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Non-Traumatic Epidural Hematoma In Sickle Cell Disease And The Role Of Neurosurgical Intervention: A Case Report

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Summary

A 9-year-old male known to have Sickle-Cell Disease, presented with a headache, projectile vomiting, blurry vision, and one-sided weakness for seven days, without any history of trauma or assault prior. CT scan showed left temporal parietal epidural hematoma with significant mass effect, ventricles effacement and midline shift. Diagnosed with spontaneous epidural hematoma and underwent emergency craniotomy for hematoma evacuation. This case report discusses a rare case of spontaneous epidural hematoma in a child less than 10 years of age and role of early surgical intervention for better clinical outcomes.

Keywords: Sickle cell, non-traumatic, epidural hematoma, neurosurgical intervention, Tanzania

INTRODUCTION

Sickle cell disease (SCD) is a genetic haemoglobinopathy inherited as an autosomal recessive trait gene. This disease results from a substitution of glutamic acid to valine at position 6 of the beta-globin polypeptide chain. These changes lead to the polymerisation of affected haemoglobins also termed Haemoglobin S in the states of deoxygenation that leads to the destruction of the red blood cells (RBC) membranes and results in sickle-shaped cells which results in a typical presentation of SCD such as Haemolysis, vaso-occlusive crises, neurovascular complications. [1] [2]

Neurological complications are among the life-threatening conditions in SCD. The prevalence of intracranial haemorrhages is 11% in SCD patients before the age of 20 years old. [1] Children with SCD often present with Subarachnoid haemorrhage without any identifiable aneurysm. However, to the best of our knowledge, currently, there have been a total of 32 cases of Epidural hematoma in Sickle Cell disease reported globally, 7 out of those died in the course of treatment.[2]The pathophysiology of non-traumatic epidural hematoma in sickle cell disease is still unclear. [3]We

present a case of a 9-year-old boy with a history of severe headache and right lower limb weakness who attended Muhimbili Orthopaedic Institute in Dar Es Salaam, Tanzania.

CASE PRESENTATION

This was a case of a 9-year-old male, known SCD patient since he was 6 years of age, not attending clinic, on folic acid 5mg once daily and had a steady state Haemoglobin (Hb) levels of 6-7g/dL for the past three years. The patient presented with a seven-day history of headaches of acute onset progressive, projectile vomiting of previously eaten meal mixed yellow strained, blurry vision associated with photophobia, right-sided weakness more on the right lower limb which was of acute onset and non-progressive with pre-existing episodes of chronic joint pain with no history convulsion, loss of consciousness, or fevers.

The patient had no history of trauma or history of similar presentation prior. The patient also had a history of blood transfusion 4 years prior.

Before admission, the patient attended a regional referral hospital where he received blood transfusion before being referred to Muhimbili Orthopaedics Institute. Upon arrival at the hospital, the patient was fully conscious with a Glasgow Coma Score of 15 E4V5M6, (spontaneous eye opening, oriented to people time and place and was able to obey motor commands) and had anisocoria dilated right eye 5mm, and left eye 3mm, pale and with frontal bossing, afebrile with a temperature of 37^{o}C and with lower limb power of 4/5 in all muscle groups. The admission haemoglobin level was low - 7.4g/dL (reference range 12-16 g/dL) and platelets were within the reference range at 120g/dL (reference range 100-300). After appropriate initial evaluations at the clinic/emergency department investigations mentioned below were ordered.

Informed written and verbal consent for treatment and case report write-ups were obtained.

INVESTIGATIONS

CT scan of the brain (Figure 1) was done which revealed left temporal parietal epidural hematoma with significant mass effect, effacement of basal cisterns and ipsilateral ventricles, midline shift of 5.4 cm to the left and unremarkable bone window

TREATMENT

The patient was evaluated and taken to the operating theatre for emergency craniotomy and epidural hematoma evacuation. Intraoperative findings were 150mls of epidural blood clots, his temporal bone was returned and a subgaleal drain was placed. The patient was transferred to the Intensive Care Unit (ICU) where he stayed for 1 day where he received Intravenous Ceftriaxone 500mg once daily, Morphine 5 mg t8 hourly, Normal saline 1250mls for 24 hours., Paracetamol 250mg 8 hourly. After a trend of stable vitals, the patient was transferred to the general ward. In the ward, he received a total of 450 mls of packed red blood cells of 150 mls per unit of transfusion, observed for 6 days then discharged. On the day of discharge, he was conscious, with the ability to walk well without support, resolved blurry vision, no headache, and no history of vomiting post-operation or fever.

OUTCOME AND FOLLOW-UP

The patient returned to the clinic for follow-up after 3 weeks, the patient reported no recurrence of prior symptoms. Upon examination, he was alert, afebrile, mildly pale, pupils bilateral equal reactive to light, not jaundiced, Healed scalp wound, and Muscle power of 5/5 in all muscle groups. CT scan (Figure 2b) showed a completely

resolved epidural hematoma in the brain window and a bone window showing no bone pathology.

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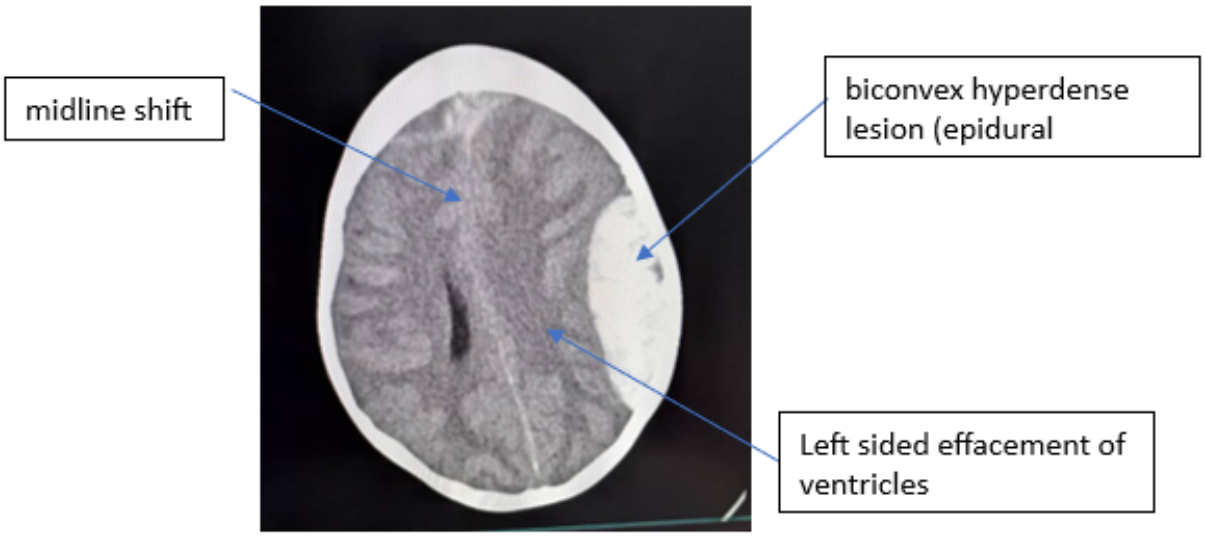

Figure 1; CT Scan brain window showing left temporal parietal epidural hematoma with significant mass effect, effacement of basal cisterns and ipsilateral ventricles, midline shift of 5.4 cm to the left.

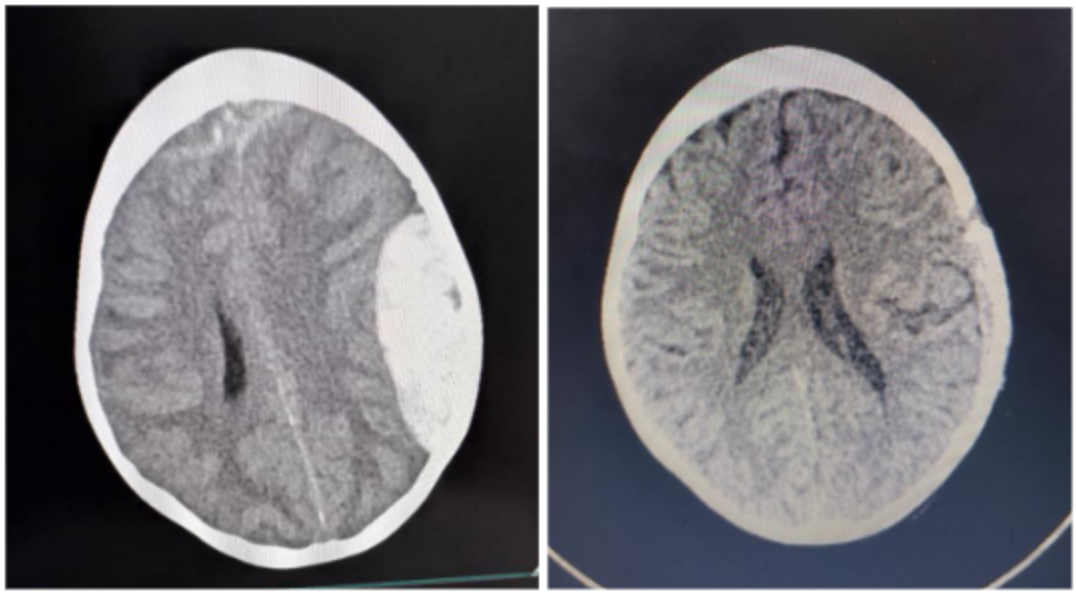

Figure 2 (a) Pre-op CT scan showing massive temporal parietal hematoma epidural hematoma with midline shift (b) control CT 3 weeks post operation showing resolved epidural hematoma

DISCUSSION

The case presented illustrates an example of CT scan and intra-operative proved epidural hematoma in a patient with sickle cell disease in Tanzania. While the incidence of epidural hematoma among patients with SCD in Africa, when missed it can lead to serious immediate and long-term complications including death. Africa carries a burden of more than 75% of all sickle cell disease cases globally [4] . Tanzania is one of the leading countries with high SCD prevalence in Africa and globally, despite having this huge number of patients there is only one case of epidural hematoma in SCD that has been published, to the best of our knowledge. [5]

Epidural hematoma is a collection of blood in the extradural space and it is almost exclusively occurs in trauma, however there are some few cases of non-traumatic spontaneous epidural hematoma that has been reported in patients with sickle cell disease and also end stage renal disease. [6] Even though uncommon, epidural hematoma in paediatric setting can present following other domestic incidences such as falling accidents, sexual violence and domestic abuse. It is important to rule out other causes through thorough clinical examination by looking into bruises and other signs pointing towards other traumatic causes. [5] In our case, thorough physical examination was done and ruled out possibilities of having a traumatic cause.

Pathophysiology of non-traumatic epidural hematoma in SCD is quite unclear but most cases reported involve other pathologies such as vasculopathy, cancer and infections [A1]. Despite their difference in etiology, epidural hematomas largely share a common clinical presentation. [5] The most common presentation being headache, anisocoria, one sided weakness, seizures, loss of consciousness and vomiting. Glasgow Coma score of less than 8 is highly associated with poor outcomes [8]

SCD starts to be clinically evident at an age of 6 months due to decline of fetal hemoglobin that masks the hypoxic stress of red blood cells. Patients tend to present with dactylitis, regressed or delayed milestones, later with frontal bossing. Our patient started to present with dactylitis at 4 months of age and also had delayed gross motor milestones; he started walking at 18 months of age. Clinical presentations among pediatric patients are Vaso-occlusive crises, frontal bossing and frequent blood transfusion. Our patient had a history of multiple hospital admissions due to Vaso-occlusive crises and also history of multiple blood transfusions and was not attending SCD clinic or initiated on Hydroxyurea therapy. Hydroxyurea use among SCD patients is associated with decrease in SCD complications and number of hospitalizations [8]

CT scan is the most common imaging modalities for cases of acute epidural hematoma for both traumatic and non-traumatic epidural hematoma, and also has a high sensitivity to these lesions. However, in contrast to other few cases we lack presence of pre-incident imaging that could have ruled out pre-existence of other pathologies prior[5]. Early neurosurgical intervention for operable epidural hematoma offers good outcome, with an average ICU stay of 4 days and an average hospital stay of 11 days [8] Our patient had an ICU stay of 1 day and a total of 6 days hospital stay which is a better outcome compared to the average stay of 11 days [8] [4]

We have presented a paediatric case of non-traumatic epidural hematoma in a patient with SCD, a rare case of epidural hematoma and neurovascular complications of SCD, number of cases being reported since the past decade has been increasing, this might be attributed due to the actual increase in these complications or advancement in radiologic technology,

health service availability, and publication opportunities [2]

In all cases that have been published the emphasis has been on clinical presentation and medical management outcomes, in our case we are highlighting the role of early neurosurgical intervention in improvement of overall clinical outcome [4.

CONCLUSION

Diagnosis and treatment of Epidural hematoma in SCD patients is still challenging. This is contributed with rarity of these cases and scarcity of proper diagnostic tools in most Africa country regions where SCD has higher prevalence. Regardless, it is important to be aware of epidural hematoma in patients with SCD. A combination of high clinical suspicion, early radiological tests and on time neurosurgical intervention improves clinical outcomes to Sickle cell patients presenting with epidural hematoma.

Competing interests None

Patient consent: Written consent Obtained.

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