

## CASE REPORT

## A case of spontaneous epidural hematoma in sickle cell disease patient

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### Introduction:

Sickle cell anaemia, an autosomal recessive disease relatively common among the black races, gives rise sometimes to neurological complications.<sup>2</sup> It is a qualitative hemoglobinopathy due to the presence of hemoglobin S. Its incidence is high in Africa and in African American populations. The average life expectancy is 42 years for men with sickle cell disease and 48 years for women.<sup>7</sup> The term "sickle cell crisis" is used to describe several acute conditions, including vaso-occlusion, aplastic crisis, splenic sequestration and hemolysis.<sup>7</sup> Most crises last between five and seven days and can be managed with fluids and analgesia. Neurologic complications of sickle cell disease are rarely described in the medical literature, and their mechanisms are not completely elucidated.

**Key words:** sickle cell crisis, epidural hematoma, sickle cell disease, craniotomy

### Case Report:

A 15 year old female patient known to have SCD since childhood with frequent admissions and blood transfusions. She is still not menstruating. Past history of Splenectomy and 2 previous ischemic strokes. The patient was maintained on: Aspirin, Hydroxyurea, Folic Acid.

She presented to ED of king Abdullah Hospital Bisha in southern region in kingdom of Saudi Arabia on 23.6.1436 with generalized body-aches, backpain and leg pains. There was no fever, or headache. On Physical Exam: T 36.8, P 78/min, BP 127/72, Sat 100%. Chest, heart and abdominal exam were normal. Extremities were all normal. Neurological examination was normal. Laboratory Investigations revealed Hb 8.1 g/dl, WBC 21,000, PLT 1,117,000, Urea 4.1, Creatinine 41, AST 18U/l, ALT 19 U/l, ALP 114 U/l, Bil 86 micromol/l, chest X ray was normal (fig.1). ECG nonspecific ST/T wave changes. She was admitted on IVF, analgesia, oxygen, antibiotics. She was stable, but in the morning she developed sudden deterioration

of consciousness. The patient was evaluated by Neurologist. Brain CT scan revealed a big epidural hematoma with midline shift (fig.2). The neurosurgeon evaluated the case and ordered for evacuation of hematoma urgently. Our anesthesiologist evaluated the case and requested blood to be ready. The patient was intubated and shifted to ICU and immediately to OR. The epidural hematoma was evacuated. The Hb electrophoresis showed HbS 86.3, HbA 3.2, HbA2 4.4, HbF 6.1. Exchange transfusion was done. Hb 9.4g/dl, WBC 22,000, PLT 3,59,000. Hb Electrophoresis after exchange Hb S 14.3%, Hb F 1.2%, HbA2 2.9%, HbA 81%. Blood culture was negative, sputum culture was negative, urine culture was negative. The patient improved and was extubated and shifted to ward. Rehabilitation was done and patient was discharged home in good general condition. Repeat brain CT after surgery revealed that the epidural hematoma is completely evacuated. Midline shift is still present with focal edema in the the left cerebral hemisphere. Small pneumocephalus is noted (fig.3).

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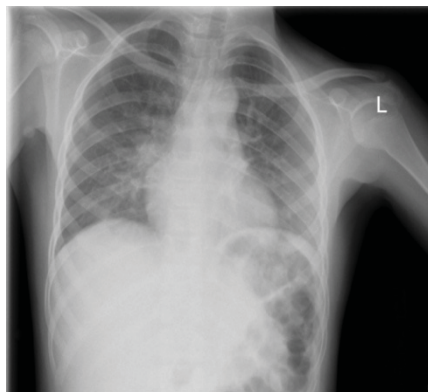


Fig.1: Chest x-ray was within normal limit

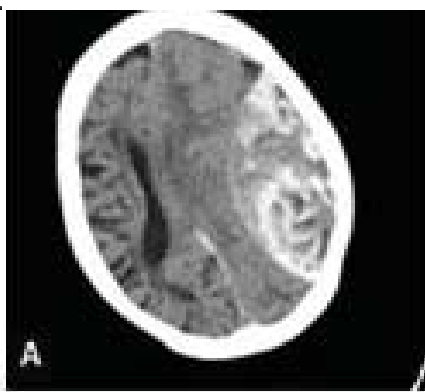


Fig.2: Brain CT Scan revealed a big epidural hematoma with mild shift

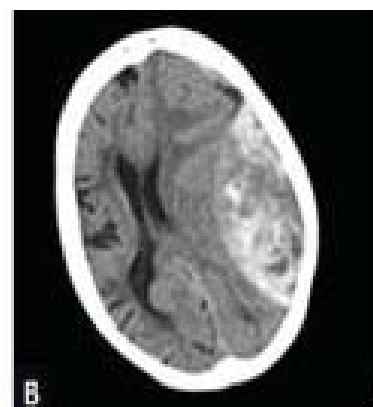


Fig.3: CT Scan revealed a big epidural hematoma

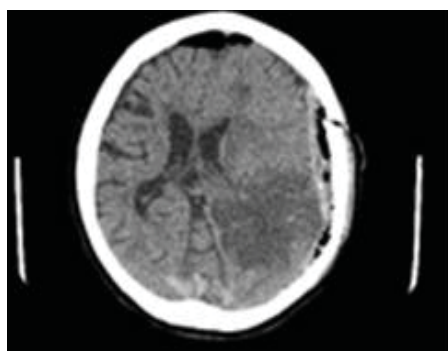


Fig.4: Repeat brain CT scan after surgery revealed that the epidural hematoma is completely evacuated. Mildline shift is present with focal edema in the left cerebral hemisphere. Small pneumocephalus is noted

#### Discussion:

Epidural haematomas are usually associated with preceding head trauma. The entity of non-traumatic spontaneous acute epidural haematoma is rare and most commonly occurs in the presence of infectious disease.<sup>10</sup> The first documentation of spontaneous EDH was by Schneider and Hegarty in 1951. It can also occur in the presence of coagulopathy, vascular malformations of the dura mater and haemorrhagic tumours.<sup>8,9,10</sup> Sickle cell disease, systemic lupus erythematosus, open heart surgery and haemodialysis have also been implicated as causative factors.<sup>10</sup> Acute neurological complications in patients of sickle cell anemia can be ischemic or hemorrhagic.<sup>13</sup> Cerebral ischemic complications are most common accounting for two-third of all neurological complications and are due to vaso-occlusive phenomenon in sickle cell

anemia.<sup>13</sup> Hemorrhagic complications are uncommon; intracerebral hemorrhage is common and subarachnoid hemorrhage and spontaneous epidural hematomas are very rare.<sup>13</sup> Spontaneous epidural haematoma constitutes a rare event that is not always easy to treat in the third world conditions.

Primary hemorrhagic stroke is an uncommon complication of sickle cell disease, with reported mortality rates of 24% to 65%.<sup>3</sup> Most reported cases are in adults. Intracranial hemorrhage accounts for approximately one-third of CVA's in patients with SCD. Hemorrhagic stroke was associated with a history of hypertension or antecedent events including transfusion or treatment with corticosteroids.<sup>3</sup> The site of bleeding may be subarachnoid, intraparenchymal, intraventricular, or a combination of these locations. Among hemorrhagic complications, intracerebral hemorrhage is common and subarachnoid hemorrhage and spontaneous extradural hematomas (EDH) are very rare.<sup>12</sup> Spontaneous extradural hematoma (EDH) is a very rare and uncommon complication of SCD. The peak incidence of ICH is between the ages of 20 and 29 in comparison to cerebral infarction, which is much more common in children. Approximately 3% of children with SCD will have a hemorrhagic stroke by 20 years of age, and 25 to 50% will die within two weeks of the event. Several cases of spontaneous EDH's have been reported. Since 1978, 15 case reports have described the occurrence of spontaneous EDH as a complication of SCD.<sup>15</sup>

In the Cooperative Study of Sickle Cell Disease, two major risk factors for hemorrhagic stroke were identified on multivariate analysis:

- Low steady state hemoglobin – RR 1.6 per 1 g/dL decrease
- Increased steady state leukocyte count – RR 1.9 per 5000/microL increase.

In addition, a small retrospective case-cohort study from two children hospitals in the US suggested that a recent history of HT, transfusions, or corticosteroid therapy was more likely in children with SCD who experienced hemorrhagic stroke compared to those with ischemic strokes.

In these case reports of EDH which have been reported, the hematomas were associated with bony infarcts of the skull and required aggressive surgical intervention. With no history of head trauma the pathogenesis of spontaneous EDH in patients with SCD is still not clearly defined(). Non traumatic spontaneous EDH has also been reported in association with other clinical conditions like coagulopathies, infectious diseases of the skull like sinusitis, vascular malformations of the dura mater and metastasis to the dura, or skull, and CKD.<sup>11</sup>

The clinical presentation of EDH in SCD patients is different from the classic description of post traumatic EDH which usually is characterized by a lucid interval. A high index of suspicion is needed for prompt diagnosis and treatment of this rare complication of SCD.<sup>14</sup> The frequent concurrence of skull infarctions along with spontaneous intracranial EDH has been noted to suggest a mechanism whereby infarction of the skull leads to periosteal elevation, disruption of the cortical bone margin, and bleeding into the epidural space.<sup>1,5,15</sup> Another theory proposes that insufficient venous drainage is the inciting event that leads to oedema and haemorrhage.

In spontaneous EDH not associated with skull infarction, the patients' skull is said to have abnormal anatomy because of a pathological process of chronic medullary haematopoiesis.<sup>15</sup> The proliferation of this hematopoietic skull tissue

with resultant expansion in response to acute anaemia results in the expanding hematopoietic tissue disrupting the inner and outer skull margins and precipitating extravasations of blood and hematopoietic tissue into the subgaleal and epidural spaces.<sup>4,6,15</sup> Despite the background haemoglobinopathy, this patient successfully underwent standard craniotomy and evacuation of EDH under general anaesthesia with complete resolution of symptom complication.

**Conclusion:** The best way of treating such a rare neurosurgical crisis of SCD is by preventing it.<sup>14</sup> As it is noticed an episode of sickle cell crisis in majority of cases just before occurrence of spontaneous EDH, we suggest that preventing sickle cell crisis would help us in preventing this rare entity.

Few simple measures like: Taking folic acid daily to help make new RBC's, drinking plenty of water daily (8-10 glasses for adults), avoiding too hot or too cold temperatures, and avoiding over exertion and stress.<sup>14</sup> Getting plenty of rest, and getting regular check-ups from knowledgeable health care providers will help such patients to prevent sickle cell crisis, hence preventing spontaneous EDH. Once EDH had developed and causing mass effect, then surgery should be performed with special precaution to avoid hypoxia, acidosis, increased blood viscosity (Hb >8.5 g/dL), dehydration, hypothermia, and stress in both intra and post operative period. Overzealous use of blood may increase the viscosity so should be avoided.

#### **Conclusion:**

Although a spontaneous extradural hematoma is a rare complication of sickle cell disease, it should be suspected when patients present with a sudden headache or other signs of intracranial hypertension.

#### **Role and contribution of Authors:**

Dr Mahmoud Abu Esleih, Consultant Internal Medicine, manage the case and wrote the initial write up

Dr Rafaat Elsayeed, Consultant Neurology,

manage the case and perform the medical management of the above patient and helped in collecting the references.

Dr Umar Farooq, Specialist Neurosurgery, helped in discussion writing and operated the case

Dr Reyad Shehab, Consultant Hematology, manage the case pre-operatively and helped in writing the discussion

Dr Sathya Moorthy, Specialist Radiology, helped in writing discussion and conclusion

Dr Khaled Habib, helped in writing the discussion

Dr Naveed Chawla, collected the references and helped in writing the discussion, he also assisted consultant neurosurgeon for evacuation of epidural haematoma.

**Conflict of Interest:** None

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