Acute soft head syndrome in a sickle cell disease patient

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ABSTRACT

Acute soft head syndrome is an extremely rare complication in children with sickle cell anemia. We present a 16-year old male patient known to have sickle cell anemia who presented to our emergency department with low grade fever, headache, skull pain and swelling. Magnetic Resonance of the brain was done demonstrating subgaleal collections overlying calvarial signal abnormalities. The combination of clinical and radiological findings were indicative of acute soft head syndrome. Acute soft head syndrome is a rare complication in children with sickle cell anemia and the pathophysiology is related to osteonecrosis and bone infarcts of the calvarium as well as secondary reactive sub-galeal collections. Treatment includes conservative management with intravenous fluids and analgesics, in addition, our patient was treated with antibiotics since differentiation clinically and by imaging can be challenging. Acute soft head syndrome should be considered in the differential diagnosis of headache and skull swelling in children with sickle cell anemia.

CASE REPORT

CASE REPORT

We present a 16-year old male patient known sickle cell anemia (HB-SS) at age of 6 months who presented to our emergency department with few hours onset of low-grade fever and headache. The headache was intractable and all over his head. He described it as pulsating and non-tolerable and could not be alleviated by analgesic. The patient did not report any history of head trauma. The patient was hemodynamically stable. On physical exam, the neurologic examination was unremarkable with normal motor power, sensations and reflexes. The medical team noticed a tender scalp swelling that was mostly located over the left hemisphere and right occipital region.

On investigation, laboratory studies showed: elevated white count (20400/mm, reference range: 4000-11000/mm), CRP (79.7mg/l, reference range: 0.0-2.5 mg/L), LDH (851IU/L reference range: <4.97, total Bilirubin (7 mg/dl reference range:0.0-1.2 mg/dl) and reticulocyte count (3.6, reference range: 4-6.5 mil/cu.mm). The hemoglobin and hematocrit levels were low (9.4 g/dl reference range:12-18 g/dl and 27% reference range: 37-54% respectively). His chest radiograph showed no acute cardiopulmonary process.

Two days after admission, non-enhanced MRI of the brain was performed and showed diffuse subgaleal collections with a predominant high T2 signal, iso to low T1 signal along with areas of high FLAIR and restricted diffusion signal in the frontal, bilateral parietal and occipital calvarium and small foci of susceptibility on T2* suggestive of punctate blood

products (Figure 1). The largest collection in the right occipital region measured 1.5 cm in maximum thickness. The underlying calvarium demonstrated high signal on FLAIR signal within the bone with areas of high signal on diffusion images in the regions of bone infarcts (Figure 1). No associated epidural hematomas or extra-axial collections. No cortical breaks or erosions were noted.

There are many differential diagnoses for headache and skull swelling in patients with sickle cell disease. In the setting of trauma, extra-axial hematomas would be the first consideration. With the lack of trauma history, differential diagnosis includes osteonecrosis, osteomyelitis with overlying abscesses/collections and finally ''acute soft head syndrome'' which is a diagnosis of exclusion.

The patient was admitted and started on intravenous fluid, broad spectrum antibiotics, analgesics and oxygen. The patient continued to improve clinically with significant improvement in his headache and resolution of his skull swellings, decrease in fever and normalization of his white count. Osteomyelitis was felt to be an unlikely possibility, however, could not be completely excluded and the patient was discharged on antibiotics including levofloxacin and clindamycin.

DISCUSSION

Sickle cell disease (SCD) is one of the most common hemoglobinopathies worldwide with a high incidence noted in African, afro-american, afro–Caribbean, Middle East and Indian descent [1]. The incidence of sickle cell disease is estimated globally between 300,000 and 400,000 neonates each year, the majority in sub-Saharan Africa [2]. The disease is caused by a mutation in the beta globin gene which results in an abnormally shaped red blood cell. The sickled, readily polymerized hemoglobin causes obstruction of the microcirculation. Patients with sickle cell disease suffer from diverse neurological manifestations.

Etiology & Demographics:

Acute soft head syndrome is an extremely rare complication in SCD and only few cases are reported in the literature [3,4] and the pathogenesis of this condition is not fully understood yet [3,5]. The most reasonable discussed mechanism is the fact that sickled RBC induces osteonecrosis and bone infarcts of the calvarium as well as cortical disruption and secondary reactive subgaleal collections [3]. Only nine cases reported associated epidural bleed that were attributed to the same pathophysiology described above but was not present in our case [5]. On the other hand, Berger et al concluded that unilateral skull swelling associated with fever and pain increases the possibility of osteomyelitis in sickle cell disease patients [6]. Although our patient had bilateral collections, he however also had fever and positive blood cultures for Granulicatella adiacens; and therefore, osteomyelitis could not be ruled out with certainty. Echocardiography performed to rule out endocarditis was negative.

Clinical & Imaging findings:

We present a case of acute soft head syndrome and the MR imaging findings in a patient with SCD who presented to the Emergency Department with severe headache, skull swelling and fever. This diagnosis of acute soft head syndrome in patients with sickle cell disease is a rare and challenging complication to diagnose [3]. Patients usually presents with headache and scalp swelling. It is a diagnosis of exclusion and needs to be differentiated from other common causes of headache in this population.

The cornerstone of diagnosis is imaging with MRI being the most sensitive modality for diagnosis [7]. CT has a limited sensitivity compared to MRI to detect these bone infarcts and is only able to demonstrate swelling and fluid [3]. Bone scan has a high sensitivity in detecting regions of osteonecrosis with photopenia surrounded by a rim of increased uptake, however, bone scans lacks the anatomic detail to delineate the extent of the abnormality and is also limited in assessing the associated soft tissue abnormalities [4]. MRI imaging features include high signal on T2 and FLAIR images, iso to low signal on T1, and high signal on diffusion within the calvarium. Within the soft tissues, there is extensive high FLAIR and T2 signal with possible associated foci of diffusion restriction in addition to susceptibility foci on T2* representing blood products. In addition to establishing a diagnosis, MRI is also important in these patients to evaluate for associated intracranial abnormalities such as intracranial extra-axial collections or bleed, which our patient did not have. It is also important to evaluate for other manifestations of sickle cell disease such as infarcts, silent ischemia, hemorrhage, and posterior reversible encephalopathy.

Treatment & Prognosis:

The management of this condition is usually conservative with IV hydration and analgesics and patients report significant improvement after few days. Commonly, as in our patient, treatment will also include broad spectrum antibiotics since differentiation clinically and by imaging can be challenging. Our patient was treated appropriately, and he was discharged home without complications. Follow up MRI one year later showed complete resolution of the extra-axial collections with persistent heterogeneous intramedullary high FLAIR signal sequelae of prior bone infarct (Figure 2).

Differential Diagnosis:

Acute soft head syndrome is a diagnosis of exclusion. It must be differentiated from osteomyelitis which needs to be treated aggressively with antibiotics in addition to the IV hydration and analgesic. Osteomyelitis show cortical destruction and periosteal reaction on computed tomography images. Post contrast MRI images show rim enhancement of the associated abscesses. Other differential diagnoses include epidural and subdural hematomas that can cause scalp swelling secondary to acute trauma. Associate fractures will tilt toward this diagnosis in the appropriate clinical context.

TEACHING POINT

The case presented illustrates a rare and benign complication seen in patients with sickle cell disease. The pathophysiology of acute soft head syndrome is related to osteonecrosis and bone infarcts of the calvarium as well as secondary reactive sub-galeal collections. The mainstay of treatment is conservative management of the vaso-occlusive crises with intravenous fluids and analgesics. Acute soft head syndrome should be considered in the differential diagnosis of headache and skull swelling in children with sickle cell anemia.

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FIGURES

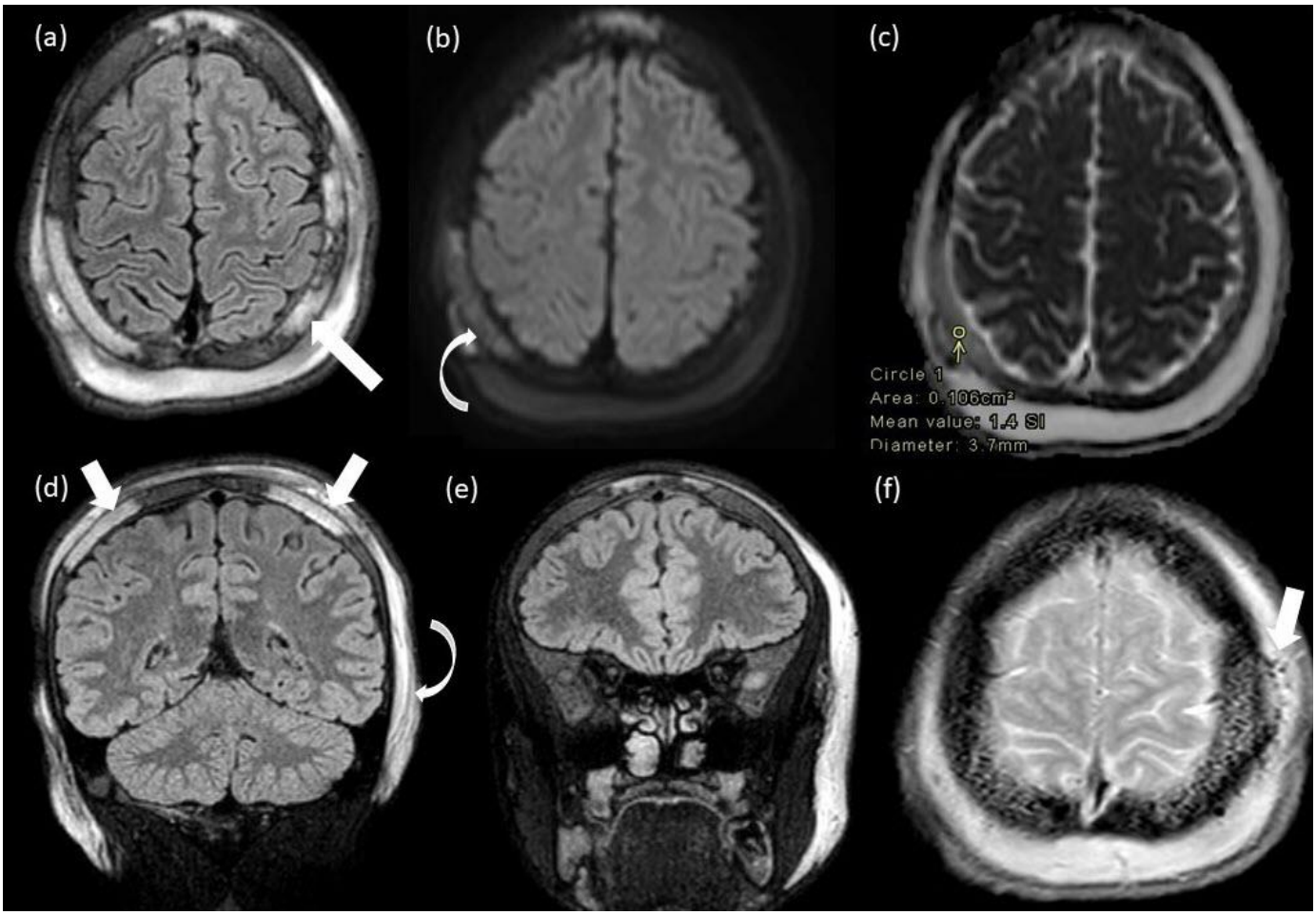

Figure 1: 16-year-old male patient with sickle cell disease and acute soft head syndrome.

Findings: (a) Axial FLAIR image near the level of the vertex showing bilateral high signal in the calvarium (white arrow). (b) Corresponding axial diffusion weighted image, (c) reconstructed ADC map showing high signal intensity within the calvarium (curved arrow) (d and e) Coronal FLAIR MRI showing extensive bilateral parietal hyper-intense signal in the calvarium (white arrow) suggestive of bone infarcts with associated extensive subgaleal collections and edema extending to the face and cheek (curved arrows). (f) Axial T2* images demonstrate punctate susceptibility foci within the subgaleal collections suggestive of associated blood products.

Technique: Non-enhanced study performed on 1.5 Tesla magnet (a) Axial FLAIR, 2mm slice thickness, TE:347.702, TR: 4800. (b) Axial diffusion with reconstructed ADC map, 5 mm slice thickness, TE: 105.406, TR: 3302. (c) and (d) Coronal FLAIR, 2mm slice thickness, TE:347.702, TR: 4800. (e) Axial T2*, 4 mm slice thickness, TE: 18.42, TR: 826.72.

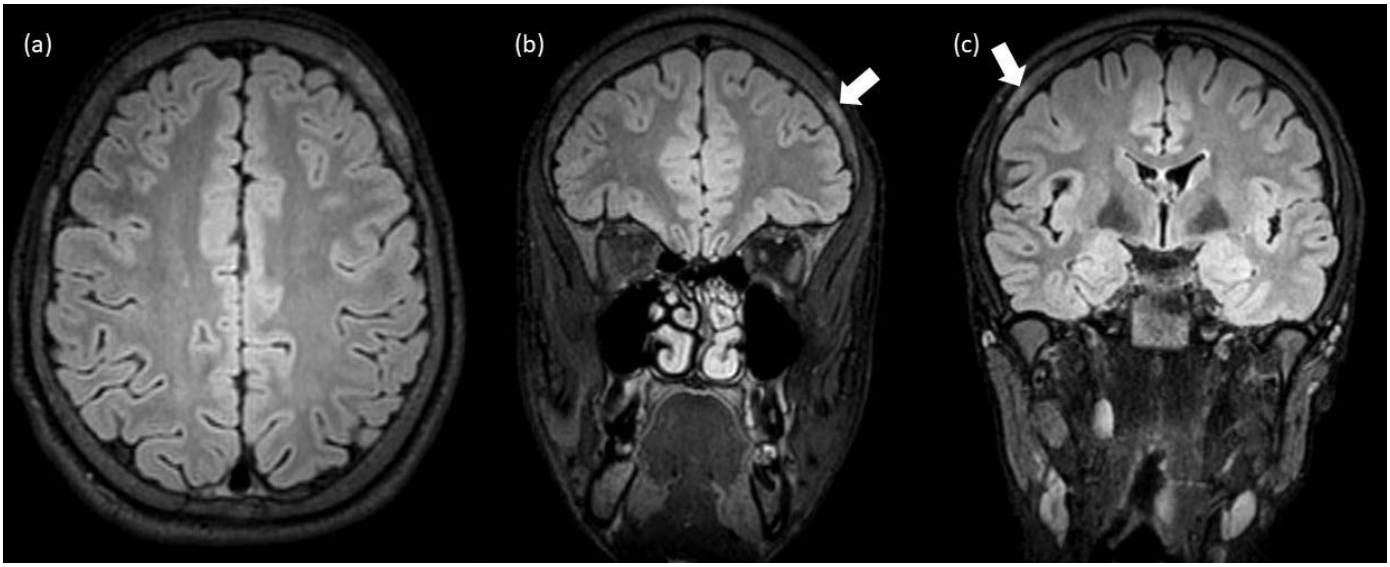

Figure 2: 16-year-old male patient with sickle cell disease and acute soft head syndrome. Follow up MRI one year after first MRI was performed.

Findings: (a) Axial FLAIR image near the level of the vertex showing resolution of the bilateral subgaleal collections. (b) and (c) Coronal FLAIR MRI showing resolution of the bilateral extra-axial collections with persistent heterogeneous intramedullary high FLAIR signal (arrows) corresponding to the areas of previous infarcts.

Technique: Non-enhanced study performed on 3 Tesla magnet (a) Axial FLAIR, 2mm slice thickness, TE:319.59, TR: 4800. (b) and (c) Coronal FLAIR 2mm slice thickness, TE:319.59, TR:4800.

Etiology	Osteonecrosis and bone infarcts as well as cortical disruption and secondary reactive subgaleal collections.
Incidence	Very rare, to our knowledge 6 cases were described in the literature.
Gender Ratio	None
Age predilection	None
Risk Factors	Sickle cell disease
Treatment	IV hydration and analgesics.
Prognosis	Good prognosis with significant improvement post treatment.
Findings on imaging	CT scan: swelling and fluid collections. Bone scan: photopenia surrounded by a rim of increased uptake. MR: high signal on T2 and FLAIR images, iso to low signal on T1, and high signal on diffusion within the calvarium.

Table 1: Summary table of acute soft head syndrome.

	Acute soft head syndrome	Osteomyelitis	Subdural and epidural hematomas in the setting of trauma.
X-Ray	-	A central area of radiolucency with a surrounding thick rim of reactive bone sclerosis.	Fracture lines.
US	Soft tissue edema is seen as areas of hypervascularity.	Limited use since it cannot assess bones. It can be useful for detecting soft tissue or subperiosteal collections which are seen as periosteal elevation with an underlying fluid collection.	Soft tissue edema is seen as areas of hypervascularity around the affected bone on color Doppler.
CT	Assessment of soft tissue swelling and fluid collections.	Shows osseous changes such as: 1. cortical destruction. periosteal reactions. 2. sequestrum formation. Poor soft tissue resolution.	Bone fractures. Assessment of soft tissue swelling and fluid collections.
MRI	1. High signal on T2 weighted and FLAIR images. 2. Iso to low signal on T1 weighted images. High signal on diffusion within the calvarium.	Bone marrow edema: 1. low signal on T1W images. 2. high signal on fluid-sensitive and post-contrast sequences Intraosseous and subperiosteal abscesses: 1. low signal on T1W images. 2. high signal on fluid-sensitive sequences. 3. peripheral enhancement on post contrast images. Sinus tract is seen as a linear fluid-filled structure extending from bone to the skin surface.	In the setting of trauma MRI is used to assess intraparenchymal contusions.
Pattern of contrast enhancement	No enhancement in the absence of superimposed infection.	1. Peripherally enhancing intraosseous lesion. 2. Non-enhancing sequestrum. 3. Sinus tract.	None
Scintigraphy	Osteonecrosis on bone scan shows photopenia surrounded by a rim of increased uptake.	In a triple-phase bone scan, technetium-99m-labelled MDP shows high tracer uptake in all three phases.	-
FDG-PET	-	Hypermetabolic activity in the affected tissues. Has the highest sensitivity in chronic osteomyelitis.	-

Table 2: Differential diagnosis table for acute soft head syndrome.

ABBREVIATIONS

CRP = C reactive protein
CT = Computed tomography
FLAIR = Fluid-attenuated inversion recovery
Hb-SS = Sickle cell anemia
LDH = Lactic Acid Dehydrogenase
MR = Magnetic Resonance
MRI = Magnetic Resonance Imaging
RBC = Red Blood Cells
SCD = Sickle cell disease

KEYWORDS

Acute soft head syndrome; sickle cell disease; osteonecrosis; osteomyelitis; headache

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