Frontal Bone Infarctions Masquerading as Bilateral Orbital Cellulitis in a Patient with Sickle Cell Disease

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Abstract:
Repeated vaso-occlusive crises (VOCs) are the hallmark of sickle cell disease (SCD). These repeated crises can lead to bone infarcts, necrosis, and, over time, degenerative changes in the bone marrow. Orbital complications in SCD patients are infrequent and usually present as orbital cellulitis. We report the appearance of orbital bone infarction intraoperatively in the case of an 18-year-old Saudi male patient who has been diagnosed with SCD and presented with severe headaches and generalized body aches. He was admitted with a case of SCD with acute VOC and started on the hospital sickle cell protocol. During the admission, the patient developed bilateral periorbital swelling and left inferior dystopia secondary to bilateral frontoparietal bone infarction, which was evident on the magnetic resonance imaging.

Keywords:
Orbital bone infarction, orbital complications, sickle cell disease, vaso-occlusive crises

Introduction
Orbital bone infarction is still a relatively rare phenomenon in sickle cell disease (SCD) cases, with few examples cited in the literature,[1,2] and it mainly affects children, due to a higher volume of bone marrow spaces in their orbital bones.[3]

Case Report
We report the case study of an 18-year-old Saudi male patient who was known to suffer from SCD. This patient was presented with a generalized body ache and headache, both of which had persisted for 2 days. His headache was most severe in the frontal region and was not relieved by his home-administered analgesics. There was no history of neck stiffness, weakness, or a decreased level of consciousness. There was no history of fever, chills, rigors, or contact with sick people. The patient was admitted to the medical ward as a case of SCD with vaso-occlusive crises (VOCs) and was started on King Fahd Hospital of the University sickle cell protocol; initial workup included complete blood count, retics, renal function test, liver function test, peripheral blood picture, urinalysis, chest X-ray, and electrocardiography. The patient was given half normal saline at a rate of 150 cc/h, and for the pain, he received 5 mg morphine sulfate every 4 h and 8 mg Xefo (lornoxicam) every 12 h. He was also kept on daily pantoprazole 20 mg and folic acid 5 mg. An intradermal skin test was made for ceftriaxone and was negative. One gram of ceftriaxone was administered every 12 h plus vancomycin 1 g every 12 h. He was also kept on daily pantoprazole 20 mg and folic acid 5 mg. An intradermal skin test was made for ceftriaxone and was negative. One gram of ceftriaxone was administered every 12 h plus vancomycin 1 g every 12 h. The patient was evaluated by neurology to rule out meningitis.

This patient was referred to the on-call ophthalmologist as a case of bilateral...
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periocular swelling; the left eye was affected first, followed by the right eye. According to the patient, he developed a similar complaint 2 years ago and was believed to be an acute allergic reaction to an antibiotic.

On examination, the patient was conscious, oriented, and vitally stable, with a low-grade fever. The uncorrected visual acuity was 0.8 in both eyes. The external examination showed swelling of the bilateral upper lid with left inferior dystopia [Figure 1]. In addition, there was no erythema or hyperemia. Extraocular motility was full in both eyes, and color vision assessment by the Ishihara test was normal. The pupils were reactive with no relative afferent pupillary defect.

Furthermore, the slit-lamp examination was unremarkable, and no disc edema or any other abnormality was noted on examining the fundus. The initial impression was orbital infarction. Hence, the ophthalmology plan was imaging to confirm the diagnosis, metronidazole 500 mg intravenous every 8 h to cover anaerobes and Vitamin C supplements.

A magnetic resonance imaging (MRI) with contrast was undertaken and showed intermediate signal intensity on T1-weighted (T1-W) image and high signal intensity on T2-W image. These results occurred with a peripheral diffusion restriction and faint peripheral enhancement, representing multiple bony infarctions and subperiosteal hematomas within the frontoparietal region bilaterally. The MRI also showed a well-defined extraconal lesion, likely to be a hematoma, arising from a superior lateral orbital wall of the left eye, causing mass effect on the ipsilateral lacrimal gland as well as the left lateral rectus muscle [Figure 2].

In addition to the periocular swelling, the patient was found to have a low-grade fever, leukocytosis, and elevated inflammatory markers. Due to ongoing clinical concern, further surgical exploration was conducted via a subbrow incision on the left side, but neither pus nor hematoma was discovered. Interestingly, the bone appeared gray, which may correlate to the bony infarction seen on MRI [Figure 3].

During the follow-up visit, the patient’s pain decreased, and the dystopia improved [Figure 4]. The patient was advised to maintain proper hydration and to take Vitamin C supplements.

Discussion

Orbital bone infarctions are clinically very similar to bone infections (osteomyelitis) in the way in which they present themselves. Periocular pain, fever, or the swelling of the eyelid are all general symptoms that can be seen in patients with early-stage SCD, whether VOC or acute osteomyelitis. Indeed, it is virtually impossible to distinguish between VOC and osteomyelitis when they are in the acute stage, as both conditions cause elevated inflammatory markers such as erythrocyte sedimentation rate, C-reactive protein, and white cell count. However, bone infarction is reportedly 50 times more common in patients with SCD than osteomyelitis.[4,5] The site and extent of bone involvement in osteomyelitis can be achieved using a contrast-enhanced MRI.[6,7] Despite this, there is still uncertainty as to whether such techniques can, in fact, accurately distinguish between acute presentations...
of osteomyelitis and infarctions in children with SCD. Umans et al.\(^6\) and Bonnerot et al.\(^7\) have an opposite opinion on the usefulness of the use of contrast-enhanced MRIs in distinguishing between the two conditions.

In this case study, the patient presented with low grade fever and severe frontal bone tenderness. Other exquisitely tender swelling was not noted. Due to the persistent pain and ineffectiveness of the usual supportive therapy for acute sickle cell crisis, the possibility of infection was suspected.

In this instance, the attempt to drain the suspected hematoma produced neither a hemorrhage nor pus. The uncertainty of interpreting the true nature of the identified subperiosteal fluid in the MRI scan was questionable. The current literature documents five former cases of similar surgical explorations; all demonstrate frank hemorrhagic fluid collection. However, all were severe cases with orbital compression syndrome, and all were diagnosed with computed tomography, with only one exception, was diagnosed by MRI.\(^{[3,8]}\)

Furthermore, having had direct sight of the orbital bone intraoperatively in the case of this specific patient, the bone was discovered to be of an abnormal shade of gray, with a clear delineation between the normal and abnormal bone. It is believed that this is the first time such an *in vivo* observation has been made regarding the appearance of the orbital wall infarction. Such a finding may help us to further understand the orbital pathology surrounding sickle cell bone infarctions.

**Declaration of patient consent**
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**
There are no conflicts of interest.

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