CASE REPORT

Bilateral orbital infarction and retinal detachment in a previously undiagnosed sickle cell hemoglobinopathy African child

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ABSTRACT

Bone infarction involving the orbit in sickle cell disease is not common. Bilateral orbital infarction in a previously undiagnosed sickle cell hemoglobinopathy has not been previously reported. In this report, we present a case of an 11-year-old previously undiagnosed sickle cell disease Nigerian girl with severe acute bilateral orbital infarction and retinal detachment to highlight that hemoglobinopathy induced orbital infarction should be considered in African children with acute onset proptosis with or without previous history of sickle cell hemoglobinopathy.

Key words: Hemoglobinopathy, Nigeria, orbital infarction, proptosis, retinal detachment

INTRODUCTION

Retinal changes and long-bone infarction are common features in sickle cell disease, orbital involvement is however, not common.[1-3] Vaso-occlusive process cause bone infarction in the orbit with subsequent acute rapidly progressive periorbital swelling, fever, marked proptosis, and subperiosteal collection which can closely mimic infectious orbital cellulitis.[3-6] Prompt diagnosis and appropriate management is required to achieve resolution without adverse sequela following this sight threatening complication.[7] Conservative management is the treatment of choice with occasional surgical intervention.[4,5,8,9] Orbital disease without prior history of vaso-occlusive crisis has been reported in a patient with beta thalassemia.[9] Authors are not aware of previous report of bilateral orbital infarction without prior vaso-occlusive crisis or the concomitant presence of retinal detachment.

Previously, undiagnosed sickle cell hemoglobinopathy (HbSS) Nigerian child presenting with severe bilateral orbital infarction and retinal detachment is presented.

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An 11-year-old girl presented with 4 days history of fever, headache, generalized body weakness, and 1 day history of bilateral painful proptosis associated with sudden loss of vision and purulent discharge. She had no nasal discharge, blockage, dental pain or dental procedure. There was no history of trauma, previous bone pains or frequent illness. Her genotype was not known; she is the fourth of seven children and a 3-year-old sibling died following fever and severe anemia. She had lived all her life in a rural community with both parents who are farmers and genotypes unknown.

She was conscious but lethargic, pale, afebrile, mildly dehydrated with bilateral sub-mandibular, and sub-mental tender lymph node enlargement. She had tender hepatomegaly and splenomegaly. Visual acuity was no light perception in both eyes with marked tender periorbital swelling, axial, non-pulsatile proptosis of 23.5 mm and 23 mm on the right and left respectively [Figure 1]. There were no extraocular muscle movements and inferior prolapsing chemosis; the right corneal was hazy with turbid anterior chamber, round and unreactive pupil, clear lens, and a retrolental greyish reflex. The cornea was clear on the left, with clear anterior chamber, small, unreactive pupil, clear lens, and a retrolental glow. Fundal view was precluded by the retrolental reflex in both eyes. Intraocular pressure was 16 mm Hg in each eye. Initial diagnosis of bilateral orbital cellulitis to rule out cavernous sinus thrombosis was made; she was investigated and commenced on broad spectrum intravenous and topical antibiotics empirically as in-patient. Ocular ultrasound

of bilateral proptosis, bullous retinal detachment, and subretinal fluid collection with no orbital collection or mass[Figure 2], Cranial computed tomographyscan exclusion of space occupying lesion, orbital collections or intracranial extension [Figure 3] as well as Hemoglobin genotype result of HbSS lead to diagnosis of bilateral severe orbital infarction in a sickle cell disease. Management with systemic/topical steroid, intravenous fluids, analgesics as well as cycloplegics produced sustained progressive reduction in proptosis, chemosis and improved general health although, her vision remained no perception of light in both eyes. She was discharged on oral medications and tapering steroid dose after 2 weeks to continue out-patient management. Patients have since, defaulted preventing repeat ocular scan and follow-up evaluation.

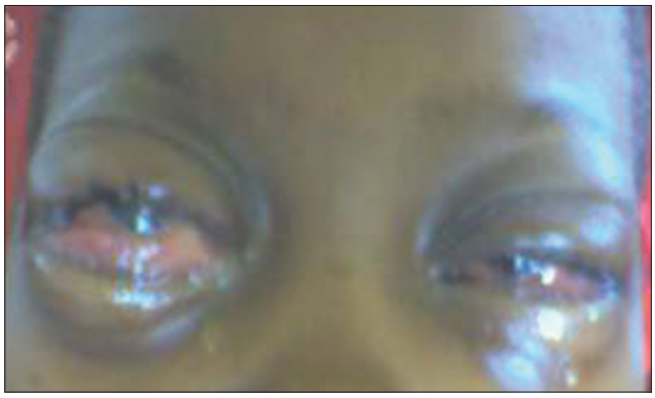

Figure 1: Severe bilateral orbital congestion with proptosis and chemosis

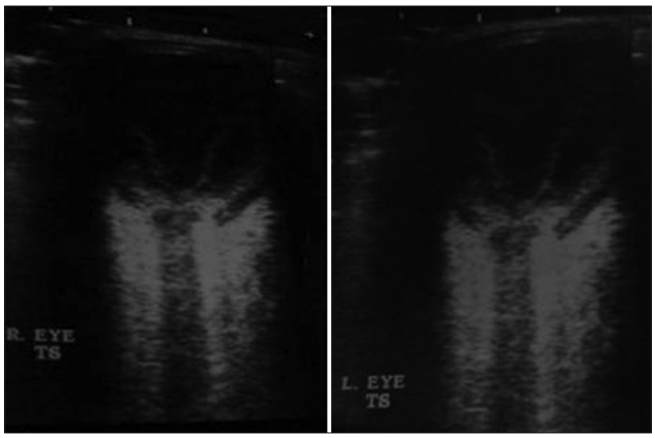

Figure 2: Ocular ultrasound scan

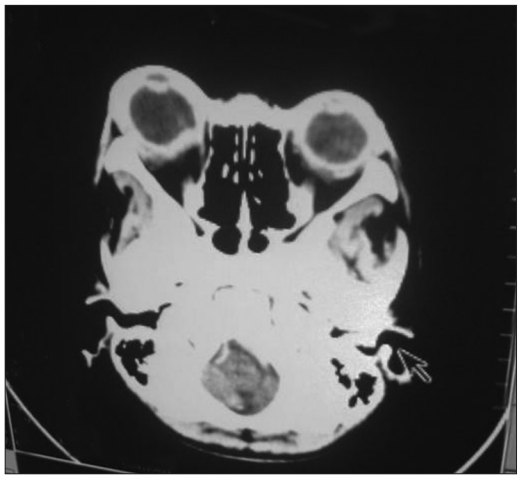

Figure 3: Cranio-orbital computed tomography scan

DISCUSSION

Sickle cell hemoglobinopathy is associated with significant morbidity in sub Saharan African. Orbital infarction, a potentially vision threatening disease of acute onset is known to closely mimic infectious orbital cellulitis;[3,4,10] previous knowledge of hemoglobin genotype aids prompt clinical diagnosis while magnetic resonance imaging (MRI) and technetium assist in making definitive diagnosis.[10] Orbital infarction requires steroid therapy and rehydration to aid resolution in addition to prophylactic antibiotics use. Delay in diagnosis would have been avoided in the index case were the genotype known or from previous history of bone pain crisis. Minimal body pains and mild dehydration level in comparison with the severity of orbital involvement was noted in this patient; Ganesh *et al*. reported the degree of severity of the orbital manifestations appeared unrelated to the severity of sickle cell disease.[10] Non-availability of MRI facility or nuclear scintigraphy studies prevented definitive documentation of bone density changes in our patient.[3,10]

Bilateral orbital infarction in a previously undiagnosed HbSS patient is not common. Retinal detachment is not a common finding in sickle cell induced orbital bone infarction; severe acute infarction with attending orbito-ocular inflammation is a probable cause of the exudative retinal detachment. This probable link requires further studies.

Hemoglobinopathy induced orbital infarction should be considered in African children with acute proptosis with or without previous history of vaso-occlusive crisis. Routine screening for hemoglobinopathy may improve the management of patients.

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