The authors have no conflicts of interest to declare.

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## Author Contributions

Najib-Georges Hanna, Asma Musleh, Hasan Khan, Emaan Chaudry, and Corinne Lahoud have all contributed equally in the conception of the idea, writing the manuscript, and supervising the project while adding some touch up to the final version

## Data Availability Statement

The data that support the findings of this study are openly available upon request. All data generated or analyzed during this study are included in this article. Further inquiries can be directed to the corresponding author.

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