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"Think Beyond Trauma!" Multiple Epidural Hematoma in Sickle Cell Disease: A Conundrum

Ruchika Bhatnagar MD [*1], Anita Kumari MD[2]

1. Associate Professor, Department of Pediatrics, Government Institute of Medical Sciences, Greater Noida, Gautam Buddha Nagar, Uttar Pradesh, India
2. Professor and Head, Department of Pediatrics, Government Institute of Medical Sciences, Greater Noida, Gautam Buddha Nagar, Uttar Pradesh, India
*Corresponding author: Dr Ruchika Bhatnagar, Associate Professor, Department of Pediatrics, Government Institute of Medical Sciences, Greater Noida, Gautam Buddha Nagar, Uttar Pradesh, 201310, India. Email: ruchu.bhatnagar@gmail.com. ORCID ID: 0000-0003-3509-0555.

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Abstract
Epidural hematoma is conventionally thought to occur secondary to trauma. Atraumatic/spontaneous epidural hematoma is of rare occurrence and the etiology spans from dural vascular malformations, neoplasms, and coagulopathy to sinus, middle ear, and orbital infections. Occurrence of atraumatic epidural hematoma in sickle cell disease is rarely reported as opposed to frequent neurological complications, such as ischemic stroke seen in 54% of cases, followed by intracerebral, subarachnoid, intraventricular, or subdural hemorrhages. This study reports the case of an eight-year-old male with Sickle cell disease (SCD). He suffered from fever followed by splenic sequestration and multiple spontaneous epidural hematomas without mass effect and successfully managed with conservative treatment.
Keywords: Anemia, Epidural hematoma, Ischemic stroke, Sickle cell disease, Vascular malformations

Introduction

Sickle cell disease (SCD) refers to the group of inherited disorders affecting the quality of hemoglobin, which include sickle cell anemia and India has been reported to have the second-highest SCD burden in the world (1). SCD conveys a greater risk of cerebrovascular disease, with an estimated stroke prevalence of 4.2% (2). Ischemic stroke accounts for the majority of cases while subarachnoid hemorrhage is the commonest subtype of hemorrhage. Hemorrhagic complications in SCD are commonly accompanied by intracerebral, intraventricular, subarachnoid, and subdural bleeding. Epidural hematoma (EDH) is conventionally linked with head injury leading to skull fracture. Various etiologies implicated in causing atraumatic EDH include dural vascular anomalies, coagulation disorders, sinusitis, and middle ear or orbital infections. Epidural hemorrhages are infrequent and multiple, atraumatic/spontaneous EDH is an extremely uncommon complication of SCD. This study reports the case of an eight-year-old male with SCD. The case had fever followed by acute onset headache and multiple spontaneous epidural hematomas without mass effect and successfully managed with conservative treatment.

Case Report

An eight-year-old boy presented to the pediatric outpatient department (OPD) with complaints of right upper limb pain for three days and a high-grade fever for one day. The child had a known case of SCD, diagnosed at the age of 3 years. Over the past five years, he was never diagnosed with any significant acute sickle cell episodes requiring hospitalization, and he was receiving hydroxyurea therapy. His eldest brother aged, 18 years, also had SCD and had undergone splenectomy at the age of five years. On general examination, the child was conscious and oriented, and

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febrile with moderate pallor, icterus. He had a blood pressure of 110/76 mmHg, a respiratory rate of 28/min, a heart rate of 122/min, and a pulse oxygen saturation of 97% on room air. His cardiorespiratory and central nervous system evaluations showed values within normal limits. On per abdominal examination, the liver was palpable 2cm below the right subcostal margin and the liver span was 13 cm, the spleen was palpable 3cm below the left subcostal margin along the longitudinal axis of the 10^{th} rib. Complete blood count revealed hemoglobin (Hb) of 7.5gm%, total leucocyte count of 7000 cells/mm^3, and platelet count of 90,000/µl. The results of other relevant blood tests are displayed in Table I. Supportive treatment measures were instituted, comprising intravenous fluids and antipyretics. Within 24 hours of admission, he developed headache, and altered sensorium without any focal neurological deficit. Clinical pallor and hepatosplenomegaly were found to increase, and this was further proved by a drop in Hb from 7.5 to 4 g/dl and a platelet count of 40,000/µl suggested splenic sequestration as the underlying culprit. On neuroimaging of the brain, the bone window appeared to be normal. On the brain window, however, multiple hyper-dense crescentic-shaped, extra-dural hemorrhages were seen along the right frontal, temporal, and left temporal regions with no features of mass effect/mid-line shift (arrows in Figure 1). There was no antecedent history of head injury or bleeding diathesis, and coagulation profile results were within normal limits. In the absence of significant mass effect/midline shift, the child was managed conservatively with adequate hydration, packed red blood cells, and platelet transfusions. The patient turned afebrile, was neurologically intact with Hb of 9 gm% and platelet count of 1.5 lakhs/µl, and thus he was discharged from the hospital after seven days. Repeat neuroimaging, which was done after three months, revealed the resolution of the epidural hematomas.

Table I: Laboratory findings of the case

Lab parameters	Day 1	Day 2	Day 7
Hemoglobin (gm%)	7.5	4	9
Total leucocyte count (cells/mm^3)	7000	5000	6500
Platelet count (cells/µL)	90,000	40,000	1.5 lakh
Blood urea (mg/dl)	46	41	-
Serum creatinine (mg/dl)	0.3	0.4	-
Serum sodium (mEq/L)	133	135	-
Serum potassium (mEq/L)	4.1	3.8	-
Total serum bilirubin (mg/dl) **Direct/Indirect (mg/dl)** **SGPT (IU)**	3.5 0.6/2.9 34	-	-
PT (sec)/aPTT(sec)/INR	15.8/54.6/1	16/51.2/1	-

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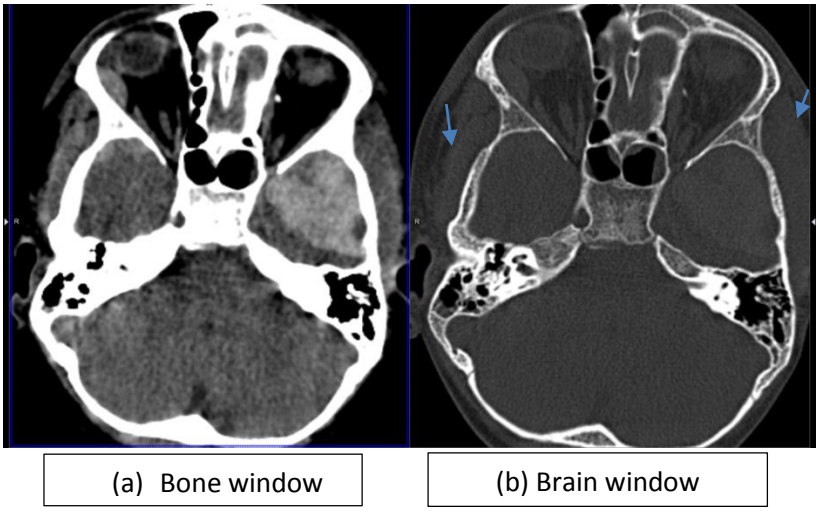

Figure 1. Non -contrast Computed Tomography (NCCT) head showing extra-axial hemorrhages (arrows) along right and left temporal convexity while bone window showing normal bone scan

Discussion

Neurological complications are frequently seen in SCD. Owing to increased cognizance, advanced neuroimaging modalities, and greater insights into the literature review, there is a growing number of reports of spontaneous EDH in children with sickle cell disease. Few reports highlighted that spontaneous EDH could be the sole presentation in an asymptomatic SCD patient (3). The majority of the reports focus on male patients who are pre-adolescent or adolescent. The most commonly reported symptoms upon admission included headache (39%), vaso-occlusive crisis (34%), and reduced consciousness (13%) (4). The present case had fever and pain in limbs followed by acute onset headache; however, few authors reported that convulsive crisis, periorbital edema, and proptosis heralded the onset of EDH in SCA patients. The most common sites of the EDH, as reported in the literature, have been bilateral involvement accounting for 40% of the cases while frontal and parietal regions, each account for 36% of the cases (4). Unlike this, the frontal and bilateral temporal regions have been involved in the index case. The three primary suggested mechanisms for spontaneous EDH in SCD are bone infarction, expansion of hematopoietic marrow, and changes in blood flow caused by increased viscosity (5, 6, 7). According to the last mechanism, slow venous blood flow caused by hyperviscosity leads to venous congestion and results in rupture (7).

Concerning the present case, fever, and subsequent dehydration triggered sickling. The sluggish blood flow across cerebral venous circulation could have led to venous congestion, rupture of veins, and occurrence of multiple extra-axial hemorrhages manifested as acute onset headache. The significant drop in Hb and platelet count and increased splenic size further substantiated the occurrence of splenic sequestration in the present case which might have also triggered the vaso-occlusive crisis (VOC). A similar occurrence of non-traumatic EDH with a sickling bony crisis was reported by Joy et al. in a 20-year-old Indian male with sickle cell anemia, however, due to large left-sided parietal-temporal epidural hematoma with midline shift and mass effect neurosurgical intervention was required in their case (8). Other authors have also enumerated VOC and rapid marrow expansion as the underlying causes of EDH, but the occurrence of disseminated

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intravascular coagulation portends a poor prognosis and an unfavorable outcome in such patients (4). Thrombocytopenia has also been reported as one of the triggers resulting in EDH, thus complicating the clinical outcome in such patients (9). EDH was also reported to be usually unilateral, and associated with overlying bony infarctions (5). Regarding this case, however, multiple EDHs without any evidence of skull bone infarction were observed. Detecting bone infarction can be difficult, and MRI appears to be the most sensitive method for identifying infarctions, whereas CT is less productive, particularly in the acute phase of the disease. (10). In the cases documented in the literature, 45% were treated conservatively, while 55% needed surgical intervention. It was noted that patients receiving conservative treatment had a survival rate of 91%, compared to a 61.5% survival rate for those who underwent surgery. The overall mortality rate among the cases was 22% (4). The present case was also managed conservatively and showed a neurologically intact outcome. This is as per the similar case profile of a 14-year-old boy, as reported by Takroni et al. (10).

Conclusion

Multiple EDH is a rare complication of SCD. Clinicians and neurologists should maintain a strong level of suspicion for diagnosing EDH, if a child with SCD suffers from headaches with or without focal neurological deficits. Fever and splenic sequestration act as triggers for VOC in such patients. Early diagnosis and specific measures can result in a neurologically intact outcome. The frequency of EDH and its predisposing factors are suggested as some potential areas for future research.

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Author's Contributions

AK conceptualized the study. AK and RB conducted a literature review. RB drafted the manuscript and owns primary responsibility for the final content. All the authors revised it critically and approved the submitted manuscript.

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Conflict of Interest

None

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