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Spontaneous Extradural Hematoma: A Rare Neurological Crisis in Sickle Cell Disease

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Abstract : *The Occurrence of spontaneous Extradural Hematoma (EDH) is rare neurological complication in patient with sickle cell disease. We report a twenty year old male patient with sickle cell disease who developed persistent headache and joint pain as a part of sickle cell crisis. One day He suddenly became altered conscious and on brain computed tomography (CT), a large Extradural Hematoma (EDH) in left parieto-temporal region with mass effect was found. No other etiologic factor was identified. Patient was successfully managed with left parieto-temporal craniotomy with evacuation of Haematoma without any undesirable sequel. He made a good recovery. We discuss the possible pathogenesis of this rare complication.*

Keywords: *Sickle cell disease, spontaneous extradural hematoma, computed tomography, craniotomy*

I. Introduction

The Sickle cell disease (SCD) is a qualitative hereditary hemoglobinopathy due to the presence of hemoglobin S [1]. Spontaneous extradural hematoma (EDH) is a very rare and uncommon complication of SCD. With no history of trauma, the pathogenesis of spontaneous extradural hematoma in patient with SCD is not clearly understood. Only 12 cases have been reported till date in medical literature [2-9]. The first documentation of spontaneous EDH was by Schneider and Hegarty in 1951[15]. Apart from Sickle cell disease, Spontaneous EDH has also been reported in association with dural vascular malformations, infections, tumor, and disorder of blood coagulation [9-14]. Here is a case of a young man having Sickle Cell Disease presented with spontaneous EDH following a sickle cell crisis. Patient was managed successfully with craniotomy and evacuation of hematoma.

II. Case Report

We report a case of a 20 year old male patient with sickle cell disease (SCD). A night before, he complained about headache associated with nausea and two episodes of vomiting. In the morning he was found altered conscious by his parents when they tried to wake him up. Patient was brought to Emergency Medicine Department of New Civil Hospital, Surat. There is no evidence trauma or convulsions. Patient had past history of multiple episodes of vaso-occlusive crisis requiring simple analgesia. On initial evaluation patient was found disoriented with pulse 64/min, BP 220/110 mmhg, and pupils were bilaterally semi dilated with Glasgow coma scale (GCS) 9/15 (E2V2M5). His Coagulation profile was within normal limits. His Hemoglobin was 8 gm% and hematocrit value was 28%. Computed Tomography imaging was performed using contiguous 6mm axial plain scan of brain from base to vertex. A large extradural hematoma in left parieto-temporal region was found with maximum width of 4.6cm and midline shift of 14 mm. Patient was transferred to emergency operation theatre and underwent an emergency left sided craniotomy under general anaesthesia. A large extradural hematoma was evacuated. Patient was managed in Surgical ICU post operatively. One unit of Whole blood (WB) and one unit of red cell concentrate (RCC) were given to this patient. Postoperative CT showed almost complete evacuation of hematoma. Patient regained consciousness one day after surgery and was discharged after seven days of hospital stay.

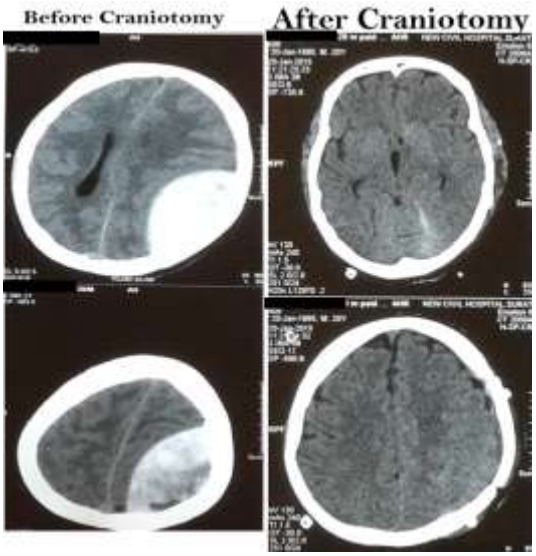
III. Discussion

The Sickle cell disease (SCD) is the most prevalent inherited blood condition worldwide resulting from single DNA mutation within the beta-globin [16]. It is common inherited disorder among people of sub-Saharan Africa, Middle East and India [17].Abnormal hemoglobin produces sickling of red blood cells under low oxygen tension leading to capillary occlusion. Affected individuals suffer constitutional manifestations, anemia, and ultimately organ damage due to micro and macro infarcts. Sickle cell anemia is associated with various complications and the average life expectancy is 42 years in males and 48 years in females [18]. Central nervous system (CNS) complications in sickle cell disease are rarely described in medical literature and may be either due to vaso-oclusive or hemorrhagic complications. Cerebral ischemic complications are common accounting for two third of all neurological complications [19]. Hemorrhagic complications are uncommon. Among

hemorrhagic complications, intracerebral hemorrhage is common and subarachnoid hemorrhage and spontaneous extrdural hematomas (EDH) are very rare [20].

Spontaneous extradural hematomas as a complication of sickle cell disease have been reported mostly secondary to bone infarction [21]. Possible mechanisms for such hematomas are: [21, 22] (1) Periosteal elevation secondary to bone infarction with disruption of the cortical bone margin, and bleeding into the epidural space; (2) Poor venous drainage leading to venous congestion and rupture of these thin-walled veins and (3) Episodes of sickle cell crisis puts extra demand over hematopoietic skull tissue causes its expansion and disruption of the inner and outer skull tables with bleeding into subgaleal and epidural spaces.

The exact mechanism is not clear in our patient as there was no evidence of skull bone infarction on CT and skull looked normal during craniotomy. However, disruption of cortical bone margin might have occurred at microscopic level in our patient who had history of multiple vaso-occlusive crisis before. Despite the hemoglobinopathy, this patient successfully recovered after surgical intervention.



IV. 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. Operative management is associated with excellent outcomes as demonstrated in our index patient.

V. Take Home Message

1. Spontaneous EDH in SCD is too rare neurological complication. High Index of suspicion is required. Any sickle cell disease patient with symptoms and signs of raised intracranial tension should be referred to cranial CT.
2. Acute drop in hematocrit in SCD patient should alert clinician and clinician should have low threshold for brain imaging for such cases.
3. Surgical Evacuation remains the standard of treatment for this condition and the presence of sickle cell haemoglobinopathy should not serve as contraindication.
4. The best way of preventing such rare neurological crisis of SCA is by preventing it. As we know occurrence of sickle cell crisis just before occurrence of spontaneous EDH in most of reported cases, we suggest the preventing sickle cell crisis would help us in preventing this rare complication.
5. How to prevent?? Simple measures like taking Folic acid daily, drinking plenty of water (8-10 glasses for adults), avoiding too hot or too cold temperature, getting enough rest, avoiding over stress, and getting regular checkups.

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