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Case Report

Acute Soft Head Syndrome as the Initial Presentation of Sickle Cell Disease in an Adolescent in Rural Western Uganda: Case Report

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Abstract

Sickle haemoglobin (HbS) is the most common abnormal haemoglobin mutation worldwide. In Uganda, according to the Uganda Sickle Surveillance Study (US3), the overall prevalence of sickle cell trait and disease is 13.3% and 0.7%, respectively. SCD presents with numerous complications. Acute Soft Head Syndrome (ASHS) or sickle cell cephalohematoma is among the rare complications of SCD. We report a 14-year-old adolescent male, a resident of rural Western Uganda, who presented with a history of non-traumatic painless scalp swelling for a week and multiple joints pain for 5 days. He had a similar swelling in the previous six months, which subsided after blood transfusion given at a nearby health facility. There was a history of on-and-off fever, easy fatiguability, and generalized body weakness. Complete blood counts revealed anemia, Haemoglobin Electrophoresis confirmed Sickle Cell Anemia. Head Computed Tomography concluded a subgaleal fluid collection. The diagnosis of acute soft head syndrome was established in a newly diagnosed SCD patient. This case highlights a rare diagnosis of acute soft head syndrome as well as a late diagnosis of SCD in adolescent in rural settings. In such settings, the pre-existing poor socio-economic background coupled with atypical presentation constitute a major impediment for timely diagnosis.

Keywords: Acute Soft Head Syndrome, Sickle cell disease, Rural.

INTRODUCTION

Sickle haemoglobin (HbS) is the most common abnormal haemoglobin mutation worldwide[1]. Uganda has one of the highest burdens of sickle cell anemia in the world[2]. According to Uganda Sickle Surveillance Study (US3) the overall prevalence of sickle cell trait prevalence of 13.3% and disease prevalence of 0.7% respectively[3]. SCD presents with numerous complications. A rare complication of SCD is Acute Soft Head Syndrome (ASHS) or sickle cell cephalohematoma, and few cases have been reported in East Africa.

This rare complication of SCD is thought to involve the expansion of intramedullary hematopoietic tissue, which disrupts both the inner and outer surfaces of the skull, potentially leading to the thinning of cortical bones[1]. Also, infarction of skull bones, along with cortical osteopenia, causes structural modifications to the bone and periosteum. These changes, coupled with the rupture of dilated periosteal blood vessels and necrosis of local vessel walls due to vaso-occlusion, result in the extravasation of blood and hematopoietic tissue into the subgaleal and epidural spaces[4]. This process underlies the non-traumatic scalp swelling characteristic of Acute Soft Head Syndrome.

Therefore, the purpose of this report is to raise awareness that nontraumatic scalp swelling is an uncommon complication of sickle cell disease and that it should not be managed with traditional practices or surgical excision.

CASE REPORT

We report a 14-year-old adolescent male not known to have sickle cell disease referred from a nearby health center to Kampala International

University Teaching Hospital in Ishaka – Bushenyi in March 2024. The patient was admitted through Accident and Emergency with a chief complaint of scalp swelling for a week and multiple joint pain for 5 days. The scalp swelling was diffuse, non-tender, and boggy, predominantly localized to the frontal and temporal regions of the head. Notably, there was no involvement of the underlying bone, and the swelling exhibited fluctuation upon palpation. The informant (mother) reported that the swelling had been progressively increasing in size over days. There was a history of headache for 1 day. There was no history of head trauma or injury. He had a similar swelling in the previous six months which subsided after blood transfusion given at a nearby health facility. The joint pain was more predominant in the knees and right elbow. There was a history of on-and-off fever, easy fatigability, and generalized body weakness but no dizziness or joint swelling was reported. The patient had paroxysms of mild dull generalized abdominal pain with normal bowel and micturition habits. However, no chest pain or difficulty in breathing. The patient had three siblings and neither the siblings nor the parents had a history of similar presentation or recurrent blood transfusion.

His initial physical examination revealed, alert and oriented patient. Severely pale conjunctiva, no jaundice, no palpable lymphadenopathy no lower limb edema. His scalp examination revealed a diffuse, boggy, non-tender, and soft swelling of the forehead and temporal part of the head. There was no periorbital involvement, pupils were equisized and reactive to light, and had no sign of meningism. His vital signs BP 110/80mmhg, pulse rate 98beat per minute, saturation 98% on rom air, respiratory rate 20 breath per minute, body temperature 36.5C. Abdominal examination showed splenomegaly (4cm below costal margin). Other systems where unremarkable.

His initial investigations showed severe anemia (HGB 6.1 g/dL), leucocytosis (38.09× 10^3 /uL), blood smear for malaria parasite not seen. He was managed with blood transfusion, IV fluids, antibiotics and analgesics. Later tests; Peripheral blood film showed anisocytosis, sickled red cells, pencil cells, and normochromic anemia. Capillary electrophoreses confirmed hemozygous haemoglobulin SS and recent transfusion due to presence of Hb A at 46.1%. S-CRP result 67.5mg/L (ref. 0.0—8.0). The diagnosis of newly diagnosed sickle cell disease with Vaso-occlusive crisis and acute soft head syndrome was established.

Table 1. Complete blood count showing normocytic normochromic anemia

	Results On admission	Reference ranges	Flags
WBC	38.09	$(4 - 10) \times 10^3$ /uL	High
RBC	2.43	$4.0 - 5.50 \times 10^{12}$ /uL	Low
HBG	6.1	(13 - 17) gm/dL	Low
HCT	20.95	(49 - 54) %	Low
MCV	78	(78 - 96) fL	Normal
MCH	30.5	(27 - 32) PG	Normal
MCHC	39.1	(30.5 – 35.5) g/dL	High
RDWs	94.3	(20.0-42.0) fl	High
RDWc	33.2	(11.6 - 14) %	High
PLT	285	$(150 - 40O) \times 10^3$ /uL	Normal

Table 2. Complete blood count on day 2 on review showing normocytic normochromic anemia

	Results	Reference ranges	Flags
WBC	27.62	$(4 - 10) \times 10^3$ /uL	High
RBC	2.68	$4.0 - 5.50 \times 10^{12}$ /uL	Low
HBG	7.0	(13 - 17) gm/dL	Low
HCT	18.64	(49 - 54) %	Low
MCV	77	(78 - 96) fL	Normal
MCH	28.9	(27 - 32) PG	Normal
MCHC	37.7	(30.5 – 35.5) g/dL	High
RDWs	97.6	(20.0-42.0) fl	High
RDWc	34.9	(11.6 - 14) %	High
PLT	248	$(150 - 40O) \times 10^3$ /uL	Normal

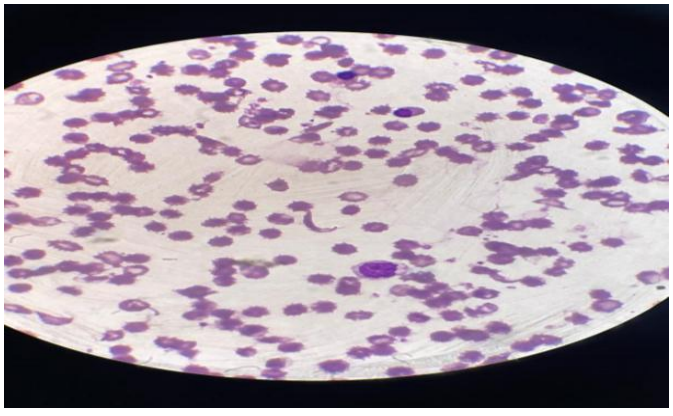

Figure 1. Peripheral film showing sickled red blood cells

Table 3. Haemoglobin electrophoresis (HB) showing predominant homozygous sickle cell disease

Haemoglobin A	46.1%3,0	
Haemoglobin A2	3.0	2.2- 3.2
Haemoglobin F	2.9%	
Haemoglobin S	48.1%	

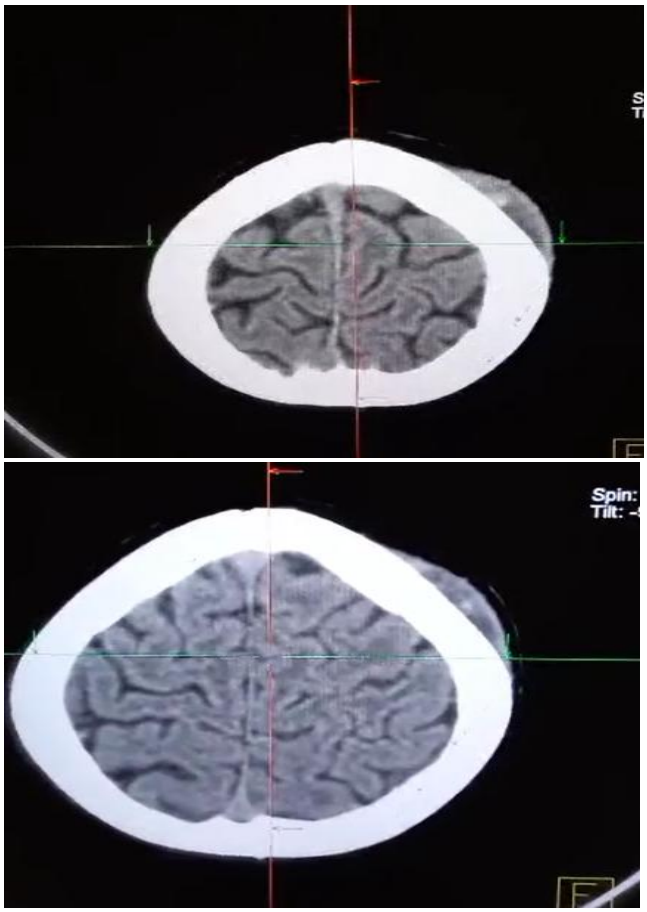

Figure 2. Head CT-Scan showing subgaleal fluid collection

One day post admission patient reports some improvement joint pain and chest pain has reduced. Post transfusion HGB 7g/dl and WBC 27.62× 10^3 /uL. On physical examination the patient was alert and moderate pale conjunctiva. His scalp examination revealed diffuse, boggy, tender, and soft swelling of the forehead and parietal part of the head although the swelling is reduced. There was no periorbital involvement. There no sign of meningism. His vital sings BP 105/80mmhg, pulse rate 82beat per minute, saturation 99% on rom air, respiratory rate 28 breath per minute, body temperature 36.2C. Abdominal examination showed splenomegaly (4cm below costal margin). Other systems where unremarkable. On the 4th day of post-admission we did non-contrast head CT scan showed subgalial fluid collection in frontal part of the head.

DISCUSSION

Acute soft head syndrome is a rare complication involving the skull in SCA, with few cases reported in the literature[4,5]. Acute soft head syndrome's precise pathogenesis is unknown, but theories include extramedullary hemopoiesis involvement of the skull's flat bones, bone infarction, osteopenia leading to cortical bone thinning, and disruption of the periosteal structure with blood leaking into the subgaleal space [4,6]. Acute soft head syndrome can be considered in deferential diagnosis and headache and scalp swelling in patients with sickle cell disease[7] The common acute soft head syndrome symptoms are headache and swelling [4,8]. Although our patient presented headache, fever, and unliteral head swelling of the scalp and forehead without periorbital involvement.

The differential diagnosis of sickle cell cephalohematoma includes trauma, osteomyelitis, bleeding diathesis, and skull hematoma. However, in our case, platelet count, prothrombin time, and partial thromboplastin test were not done due to financial constraints, also our patient had no recent history of head trauma. Although in our patient we didn't perform transcranial Doppler and central nervous system examination was normal this ruled out the possibility of cerebral ischemia. However, separating severe VOC from osteomyelitis in people with SCA may be challenging [6]. The likelihood of osteomyelitis was ruled out in the absence of fever, significant parietal bone tenderness, hyperemia, and no warmness surrounding the mass that may suggest severe acute inflammation and acute osteomyelitis are frequently linked. Due to clinical improvement and the accepted diagnosis, aspiration of the swelling to further describe it was discouraged.

Both magnetic resonance imaging (MRI) and computed tomography (CT) scans are valuable tools for the differential diagnosis of skull swellings in sickle cell anemia patients. However, the imaging findings for bone infarction are often limited to evidence of soft tissue swelling and fluid collections[8]. But MRI surpasses CT in sensitivity when it comes to detecting bone infarction.

A comparable case study from Nigeria described an 11-year-old patient with known sickle cell disease who developed head swelling two days

after admission and was diagnosed with Acute Soft Head Syndrome (ASHS). A comprehensive diagnostic evaluation, such as a clotting profile and a transcranial Doppler of the scalp, to rule out other potential causes of the swelling. The patient responded well to conservative management, with the swelling progressively resolved by the second week[4]. A similar case study from Saudi Arabia reported a 15-year-old male with sickle cell disease who developed Acute Soft Head Syndrome (ASHS) 48 hours after admission. Following a thorough investigation of imaging, including X-ray, CT, and MRI, the patient was managed conservatively with intravenous fluids and analgesia[7].

In spite of the further diagnostic techniques used in these other cases, the management across all three cases were similar, focusing on conservative methods such as intravenous fluids, analgesia, blood transfusion, and monitoring. The outcomes were also similar, with the swelling resolving progressively, which reinforces the effectiveness of conservative management even when advanced diagnostic tools are unavailable. Conservative and supportive therapy has been the key therapy in most of the cases.

In this case, the patient was managed conservatively with blood transfusions, antibiotics, analgesics, and intravenous fluids. A non-contrast head CT scan performed on the fourth-day post-admission revealed subgaleal fluid collection, confirming the diagnosis. The patient showed significant improvement in symptoms, including a reduction in head swelling, and was subsequently discharged. The patient was linked to the sickle cell clinic for future flow-up and also planned for follow after 1week in the medical OPD.

CONCLUSION

Poor socio-economic status and the atypical presentation of SCD create significant diagnostic challenges in rural settings. To address this, targeted CME programs that emphasize the importance of clinical diagnosis and conservative management of ASHS are crucial. Incorporating ASHS into SCD guidelines and strengthening primary care through newborn screening and community awareness can improve early diagnosis and outcomes. Conservative management, without the need for aspiration, remains effective in treating this uncommon complication, underscoring the need for awareness and appropriate clinical approaches.

Conflict of Interest

The authors declared that they have no conflict of interests.

Consent for publication

Informed consent was obtained from the patient's mother to publish this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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