these ampoules cannot have a negative systemic affect. However, systemic toxicity is rare with regional anesthesia.[3] In addition, the assessment of endocrine stress response in anesthesia and surgery is controversial.[4] Removal of a larger tumor from the neck requires drainage. Following the surgery, the penrose drain was used and as a precaution it was not removed for 2 days. Closure with 5,0 Nylon gave us an opportunity for a cosmetic scar. The whole operative procedure lasted 1 hour. The patient's optimism and recovery in the post-operative period were on a high level. Certainly, the main reasons could be short medical procedure and home environment during the recovery period. For shorter procedures, LA offers the advantages of convenience and comfort to the patient.[2]

Neck surgery under LA in an outpatient environment had a lot advantages for the patient in this case. It saved his time. The patient was conscious during the surgery. He avoided possible side effects of the general anesthesia such as nausea, vomiting, muscle aches, and generally longer medical procedure. The same-day surgery gave him an opportunity for smooth postoperative course, shorter medical procedure, and shorter recovery time. Lastly, the whole procedure was much less expensive.

The scope of outpatient surgical care for head and neck surgery will undoubtedly increase.[5] Neck surgery under LA with outpatient conditions also shows great potential in adequately selected patients. Understanding of neck *in vivo* anatomy, appropriate application of LA and experience in emergency surgical care are presumably the main requirements that are placed in front of the surgeon.

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Bilateral Subperiosteal Hematoma and Orbital Compression Syndrome in Sickle Cell Disease

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Abstract: A 14-year-old boy with sickle cell disease presented with preseptal cellulitis findings as proptosis, eyelid edema, and hyperemia. His best corrected visual acuity in the right eye was 20/20 and 16/20 in the left eye. He had limited ductions in vertical and lateral gazes in both eyes. Bilateral venous tortuosity was observed in posterior segment examination. Orbital bone infarction and subperiosteal hematoma were seen in magnetic resonance imaging. He was diagnosed as having orbital compression syndrome secondary to vaso-occlusive crisis of sickle cell disease and was treated with intravenous ampicilin-sulbactam and methylprednisolone.

Key Words: Orbital compression syndrome, sickle cell disease, subperiosteal hematoma

Sickle cell disease (SCD) is an autosomal recessive disorder and is associated with high morbidity and mortality. It is known that people who are carriers generally do not have any medical problems and SCD is traditionally considered a benign condition by ophthalmologists.[1,2] Several studies have reported ocular complications in SCD, but these complications have been described as the consequence of trauma, exertion, and associated systemic disorders.[3] Ocular manifestations of SCD may include anterior segment ischemia, secondary glaucoma, angioid streaks, retinopathy, and retinal artery occlusions.[4] In addition, patients with SCD are at risk for orbital compression syndrome (OCS) secondary to orbital bone infarction, in the setting of vaso-occlusive crises.[5] In the present study, we aimed to report a patient with OCS who was diagnosed as having SCD.

CLINICAL REPORT

A 14-year-old boy patient who was hospitalized in Pediatric Clinic because of avascular necrosis in his left femur head was referred to Ophthalmology Department with bilateral eyelid hyperemia and edema, proptosis, and low vision in his left eye. He had no history of trauma or insect bite and these symptoms occurred in the fourth day. On ophthalmological examination, he had severe bilateral eyelid edema and hyperemia. He had limited ductions in vertical and lateral gazes in both eyes (Fig. 1). Best corrected visual acuity (BCVA) in his right eye was 20/20 and 16/20 in his left eye. Pupil reactions and anterior segment examination were normal in both eyes. Bilateral venous tortuosity was observed in posterior segment examination. Coagulation parameters were within normal limits, but his hemoglobin and platelet levels were low. Serum fibrinogen level was 618.5 mg/dL (175–400 mg/dl). One unit of erythrocyte and fresh frozen plasma transfusion was made. Magnetic resonance imaging of orbits showed calvarial infarction including the frontal and parietal bones and bilateral subperiosteal hematoma in the orbital walls (Fig. 2). His transcranial doppler ultrasonography was normal. He was diagnosed as having OCS secondary to vaso-occlusive crisis of SCD. Patient was treated with intravenous ampicilin-sulbactam and methylprednisolone (1 mg/kg). Eyelid edema was resolved at the sixth day and it was totally recovered after 13 days (Fig. 3). BCVA was 20/20 in both eyes at the 13th day.

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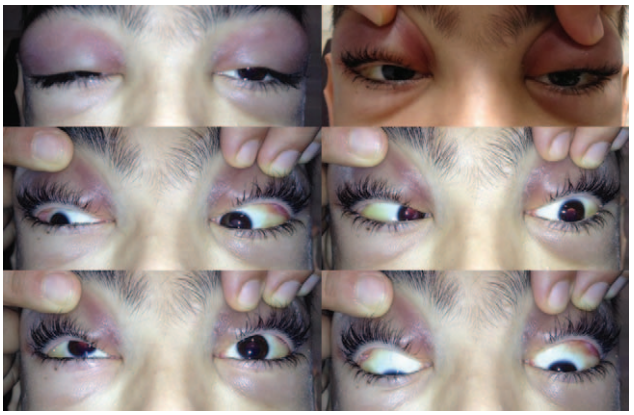

FIGURE 1. Limited ductions in vertical and lateral gazes in both eyes before the treatment.

DISCUSSION

SCD is an inherited hemoglobinopathy which results from a point mutation of beta globulin. An abnormal hemoglobin S (HbS) molecule is formed and this form tends to polymerize under conditions of hypoxia and acidosis and HbS may block capillaries (vaso-occlusion crises) which explains almost all clinical manifestations of SCD.[1,6] Bone infarction is common in SCD; however, involvement of the orbit is rare. Orbital wall infarction typically occurs in young patients because there is more marrow space in the orbital bone in children than in adults.[7] In addition, it is more common in males than females. Our patient was a 14-year-old boy.

Infarction of orbital bones during vaso-occlusive crises in SCD presents acutely with a rapidly progressive periorbital swelling.[5] Hematomas frequently complicate the condition and, along with the inflammatory swelling, may lead to OCS. The main mechanism for development of OCS is orbital bone infarction with subsequent inflammatory response that can rapidly spread to the orbit resulting in orbital pain and proptosis and other features of OSC.[7] Most of the cases were bilateral just as our case.[8,9] Fever is seen mostly and swelling of the eye lids is one of the most common finding. The condition is therefore sight-threatening, and necessitates prompt diagnosis and appropriate management for resolution without adverse sequel.[4]

This condition needs to be differentiated from orbital cellulitis, pseudotumor or any other neoplastic disorders of the orbit. The presence of leukocytosis, elevated erythrocyte sedimentation rate and C-reactive protein can occur in both bone infection and infarction.[10] Imaging techniques are frequently used in the evaluation of patients, both for demonstrating the bone infarction and excluding other orbital diseases causing proptosis. In OCSs subperiosteal hematomas are common and appear to result from bone marrow infarctions.

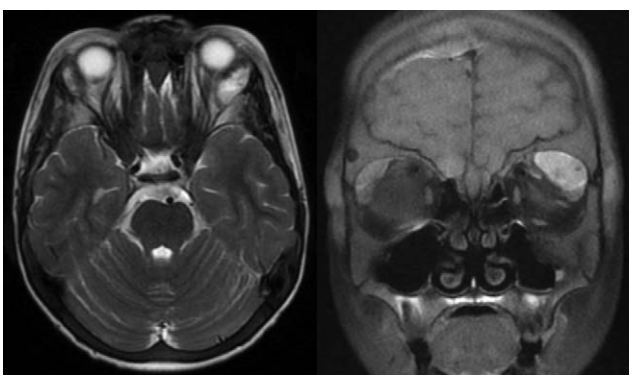

FIGURE 2. Bilateral subperiosteal hematoma in the orbital walls.

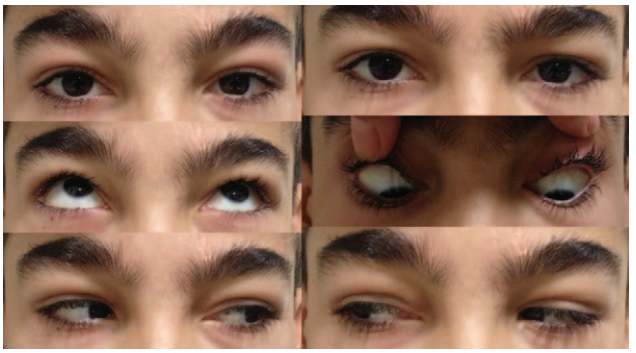

FIGURE 3. Ocular findings were totally recovered after 13 days.

The majority of OCS cases resolve with conservative treatment that includes steps to combat the vaso-occlusive crisis and use of systemic steroids under wide-spectrum antibiotics cover.[5] Supportive care is effective, unless optic nerve dysfunction or large hematomas is present, which would indicate that surgical evacuation is warranted to prevent loss of vision and to speed recovery. Our patient's response to conservative treatment was quite well and he had no need to surgical treatment.

In conclusion, OCS is a sight-threatening condition that occurs in the patients with SCD. One should be careful when patients with SCD present with proptosis, restricted extraocular motility, eyelid edema, and optic neuropathy. Children with SCD are susceptible to infections, and empirical use of broad-spectrum antibiotics should be considered if infectious process is suspected. An ophthalmologist should always be consulted, as early evaluation and surgical intervention if evidence of optic nerve dysfunction or large hematoma is present can be vision saving.

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