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Case Report
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Orbital Compression Syndrome in Sickle Cell Disease

Riham Salim Alhenaki and Ali Omari*

King Abdulaziz Medical City, Saudi Arabia

Abstract

Background: Cases of orbital compression syndrome are exceptionally reported in sickle cell disease patients. The Orbital involvement in sickle cell disease is rare but it considered one of the rare complications.

Case Presentation: Here we report a 12-year-old Saudi boy known to have sickle cell anemia developed orbital compression syndrome.

Conclusion: OCS should be considered in the differential diagnosis in any with SCD present with proptosis, decreased extraocular motility, eyelid edema, and optic neuropathy, early intervention and recognition can save their vision.

Introduction

Sickle cell disease is a genetic disorder of hemoglobin production characterized clinically by anemia, recurrent painful crisis, splenic and hepatic sequestration, acute chest syndrome, stroke and bacterial sepsis. The highest prevalence of the disease in Saudi Arabia is in the eastern and the southwestern provinces respectively. Orbital involvement in sickle cell disease is rare [1-3]. One of the rare orbital complications of sickle cell disease is orbital compression syndrome which present with acute proptosis, periorbital pain, limited motility, and potentially compressive optic neuropathy.

Case Presentation

A 12-year-old Saudi boy known to have sickle cell Anemia admitted in King Abdullah Specialized Children Hospital in Riyadh due to fever, bilateral eyelids edema and proptosis of the left eye for 10 days prior to admission, there was no history of trauma or insect bite he was admitted initially to a local hospital with vaso-occlusive crisis and managed with IV fluids, antibiotics and pain killers. He had past history of multiple admissions due to vaso-occlusive crisis. He is on folic acid and prophylactic oral penicillin but not compliant.

On initial examination, he looked sick, pale and in pain. His temperature was 38.3°C other vital signs were normal with O_2 saturation at room air 100%. His weight was 34 kg (10^{th} to 15^{th}) centile; height, 143 cm (5^{th} to 10^{th}) centile. He had bilateral eyelid edema and proptosis more in left eye. Ocular motility was restricted in left eye of both elevation and abduction. Pupils were reacting to light bilaterally. There was back pain and restriction of the range of motion of the left hip joint with no other signs of inflammation.

Laboratory results showed a hemoglobin of 6.4 g/dL; mean cell volume 79.2 fl; white cell count 6.71 109/L, with neutrophils 53%; platelets 496 109/L. Blood urea was 2.9 mmol/L, and serum creatinine was 38 mmol/L. Urine examination was normal and culture reported negative. Blood culture also negative. Enhanced CT scan of the orbit shows left orbital extracoronal superior lateral well-defined homogeneous cystic appearing lesion with a peripheral rim of hyperdensity seems arising from the left superolateral orbital wall with mass effect causing proptosis (Figure 1). It measures 2.6 cm × 2.2 cm × 1.0 cm. There is no adjacent fat stranding. The eye globe and extraocular muscles appear intact. Bilateral prominent lacrimal glands are noted. The visualized part of the brain is unremarkable. No osseous destructive lesion. With final impression of Left orbital lesion. Could represent hematoma with orbital compression syndrome. Magnetic Resonance Imaging (MRI) of the orbit was ordered too but was refused by the family.

Patient was managed in Hematology ward with hydration, analgesia, Intravenous (IV) vancomycin and ceftriaxone. He received PRBC packed red blood cells transfusion. Ophthalmology was consulted with impression of mild supraorbital compression not affecting the optic nerve with normal vision decision for supportive care and daily follow-up.

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Correspondence:
Riham Salim Alhenaki, King Abdulaziz Medical City, Al Manar, Riyadh, 11426 SA, Saudi Arabia, Tel: 966558774334; E-mail: reham429@gmail.com

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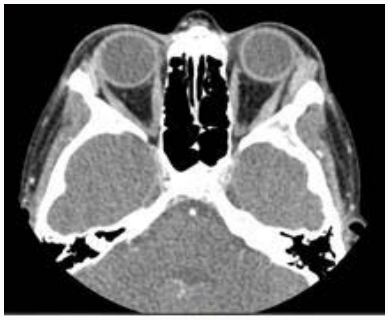

Figure 1: CT scan of the orbits of the eye.

During hospital stay he showed gradual improvement with reduction in swelling and improvement of ocular movement.

Discussion

Sickle cell disease is a hereditary hemoglobinopathy resulting from a single amino acid substitution in the chain of the beta globin molecule. Sickle shaped red blood cells will be formed under hypoxic and acidotic conditions [4,5].

Sickle cell disease can cause vaso-occlusion in any vessel as in vascular bed in the eye which cause ophthalmologic complications in the conjunctiva, anterior segment, retina, choroid, or optic nerve with vision-threatening consequences [6], however, orbital involvement is a rare finding [7]. Orbital Compression Syndrome (OCS) is an acute condition characterized by proptosis, eyelid edema, periorbital pain, restriction of extra-ocular motility, with or without decreased visual acuity [5,8].

Because of increase marrow space in the orbital bone in the youth more than that in adult, orbital wall infarction occurs in children [4]. In a report of 36 cases of OCS the mean age was 13 years and the youngest age was 2 years [9]. The presentations of pain and eyelid edema occur in all cases, bilateral proptosis, chemosis, limited ocular motility, and vision loss occur in most severe cases [5]. Fever at presentation occur in almost all reported cases, and with association with pain crises in more than two third of cases [8]. One third of cases reported with bilateral orbital involvement [8]. The differential diagnosis of a sickle cell patient with acute periorbital pain and swelling includes OCS, orbital cellulitis and abscess, osteomyelitis, orbital tumor, hemorrhage, and infarction [8]. The presence of leukocytosis and fever are nonspecific [10]. MRI is the imaging modality of choice to confirm the diagnosis of OCS in most of the reported cases [10]. In our case urgent CT done in ER shows findings consistent with OCS diagnosis. Most cases of OCS managed with conservative treatment [5,8,11]. Orbital pressure caused by inflammatory component of orbital bone infarction can be relieved by administration of intravenous corticosteroids. Also, antibiotic coverage is advisable as it is often difficult to clinically differentiate osteomyelitis from bone infarction [4,5]. If there are signs of optic nerve dysfunction or large hematomas surgical exploration and evacuation of hematoma is warranted to prevent vision loss and to speed recovery [4,11]. Among the reported cases, only two cases end with permanent visual loss.

Conclusion

Children with SCD present with proptosis, decreased extraocular motility, eyelid edema, and optic neuropathy, OCS should be considered in the differential diagnosis. Empirical use of broad-spectrum antibiotics is advised since it is difficult initially to differentiate OCS from other infectious causes.

Early consultation to ophthalmology with early evaluation and close follow up is mandatory, surgical intervention is warranted if optic nerve dysfunction or large hematoma is present to save the patient's vision.

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