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SPONTANEOUS BILATERAL FRONTAL EXTRADURAL HEMATOMAIN A SICKLE CELL DISEASE: A CASE REPORT OF RARE NEUROSURGICAL CRISIS.

Saniya Sharma Doon Medical College, Dehradun

Rahul Awasthi* Max Institute of Neurosciences, Dehradun *Corresponding Author

ABSTRACT Spontaneous extradural hemorrhage (EDH) is a rare complication of sickle cell disease and is often associated with sinus infection, otic infections, dural vascular malformations, blood coagulopathies and skull bone infarction. We report an eighteen year old boy with sickle cell disease who presented with history of persistent headache, fever and one episode of loss of consciousness. Magnetic resonance imaging (MRI) of brain revealed bilateral frontal extra-dural hematomas (EDH) compressing on the brain. A bilateral frontal craniotomy and evacuation of the hematoma was performed and he made an uneventful recovery.

KEYWORDS : Spontaneous, Extradural, Hematoma, MRI

Introduction:
Sickle-cell disease is common in many parts of India, where the prevalence has ranged from 9.4 to 22.2% in endemic areas (1). Acute neurological complications in patients of sickle cell disease can be either ischemic or hemorrhagic. Primary hemorrhagic stroke is a not uncommon complication of sickle cell disease and spontaneous extra-dural hemorrhage (EDH) is an even more rare complication. The various causes reported include: Sinus infection, coagulopathies, vascular malformation of dura, middle ear or orbital infection, and tumor (2,3).

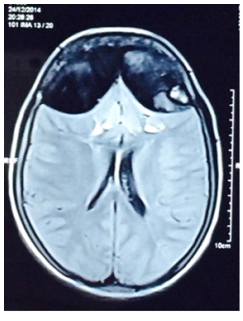
Case Report
An 18-year-old boy, first son of non-consanguineous marriage and a known case of sickle cell disease, presented with a history of persistent headache, fever and one episode of loss of consciousness. There was no history of trauma, seizures or limb weakness. He had been given blood transfusion one year back. On admission his vital signs were as follows: temperature 99.6°F, pulse 110 beats per minute and blood pressure 100/70 mm Hg. On neurological examination patient was drowsy and did not obey verbal commands. His GCS (Glasgow Coma Score) was $E_3V_3M_5$ with both pupils semi-dilated and sluggishly reacting to light at the time of admission. He had no other focal neurological deficit.

The investigations revealed: hemoglobin of 7.5 gm/dL, leukocyte count 9600/cu.mm, hematocrit of 24.1% and platelets 1.1 lakh/cu.mm. Peripheral blood smears showed microcytic hypochromic red blood cells (RBC) and sickle-shaped RBCs. Magnetic resonance imaging (MRI) of brain revealed symmetrical large acute extra-dural hematomas in bilateral frontal region, which were 6.0× 6× 6.2 cm on left side and 6.5× 4.2×7 cm on the right side causing significant mass effect on brain parenchyma and anterior corpus callosum. The calvarial and visualized facial bones appear thickened. There was no skull fracture, osteomyelitis or brain infarction [Figure 1]. The patient was taken up for urgent craniotomy for evacuation of the hematoma. A bicoronal skin flap was raised and bifrontal craniotomy was performed. The extra-dural hematoma was evacuated, leaving a thin rim of hematoma that was densely adherent to the superior sagittal sinus. The skull bone was soft and thinned out. The bone flap was of normal in color and consistency. Histopathological examination of blood clot showed presence of sickle shaped RBCs. The patient had an uneventful postoperative recovery. Post operative CT scan showed complete evacuation of hematoma with re-expansion of brain [Figure 1a].

Discussion
Spontaneous EDH is rarely reported in literature hence its incidence is not known. First documentation of this was done by Schneider and Hegarty in 1951. According to Babatola et al total 13 cases of spontaneous EDH in sickle cell disease [4].

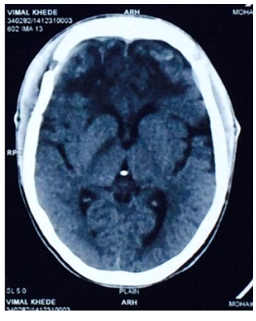
The lucid interval which is characteristic of post-traumatic EDH, is absent in cases of EDH in sickle cell disease patient and they usually present with features of raised intracranial tension. Acute neurological complications in patients of sickle cell disease can be divided into ischemic and hemorrhagic. Cerebral ischemic complications are most common neurological complications [2] and are due to vaso-occlusive phenomenon in sickle cell disease. Among hemorrhagic complications; intracerebral hemorrhage is common while subarachnoid hemorrhage and spontaneous extra-dural hematomas are very rare [3]. Spontaneous EDH as a complication of sickle cell disease often occurred along with skull infarction [5]. Possible pathophysiological mechanism responsible for such hematomas are : (a) Skull infarction causes periosteal elevation with disruption of the cortical bone margin, and bleeding into the epidural space;(b) Involvement of epidural vessels in the vicinity of infracted bone can rupture spontaneously and form such hematomas; (c) One theory suggests insufficient venous drainage leading to venous congestion (oedema) and rupture of these thin-walled veins (haemorrhage); (d) Another theory proposes expanding hematopoietic tissue causing disruption of the inner and outer skull tables with bleeding into subgaleal and epidural spaces[5,6].

Conclusions
A high index of suspicion is needed for prompt diagnosis and treatment of this rare complication of SCD. Best way of preventing such a rare crisis of sickle cell disease is by taking of folic acid (1mg) and penicillin (till 5 year of age), malaria chemoprophylaxis and blood transfusion, will help such patients to prevent sickle cell crisis, thus preventing spontaneous EDH.



1a

Figure 1a: MRI Showing bifrontal extradural hematoma



1b

Figure 1b: Post-operative Computed Tomography brain showing evacuation of hematoma

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