Case report

Multiple Scalp Infarctions and Bilateral Orbital Compression in A Child With Sickle Cell Disease

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ABSTRACT:
Orbital compression syndromeis one of Orbital complications in sickle cell disease (SCD) in which a vaso-occlusive process affecting the marrow space of the orbital bone results in subperiosteal hematomas as a result of bone marrow infarctions. It is one of the unusual manifestations of SCD. Usually presents as frontal headache, fever and eyelid oedema. Here we report an 8 year-old Sudanese girl known to have SCD since the age of one year based on HB electrophoresis, presented with bilateral orbital compression syndrome and multiple scalp infarctions, treated conservatively but at the end she undergoes operative orbital decompression when she start to developsings of optic nerve dysfunction and rapidly increasing hematomas to prevent loss of vision and to speed recovery, The ophthalmologist found that the orbital hematomas get secondary infected swab for culture and sensitivity taken and she received Gentamycin for two weeks and fortunately her vision was preserved.

Key words: Sickle cell diseases, orbital compression syndrome, scalp infarction.

Retinopathy, retinal artery occlusions and anterior segment ischemia all are Ocular manifestations of sickle cell disease[1] but infarction of the orbital bones has infrequently beenreported,which presents as acute proptosis,ocular pain, ocular motility affection and compression of the optic nerve, resulting in orbital compression syndrome[1].

To best of our knowledge, 36 cases of OCS have been reported[2- 8]. Most of the cases, 83% occur in homozygous haemoglobin S.Orbital wall infarction typically occurs in youths because there is more marrow space in the orbital bone in children than there is in adults[9]. The mean age of presentation in the36 reported cases was 13 years, with the youngest reported age at 2 years[6]. Mostcases of OCS resolve with conservative treatment[2,10,11]. Among the reported cases, only one case ended with permanent visual loss in the patient left eye. This patient had severe bilateral disease and was managed conservatively[2].

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CASE REPORT:

We report here an eight years oldSudanese girl known to have sickle cell disease (HbSS) since the age of one year based on HB electrophoresis. She presented with fever, cough, pain and swelling of her ankles for 3 days. The pain was severe and progressive to the degree that made her unable to walk or do daily living activities. She had history of repeated admissions with painful crisis.

On examination she was ill, febrile temp 40 °C, very pale, in pain, her weight and height both below 3rd percentile for her age and sex. She had 4 cm tender hepatomegaly and 6 cm spleen. Chest examination revealed signs of right upper lobar consolidation in form of bronchial breathing and crackles. Her O_2 saturation was 96% on room air.

Her investigations showed: HB 4mg/dL, TWBCS $20x10^3$cell/mm^3, Platelets count $386x10^3$cell/mm^3, Reticulocytes count 10%, blood film for malaria –ve, urine analysis was clear, chest X ray showed upper right lobar consolidation, renal and liver function tests were normal.

She commenced onoxygen, blood transfusion, I.V. ceftriaxone, Ibuprofen and I.V. hydration, she became afebrile and her ankles swellings completely resolved. After 10 days of hospital stay she showed marked improvement and was planned for discharge but suddenly she develop headache, bilateral proptosis (Figure 1) and multiple boggy tender scalp swellings.

MRI brain and orbits was done for her andrevealed bilateralorbital hematomas and multiple small pocket collections in the scalp (small hematomas) (Figure 2&3).

Bone scan with angiography confirm multiple scalp infarction including the orbital bone (figure 4)

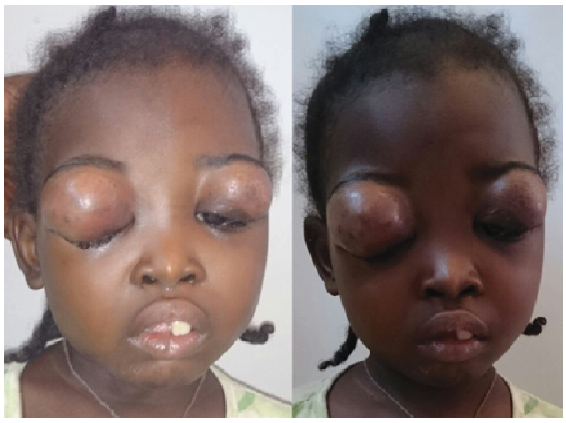

Figure 1: Acute bilateral proptosis

Ophthalmologist was consulted and confirmed the diagnosis of orbital compression syndrome and advice to continue on conservative management (I.V.hydration and analgesia) with daily follow up to the eye movement and pupillary reactions because her fundal examination was normal at that time and there was no compression on the opticnerve.

She was doing well on conservative management, but after 7days the eye swelling acutely increases along with the scalp swellings, she developed painful eye movement specially the upward gaze, and she develops sluggish pupillary reaction and she became febrile again. Operative orbital decompression and evacuation of the hematomas were done for her but the ophthalmologist found that the hematomas got secondary infected and he took sample for culture and sensitivity which revealed klebsiella pneumonae sensitive to Gentamycin.

The patient receive Gentamycin I.V. for 2 weeks with regular ophthalmology follow-up and she responded very well and discharge with her vision preserved (Figure 5).

DISCUSSION:

Sickle cell disease (SCD) is an autosomal recessive genetic disorders resulting from the presence of an abnormal mutated form of haemoglobin (haemoglobin S)[12], the cells contains this abnormal haemoglobin become sickle shaped and rigid when exposed to some conditions like hypoxia and acidosis. These cells have short lifespan that leads to chronic haemolysis. Due to their rigidity and abnormal shape these cells tend to block small vessels leading to vaso-occlusion which explains most of the clinical manifestations of sickle cell disease.

Ophthalmologic complications can result from vaso-occlusion in any part the eye, which can affect the conjunctiva, retina, choroid, or even the optic nerve which

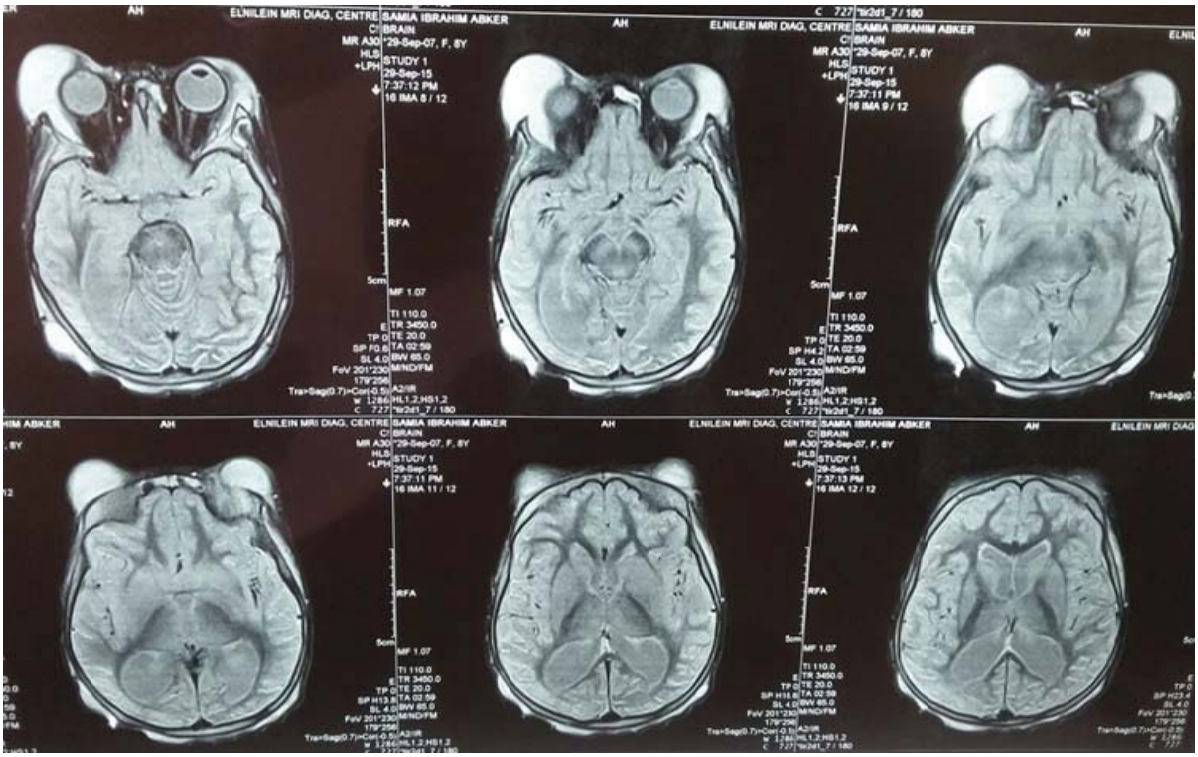

Figure2: Brain MRI displayed: Axial T1W1 and coronal T1W1 reveals well defined mass lesions with iso-intense signals intensity and include hyper-intense areas .axial and coronal T2W1 and FLAIR shows the mass lesions with high signals representing eyelids oedema with pocket collection of portentous material (sub-acute hematoma). The mass seen arise from the roof of the orbital wall extending laterally intra-orbital affecting both orbits, pushing ,displacing and compressing the eye globes medially and anteriorly causing proptosis (RT 3.5x2.3 cm)(LT 2.3x1.5 cm).

can be vision-threatening[13], but orbital involvement is a rare presentation of SCD[14].

Orbital compression syndrome (OCS) is an acute condition characterized byeyelid oedema, proptosis, ocular painand affection ofextra-ocular motility, sometimes with decreased visual acuity secondary to optic nerve compression[2,10]. Its presentation ranges from just pain and eyelid oedema, to the most severe form that presents with bilateral proptosis, painful limitation of ocular motility, and sometimes even acute loss of vision[10]. Our patient had acutely developed the severe symptoms and signs of the severe form of OCS except that her vision was not affected at the start.

The mechanism behind the development of OCS is formation of hematomas which may be orbital (subperiosteal) or intracranial (epidural) as a consequences of orbital bone infarction with inflammatory response that results in orbital pain and proptosis and other features of OSC[9].

Our patient had bilateral orbital hematomas and orbital bones infarctions as well as multiple infarctions of the bone of her skull.

Similar to our patient, more than two thirds of reported cases presents with pain crises elsewhere[2]. Bilateral orbital compression syndrome is reported only in one third of all reported cases of OCS[2]. There is really a challenge to differentiate orbital

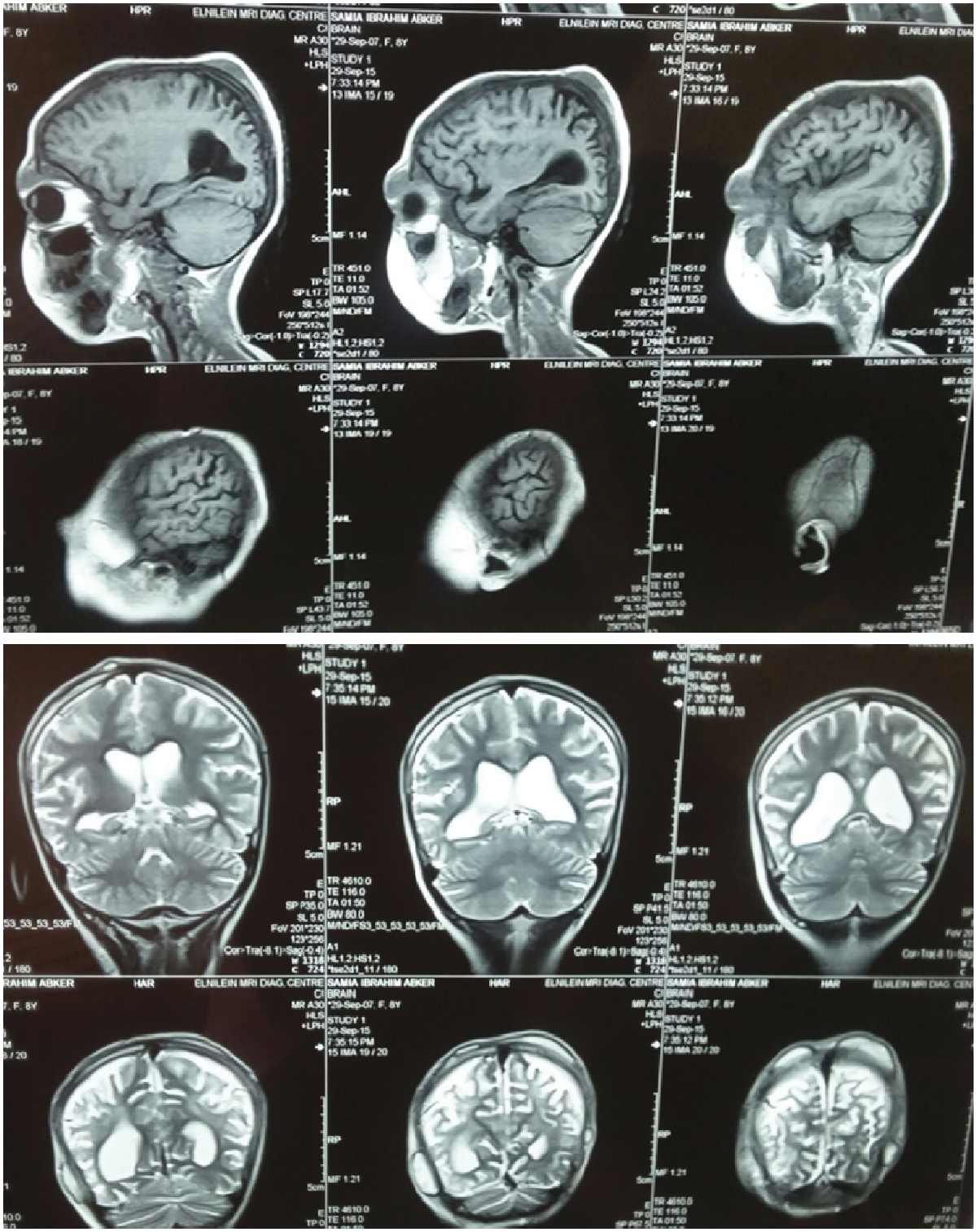

Figure 3: Brain MRI displayed: Multiple small pocket collection noted in different sites in the scalp prominent in the temporal and parietal areas.

compression syndrome and orbital cellulitis and also for orbital bone involvement whether it is bone infarction or infection. This differentiation is crucial and necessary because the management will be totally different.

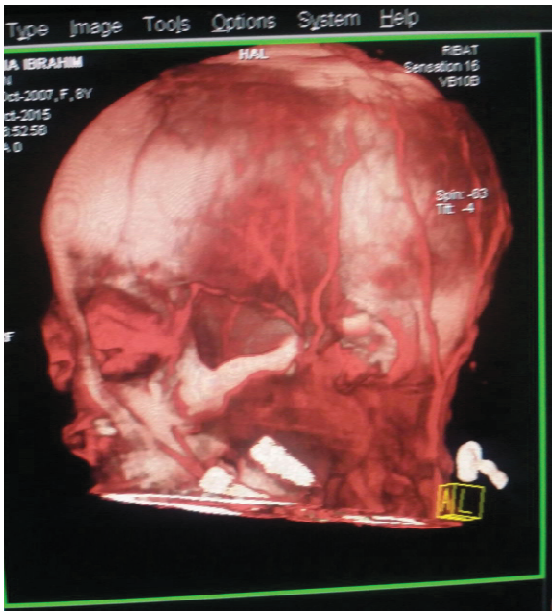

Figure 4: Reconstructed CT bony skull and angiogram confirms bone infarction.

Marrow infarction can be confirmed by bone scan in 95% of cases by demonstrating decreased tracer uptake, in caseof infection the uptake is normal[10]. In our patient the bone scan confirmed the presence of multiple skull bone infarction involving the orbits. The imaging modality of choice for evaluation of OCS is MRI in most of the reported cases, as it clearly identifies hematomas and their compression effect as in our patient.

OCS isusually treated by conservative management with critical observation for signs of optic nerve dysfunctions. Sometimes steroids are used to decrease the inflammation around the orbit. Surgical exploration and evacuation of hematoma is indicated if there are signs of optic nerve dysfunction or rapidly enlarging hematomas to prevent optic nerve compression and vision loss and also to speed recovery[9,15]. In case of our patient we used both modalities of treatment when appropriate.

Figure 5: Resolved orbital compression syndrome after surgical decompression.

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