Article published online: 2022-09-20

CASE REPORT

Spontaneous extradural and subgaleal hematoma: A rare neurosurgical crisis of sickle cell disease

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ABSTRACT

Extradural hematoma (EDH) in absence of trauma is a rare entity with only few cases reported in literature. The various causes reported include: Vascular malformation of dura, coagulopathies, sinus infection, middle ear or orbital infection, and tumor. Occurrence of spontaneous EDH as a complication of sickle cell disease is even much rarer. We report a case with sickle cell disease who presented with spontaneous extradural and subgaleal hematomas following an episode of vaso-oclusive crisis. He was managed successfully with surgery. The association of epidural hematomas in sickling hemoglobinopathies is reviewed. In all cases, we noticed one episode of sickle cell crisis just before the occurrence of spontaneous EDH. Perhaps this crisis puts an extra demand over the hematopoietic skull tissue disrupting inner and outer skull margins leading to spontaneous EDH and subgaleal hematoma.

Key words: Sickle cell disease, skull infarctions, spontaneous extradural hematomas

Introduction

Sickle cell disease is a common inherited blood disorder among people of African descent but also occur in the Mediterranean, India, and the Arabian Peninsula. The clinically important variants include homozygous hemoglobin SS (sickle cell anemia) and the compound heterozygous variants sickle B thalassemia, sickle C (SC), and sickle D (SD) diseases. 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. Central nervous system (CNS) complications may be either due to vaso-oclusive or hemorrhagic complications. Cerebral ischemic complications are common accounting for two third of all neurological complications. Hemorrhagic complications are uncommon among hemorrhagic complications intracerebral hemorrage is common, subarrachnoid, or spontaneous extradural hematoma (EDH) are less common.

Access this article online	
Quick Response Code:	Website: www.asianjns.org
	DOI: 10.4103/1793-5482.144177

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Case Report

An 18-year-old boy, a known case of Sickle cell disease, was admitted to medicine ward of our hospital with complaints of low back pain, chest pain, and pain around knee joint. He was managed conservatively as a case of sickle cell vaso-occlusive crisis. His symptoms improved, and he was discharged after three days of conservative management. Two days after discharge to his home, he developed headache followed by a boggy swelling over his Lt Parietal area [Figure 1]. As sensorium decreased, he was again admitted to medicine ward. Computed tomography (CT) scan of head showed Rt parietal biconvex heterogeneously hypodence extradural lesion with a volume of 70 cc and midline shift of 6 mm along with Lt parietal subgaleal hematoma [Figure 2]. Bone window in CT scan and X-ray of the skull showed an increase in marrow proliferation with thin cortical bone margins [Figures 3-5]. He was transformed from medicine ward to Neurosurgery ward. On initial evaluation in our

How to cite this article: Mishra SS, Senapati SB, Gouda AK, Behera SK, Patnaik A. Spontaneous extradural and subgaleal hematoma: A rare neurosurgical crisis of sickle cell disease. Asian J Neurosurg 2017;12:47-50.

ward, he was found to be disoriented with pulse 64/min, BP 130/80 mm Hg, Pupil Rt 5 mm and Lt 3 mm in size with GCS of E3V3M5. Patient attendant gave no recent history of head trauma or bleeding disorder in past. His hematological profile was as follows: Hemoglobin 7.4 g/dL, hematocrit 30%, and platelet count 250,000 platelets/mm^3. PT, aPTT, and INR was within normal limit. Peripheral blood smear shows microcytic hypochromic RBC without premature cells.

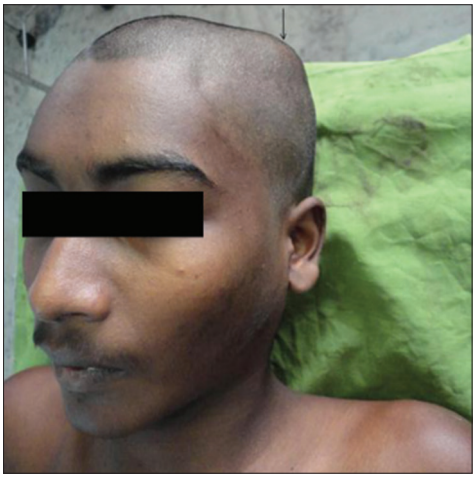

Figure 1: Arrow showing subgaleal hematoma over Lt Posterior parietal area

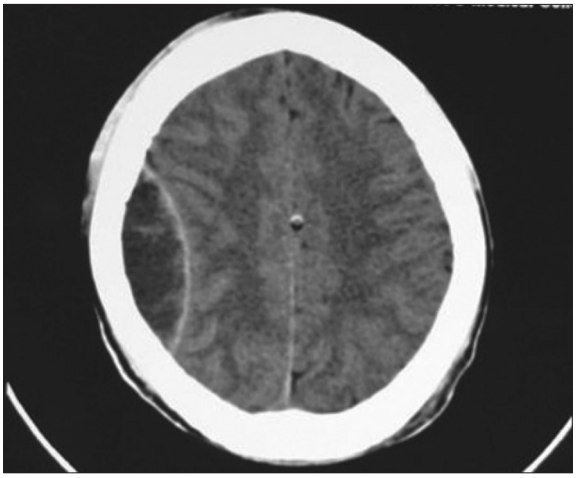

Figure 2: Axial CT scan of head shows Rt parietal heterogeneously hypodence biconvex extradural lesion

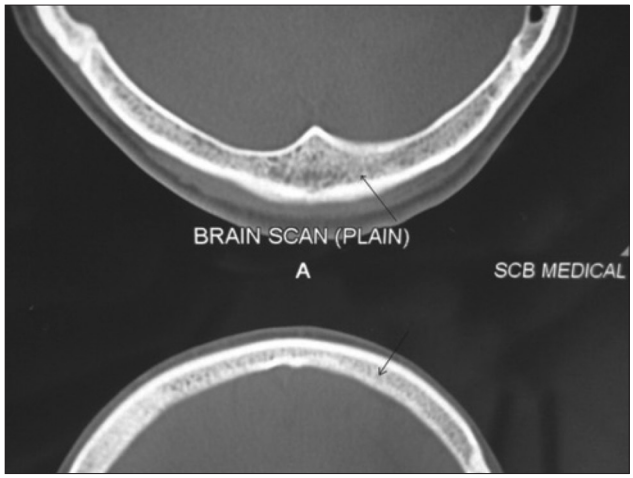

Figure 3: Bone window in CT scan showing increased marrow proliferation with thin cortical bone margins

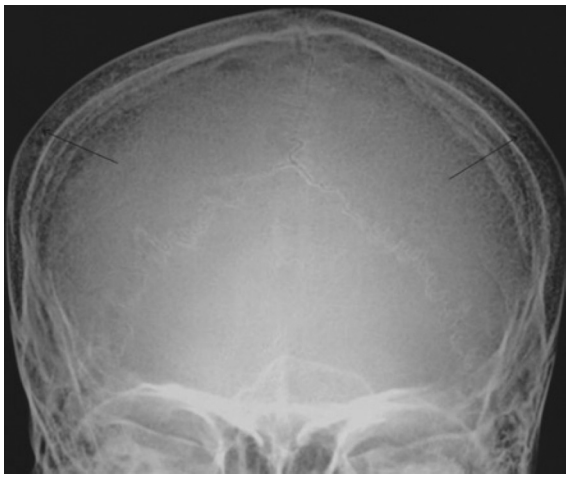

Figure 4: X-ray skull showing increased marrow proliferation with thin cortical bone margins

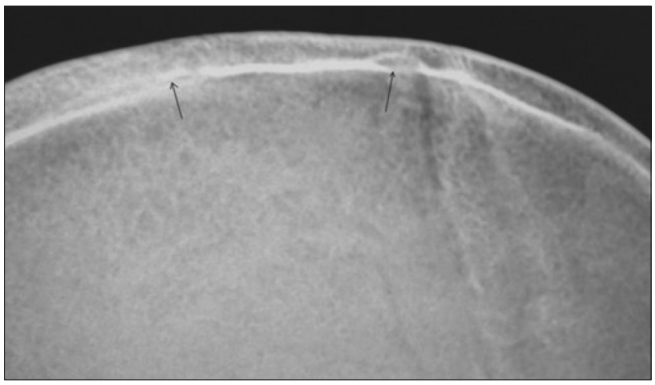

Figure 5: X-ray skull showing break in continuity of inner skull margin

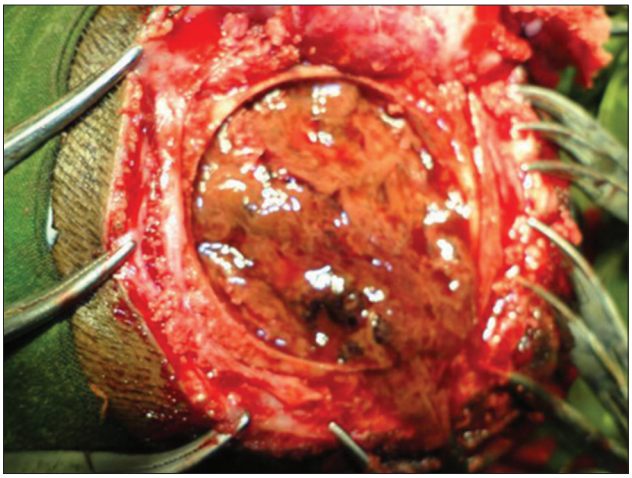

Figure 6: Trephine craniotomy showing altered blood and its degraded product in extra dural space

When we analyzed CT scan, there were two possibilities; biconvex heterogeneously hypodence extradural lesion could be either a case of chronic EDH or a case of acute EDH containing unclotted blood and looking hypodence due to low hemoglobin level. As there was mass effect with midline shift 6 mm, we planned for surgery. Rt parietal trephine craniotomy done liquid altered blood came out, below liquid blood there was a layer of yellowish plaque which was sucked out [Figure 6] and send for cytological and culture sensitivity study which later came as having no malignant cells, containing blood degraded products and free from any organism. Underlying dura was intact without definitive bleeding point. When we examined the bone flap, we found that there was a gross bone marrow proliferation with thin cortical bone margin [Figure 7]. Inner bone margin was papery thin with areas of blackish discoloration and pinpoint bleeding sites [Figure 8]. Histo pathological examination of a bone piece reviled hyper-proliferative bone marrow [Figure 9]. Post operative period was uneventful. He was discharged on 7th postoperative day with GCS of 15/15. On follow up he is doing well [Figure 10].

Discussion

Spontaneous intracranial epidural hematoma is rare entity. First documentation of this was done by Schneider and Hegarty in 1951. The various causes reported include: Vascular malformation of dura, coagulopathies, sinus infection, middle ear or orbital infection, and tumor. Occurrence of spontaneous EDH as a complication of sickle cell disease is rarer. We reviewed reported cases including each case references available on Pubmed and scholar, and google. We found that only nine cases were documented in literature. Out of nine cases, skull infarction was the probable cause of spontaneous EDH in seven cases,[1-7] which was confirmed by doing either preoperative or post operative MRI showing infarction in skull bone. Hyper-proliferative bone marrow disrupting the inner and outer skull margins and precipitating extravasations of blood into the subgaleal and epidural spaces was probable

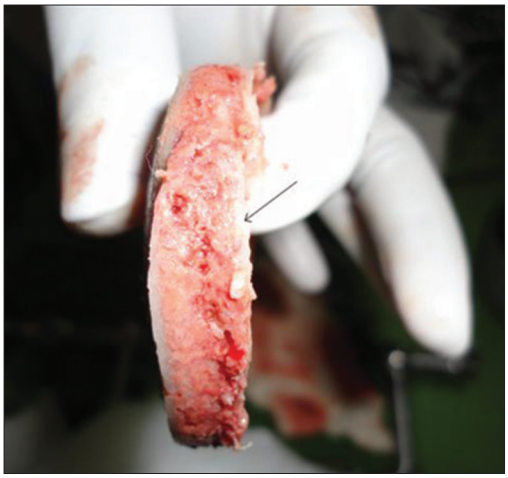

Figure 7: Craniotomy bone clearly showing increased hematopoietic skull tissue Proliferation with thin cortical bone margin

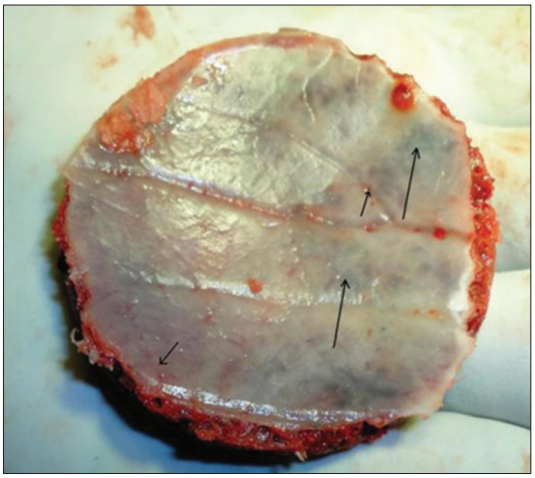

Figure 8: Inner surface of Craniotomy bone showing papery thin cortex with areas of Blackish discoloration and pinpoint bleeding sites

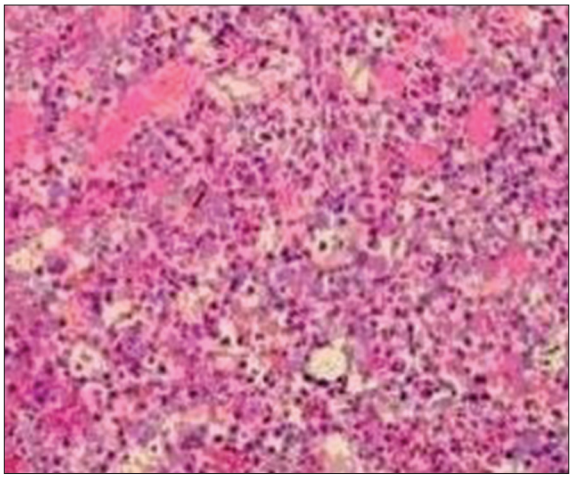

Figure 9: Histo pathological examination of a bone piece reviled hyper-proliferative bone marrow

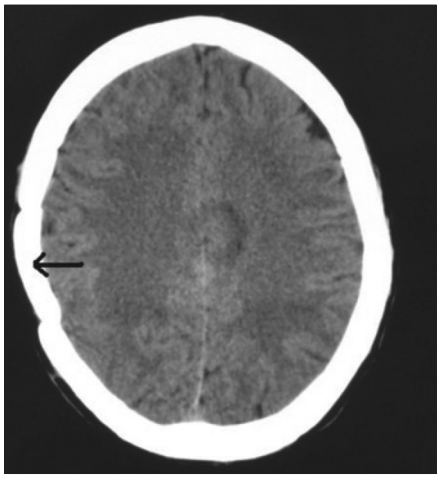

Figure 10: CT scan after 3 months of surgery at the time of follow up, showing previous craniotomy site with normal brain scan

cause in two cases.[8-10] In our case, preoperative CT scan and X-ray finding of increased marrow proliferation, thin cortical bone margin with intra operative finding of papery thin inner cortex, areas of blackish discoloration, and pinpoint bleeding sites points toward the second possible cause. In all cases, we noticed one episode of sickle cell crisis just before the occurrence of spontaneous EDH. Per halves this crisis puts an extra demand over the hematopoietic skull tissue disrupting inner and outer skull margins lading to spontaneous EDH and subgaleal hematoma.

Conclusion

The best way of treating such a rare neurosurgical crisis of sickle cell disease is by preventing it. As we noticed an episode of sickle cell crisis in all 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 red cells, drinking plenty of water daily (8-10 glasses for adults), avoiding too hot or too cold temperatures, avoiding over exertion and stress, 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.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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