Orbital Bone Infarction Masquerading as Periorbital Cellulitis in a Child with Sickle Cell Anemia

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Patient: Female, 4-year-old

Final Diagnosis: Orbital bone infarction • sickle cell orbitopathy

Symptoms: Eye swelling and redness

Clinical Procedure: —

Specialty: Ophthalmology

Objectives: Rare disease

Background: Sickle cell orbitopathy is a rare complication of sickle cell disease that closely mimics other conditions, such as orbital cellulitis and osteomyelitis. We report a case of painless orbital bone infarction masquerading as periorbital cellulitis in a child with sickle cell anemia.

Case Report: A 4-year-old Saudi girl with sickle cell disease presented to our hospital with vaso-occlusive crisis characterized by bilateral lower limb pain and painless left orbital swelling. On examination, she had swelling of the left upper eyelid with redness and mild ptosis (margin reflex distance 1 was 2 mm) without proptosis. Magnetic resonance imaging with contrast showed bilateral sub-periosteal heterogeneous collections (2×0.8×2.1 cm in the superolateral wall of the left orbit and 1×0.6 cm in the inferolateral wall of the right orbit), with intermediate-to-high T1 signal intensity and high T2 signal, causing a mass effect on the adjacent superior and lateral rectus muscles. The patient was treated with systemic antibiotics and supportive treatment for vaso-occlusive crisis under the care of the pediatric team and was discharged without complications.

Conclusions: The diagnosis of sickle cell orbitopathy can be challenging, and an accurate diagnosis is essential to ensure appropriate management. Thus, we report the case of a 4-year-old child with painless sickle cell orbitopathy masquerading as pre-septal cellulitis.

Keywords: Sickle Cell Disease • Vaso-Occlusive Crisis • Orbital Cellulitis • Orbital Bone Infarction • Sickle Cell Disease Orbitopathy • Orbital Osteomyelitis

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Background

Orbital bone infarction, also known as orbitopathy, is a rare complication of sickle cell disease (SCD) and is characterized by an acute onset of orbital pain, erythema, tenderness, and swelling. Moreover, SCD orbitopathy without pain is an extremely rare presentation, which has been described in the literature [1-3]. Magnetic resonance imaging (MRI) is the modality of choice for detecting orbital bone infarction. [4] However, diagnosing SCD orbitopathy remains challenging because of its similarities to more common conditions, such as orbital cellulitis and osteomyelitis [5,6]. Most patients respond well to medical treatment alone, including aggressive rehydration and blood transfusion, with no adverse effects [2,7]. However, in rare cases, surgical intervention can be required. Timely diagnosis and appropriate treatment of this rare condition are crucial to prevent irreversible vision loss [2]. We report the case of a 4-year-old Saudi girl with painless SCD orbitopathy mimicking periorbital cellulitis.

Case Report

A 4-year-old Saudi girl with known SCD on hydroxyurea and folic acid was referred to our tertiary care hospital with vaso-occlusive crisis (VOC) characterized by a 1-day history of bilateral lower limb pain and left painless orbital swelling. The patient was admitted to the Pediatric Department for the management of VOC and was referred for an ophthalmology opinion to rule out left pre-septal cellulitis. The patient’s mother reported that there was no history of upper respiratory tract infection or sinusitis. There was no history of insect bite, eye trauma, or surgery. The patient’s vital signs were stable, and she had a low-grade fever (37.8°C). On inspection, she had left eye swelling and redness without ocular discharge. Due to her lower limb pain, she was not cooperative during the ophthalmological examination. However, the right eye was normal. The left eye had upper eyelid swelling with redness (Figure 1) and mild ptosis (margin reflex distance 1 was 2 mm), without proptosis. She was able to fix and follow with both eyes. The extraocular movement examination was limited; however, she was not very cooperative. The pupils were round and regularly reactive to light, without a relative afferent pupillary defect. Anterior segment examination of the left eye showed mild conjunctival injection without chemosis, mild subconjunctival hemorrhage on the temporal side, clear cornea, a normal iris and lens. Fundus examination of the left eye showed a clear vitreous chamber, a normal optic nerve without swelling, and normal macula and blood vessels, so no further examinations were required.

As a temporary measure, artificial tears and a cold compress were applied. Additionally, the patient received aggressive intravenous hydration and narcotic analgesia to control the pain. While the initial impression was left pre-septal cellulitis, the patient was started on intravenous antibiotics (vancomycin 10 mg/kg every 6 h and ceftriaxone 40 mg/kg twice daily). A computed tomography (CT) scan without contrast was immediately performed to rule out involvement of deeper orbital structures, and she was reviewed daily to monitor her response to treatment and for signs of emerging orbital cellulitis. Initial laboratory findings showed a normal white blood cell count of 14.5×10⁹/L, with high neutrophils 71.5% (normal 20-60%), anemia (hemoglobin 89.5 g/L, reference range 115-135 g/L), with high hemoglobin S quantity of 77.9%. The C-reactive protein (CRP) level was high (116.6 mg/L, reference range 0-5 mg/L). Blood cultures taken on admission were reported as negative after 48 h. During follow-up, her fever subsided, and her left eye redness improved. The patient was cooperative and not in pain. On examination, uncorrected visual acuity was 20/40 in both eyes; extraocular movement of the left eye showed limitation of supraduction -2 and limitation of abduction -1 (Figure 2).

CT showed a left eye superior lateral subperiosteal collection measuring about 2.4×1.4×1.1 cm, with evidence of mass effect.

Figure 1. Left eye upper eyelid swelling and erythema.
Figure 2. Extraocular movements. Limitation of supraduction -2 and limitation of abduction -1 in the extraocular movement of the left eye.
on the adjacent lateral rectus and superior rectus muscles with a tiny focal area seen inside the collection. Another small subperiosteal collection measuring about 1.1×0.5 cm was seen at the inferolateral aspect of the right orbit. There was also evidence of left eye pre-septal cellulitis, as evidenced by mild soft tissue swelling as well as periorbital subcutaneous fat stranding and thickening (Figure 3). An MRI with contrast was advised for proper evaluation of the subperiosteal collection.

The patient’s high hemoglobin S was a contraindication for general anesthesia, owing to the risk of worsening hypoxia and further sickling. Therefore, the MRI was performed under...
Figure 5. Improvement in extraocular muscle movement with marked resolution of the periorbital swelling.

general anesthesia after a blood exchange. MRI with contrast showed bilateral subperiosteal heterogeneous collections, a large collection (2×0.8×2.1 cm) in the superolateral wall of the left orbit, and a small collection (1×0.6 cm) in the inferolateral wall of the right orbit. With intermediate-to-high T1 signal intensity and a high T2 signal, these signals were consistent with subacute hematoma rather than abscesses, tumors, or other potential differential diagnoses of heterogeneous collection [8]. Consequently, this caused a mass effect on the adjacent superior and lateral rectus muscles. Bilateral osseous changes of the sphenoid and lateral orbital wall were also visible (Figure 4).

Based on the clinical presentation and different signal intensities on MRI, the patient was diagnosed with a subperiosteal hematoma, predominantly affecting the left eye, with osseous changes related to bone infarctions from SCD. Although there was evidence of left eye pre-septal cellulitis on clinical examination and CT, the patient showed a slight improvement in swelling after starting systemic antibiotics. On the other hand, there was a dramatic improvement in extraocular muscle movement with marked resolution of the periorbital swelling in the day after blood transfusion (Figure 5).

The patient completed a course of systemic antibiotics and conservative treatment for VOC under the care of the Pediatric Department. She was discharged without complications and did not require any further follow-up with the ophthalmology service.

Discussion

VOC is the most common complication of SCD, accounting for 95% of all admissions. It commonly affects the long bones, including the vertebrae and extremities [9]. However, other uncommon sites can be affected; for example, although orbital bone involvement is rare, it has been previously reported in the literature. Nevertheless, the incidence of this condition has not been studied [10-12]. It mostly occurs in children, as they have a larger marrow volume than do adults [13]. Fever, vision loss, and orbital signs, such as ophthalmoplegia, lid edema, and ptosis, are among the most common presentations. Even though most patients with VOC present with painful episodes, a painless SCD orbitopathy is a very uncommon presentation. However, the mechanism of this painless presentation is not well understood. In 2016, Cameron et al reported the first case of SCD orbitopathy with painless VOC [14]. SCD orbitopathy can result in orbital compression syndrome, eventually leading to permanent vision loss. Thus, timely diagnosis and prompt treatment are essential [3]. Orbital bone infarction provokes an inflammatory response, and if it involves the sphenoid bone, subperiosteal hemorrhage and hematomas are common presentations. Although CT is helpful and readily available in the acute setting, MRI is superior in confirming and diagnosing bone infarctions [15]. In our case, the MRI findings were consistent with those in the literature, as bilateral subperiosteal collections were larger in the left orbit and demonstrated high T1 and T2 signal intensity. Additionally, the findings showed orbital bone wall alteration, suggesting infarction. Moreover, the levels of inflammatory markers, including the erythrocyte sedimentation rate and CRP level, were high. Thus, it is very difficult to differentiate SCD orbitopathy from osteomyelitis and orbital cellulitis, as they share almost the same clinical and radiological findings. Since the management of these conditions differs, establishing a correct diagnosis is essential. SCD orbitopathy dramatically improves with conservative management. Ganesh et al reviewed 24 SCD orbitopathy cases and demonstrated that 20 patients recovered completely with only conservative management, including intravenous fluids and analgesics. Transfusion and exchange were used only in cases that persisted despite treatment [3].
In our case, the ocular symptoms improved slightly with conservative management, but after blood exchange, the symptoms improved dramatically. Surgical evacuation demonstrated an excellent outcome in patients with signs of optic nerve dysfunction and evidence of compressing hematoma, which did not respond to conservative treatment [3].

Conclusions

We report the case of a 4-year-old child with painless sickle cell orbitopathy masquerading as pre-septal cellulitis. Although rare, orbital bone infarction should be considered in every patient with VOC presenting with orbital swelling or pain. The diagnosis can be challenging, as it has similar clinical and radiological features to other inflammatory conditions. However, accurate diagnosis is essential to ensure appropriate management.

Ethics Statement

This study was conducted in accordance with the World Medical Association Declaration of Helsinki.

Declaration of Figures’ Authenticity

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