Case Report

Spontaneous epidural hematoma: A case report of rare crisis of sickle cell disease

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Abstract

Introduction: Sickle cell disease (SCD) is defined as an autosomal recessive disorder characterized by the production of abnormal hemoglobin S and is correlated with high morbidity and mortality. The clinical consequences of SCD include pain crisis, acute chest syndrome, and strokes. Spontaneous epidural hematoma is a rare manifestation in sicklers with few cases reported in the literature.[6] The pathophysiology is not completely understood. However, a few explanations have been reported over the years that include vaso-occlusion of the bone resulting in bone infarction, microfracture due to rapid expansion of hematopoiesis of the inner cortex, and sludging of the sickle cells in the diploic veins—all result in leaking of blood in the epidural or in the subgalea space. **Patient Concerns:** A 14-year-old boy known to have SCD (Hb SS) presented to the Security Forces Hospital with a history of diffuse headache associated with nausea that started 12 h prior to presentation. **Diagnosis:** Computed tomography (CT) showed bilateral frontal epidural hematoma and subgaleal space. **Intervention:** A multidisciplinary team was created (hematology, neurology, neurosurgery, and interventional radiology) and a plan was formulated as follows: Continuous monitoring of the patient's neuro vital signs and transfuse the patient with blood and platelets in addition with Levetiracetam. **Outcomes:** The patient was discharged after 9 days of hospital admission. He has remained symptom-free post-transfusion. Post-discharge CT scan showed a reduction in the hematoma size. **Conclusion:** A high index of suspicion is needed for a prompt diagnosis and treatment of this rare complication of SCD. The management strategy of EDH depends on the level of consciousness of the patient upon presentation. Surgical approach with craniotomy and evacuation or conservative management have been used with full recovery of the patients.

Keywords: Rare, Saudi Arabia, sickle cell disease, spontaneous epidural hematoma

Introduction

Sickle cell disease (SCD) is defined as an autosomal recessive disorder characterized by the production of abnormal hemoglobin S and is correlated with high morbidity and mortality.

Saudi Arabia has a population of approximately 23.98 million. Information about the prevalence of SCD in Saudi Arabia is inconsistent.

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The pathophysiology is not completely understood. However, a few explanations have been reported over the years.

Case Report

Patient information

A 14-year-old boy known to have SCD (Hb SS) presented to the Security Forces Hospital with a history of diffuse headache associated with nausea that started 12 h prior to presentation.

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The patient denied any history of fever, neck pain, altered consciousness, skin rashes, trauma, and bleeding disorder in the past.

The patient was recently discharged from the hospital due to vaso-occlusive and hemolytic crises which were treated with blood transfusion and hydration.

The patient descends from a family with a history of Hb SS with three affected siblings.

Clinical findings

The patient's examination revealed a fully conscious, pale, jaundiced but not febrile with pulse 64 beats/min, BP 130/80 mmHg, pupil Rt 5 mm, and Lt 5 mm in size with a Glasgow Coma Score of 15\15 (E4V5M6). There was no evidence of head trauma and no abnormal neurologic signs. A boggy swelling was noted over his right partial area.

Diagnostic assessment

The initial laboratory test showed hemoglobin (Hb) of 8.9 mg/dL (hematocrit: 0.27 – mean corpuscular volume: 78.2 Fl), platelet count was 81,000, erythrocyte sedimentation rate (ESR): 112, and C-reactive protein (CRP): 247.66 [Table 1].

The patient underwent head computed tomography (CT) that showed bilateral frontal epidural hematoma (EDH), largest on the left side reaching a maximal thickness of 1.8 cm [Figure 1].

The left parietal epidural hematoma was 9.7 mm in maximal thickness with interior inferior hypodensity representing a swirling sign indicative of the ongoing active hemorrhage [Figure 2]. Small-side subgaleal space was also noted in the contralateral side.

There was a mild mass effect but there was no apparent midline shift. No herniation, or hydrocephalus, acute territorial infarction, or parenchymal contusions were noted. The ventricular system, basal cisterns, and posterior fossa structures were normal. The visualized bony structures show no fractures [Figure 3].

A multidisciplinary team was created (hematology, neurology, neurosurgery, and interventional radiology) and a plan was formulated as follows: Continuous monitoring of the patient's neuro vital signs and transfuse the patient with blood and platelets. In addition, Levetiracetam was initiated to prevent any seizure activity with the possibility of and the need for urgent surgery in case of any neurological deficits suggestive of intracranial bleeding development.

Therapeutic intervention

The patient was kept in the intensive care unit for observation as conservative management was chosen for him.

Table 1: Laboratory test

Lab	Result	Normal value
Complete blood count		
WBC (10 $\times^{9}$/L)	6.23	4.5-13.5
RBC (10 $\times^{12}$/L)	3.48	3.8-6.5
HGB G/L	89.0	11.5-180
HCT %	0.272	0.35-0.52
MCV FL	78.2	77-98
MCHC G/L	327.0	310-360
PLT	81	150-400
Inflammatory markers		
ESR mm/HR	112	0-20
CRP	247.66	Less 5.0
Electrolytes		
NA mmol/L	136	136-145
K mmol/L	3.4	3.5-5.1
UREA mmol/L	4.6	2.76-8.07
CR umol/L	40	62-106
Lactic acid dehydrogenase		
LDH U/L	1788	135-225
Liver function tests		
ALT U/L	18	UP TO 41
AST U/L	36	UP TO 40
ALK.PHOS U/L	359	82-331
BILIRUBIN TOTAL umol/L	44.6	0-17.1
CONGATED umol/L	32	0-3.4
GAMMA GT u/L	54	8-61
Iron studies		
Iron umol/L	30.4	5.83-34.5
Transferrin g/L (TIBC)	1.51	2.0-3.6
Transferrin	81	15-45%
Haptoglobin		
Haptoglobin g/L	<0.1	0.3-2.0
Electrophoresis		
HB A%	75.1	
HG A2%	3.3	2.2-3.7
HB S%	21.6	
Reticulocyte count		
Retics %	4.31	0.5-1.5
Coagulation profile		
PT SEC	14.2	10.0-14.1
INR	1.23	0.86-1.2
APTT SEC	38.9	24.6-40.1

During his stay, the patient received blood and platelet transfusion and his hydroxyurea increased from 1,000–1,500 mg once daily and Levetiracetam was added to the patient as a prophylactic measure and he underwent imaging again to ensure size reduction.

Follow-up and outcomes

The patient was discharged after 9 days of hospital admission. He has remained symptom-free post-transfusion. Post-discharge CT scan showed a reduction in the hematoma size.

No other inherited or acquired risk factors for bleeding have been recognized [Table 2].

Table 2: Laboratory test		
Lab	**Result**	**Normal value**
Factor XIII		
Factor XIII %	59.3	75.2-154.8
Platelet function analyzer		
Col/Epi	166	80-175 (closure times in s)
Col/ADP	101	71-118 (closure times in s)

Discussion

SCD is defined as an autosomal recessive disorder characterized by the production of abnormal hemoglobin S and is correlated with high morbidity and mortality.[1] In SCD, a single amino acid substitution in the β-globin chain leads to the polymerization of mutated hemoglobin S, damaging erythrocyte morphology, and endurance.[2]

Saudi Arabia has a population of approximately 23.98 million. Information about the prevalence of SCD in Saudi Arabia is inconsistent, but studies have conveyed that SCD is a relatively common genetic disorder in this specific part of the world.[3]

The clinical consequences of SCD include pain crisis, acute chest syndrome, and strokes.

Epidural hematoma (EDH) is a collection of blood between the dura and the inner part of the skull.[4] It is almost always caused by trauma associated with skull fracture as a result of bleeding from ruptured middle meningeal vessels or diploic veins.[4] Spontaneous EDH is rarely reported in the literature and its incidence is not known.[5]

Spontaneous epidural hematoma is a rare manifestation in sicklers with few cases reported in the literature.[6]

The pathophysiology is not completely understood. However, a few explanations have been reported over the years. Vaso-occlusion of the bone results in bone infarction and leaking of blood in the epidural or in the subgaleal space. The other explanation includes microfracture due to the rapid expansion of hematopoiesis of the inner cortex leading to extravasation of blood and hematopoietic tissue. Sludging of sickle cells in the diploic veins leads to insufficient venous drainage and blood oozing due to vascular injury and elevated backpressure is another proposed mechanism.[6-9]

The clinical presentation of EDH in SCD patients is usually preceded by sickle cell crisis in most of the reported cases and it differs from the classic description of post-traumatic EDH which is characterized by a lucid interval.[8]

Prior to the EDH, patients typically present with crises, which are difficult to control with analgesics and fluids. Soon after, the affected patients may experience headaches followed by a rapid decline in the neurologic state. Other manifestations include hemiparesis or hemiplegia, and some patients present with soft scalp swellings that develop rapidly and without evidence of trauma and drop in hemoglobin which cannot be solely attributed to the crisis.[10-12]

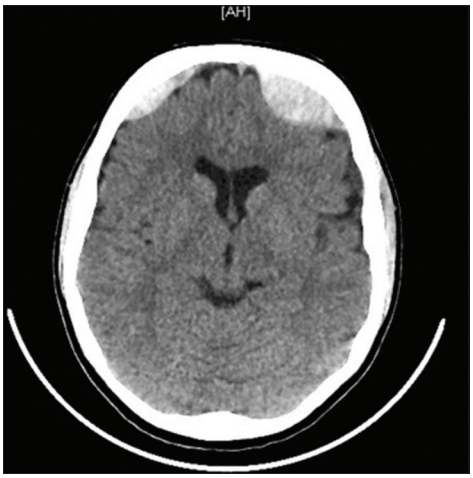

Figure 1: Unenhanced CT scan of the brain showing bilateral frontal epidural hematoma.

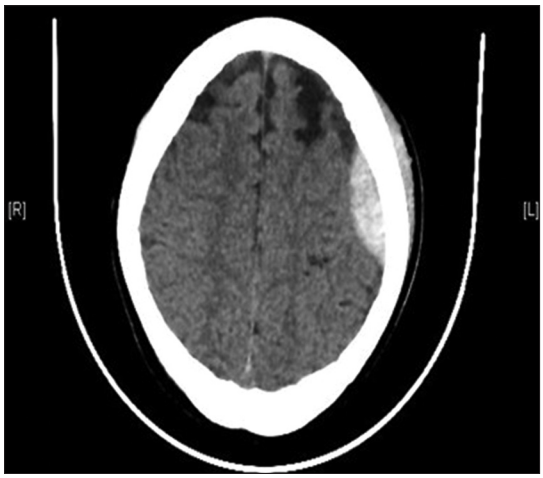

Figure 2: Left parietal epidural hematoma 9.7 mm in maximal thickness.

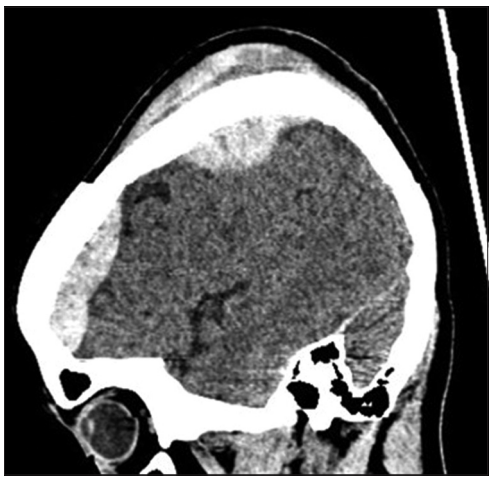

Figure 3: Subgalea hematoma

A high index of suspicion is needed for prompt diagnosis and treatment of this rare complication of SCD. Workup labs to request when evaluating the patient with suspect EDH include complete blood count and hemolysis panel (liver function tests, clotting profile, and lactic acid dehydrogenase, haptoglobin, and reticulocyte count). Imaging modalities such as ultrasound can help in determining if scalp swellings are hematomas while head CT scans are the definitive modality of diagnosis.[13]

The management strategy of EDH depends on the level of consciousness of the patient upon presentation. A surgical approach with craniotomy and evacuation is the definitive treatment for the unconscious patient. The other approach includes conservative management with close follow-up to document resolution of the hematoma or referral for the surgery if needed. Full recovery has been documented using both approaches.[9,14]

Conservative management was chosen for our patient as his clinical and radiological conditions were stable.

SCD has a devastating effect on the patient's quality of life. As a primary care physician, following the patient post-discharge and during the attack-free period leads to early detection of the disease complication and a better quality of life.

Conclusion

Spontaneous epidural hematoma is a rare complication of SCD. A high index of suspicion is needed for prompt diagnosis and treatment of this rare complication of SCD. The best way of treating such a rare crisis of SCD is by preventing it with strict control of the disease with the use of hydroxyurea, folic acid, and adequate hydration.

As a primary care physician, following the patient post-discharge and during the attack-free period lead to early detection of the disease complication and a better quality of life.

Declaration of patient consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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The authors whose names are listed above certify that they have no affiliations with or involvement in any organization or entity with any financial interest or non-financial interest in the subject matter or materials discussed in this manuscript.

Conflicts of interest

There are no conflicts of interest.

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