



Acute Soft Skull Syndrome in an Adult Male with Sickle Cell Anemia in Sudan: A Case Report

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Abstract

Acute soft skull syndrome is an uncommon complication of patients with sickle cell anemia. Here, we report a case of an adult patient in Sudan with the acute soft syndrome, with our aim of providing more knowledge on this type of complication. The 20-year old patient, with a known history of sickle cell anemia, presented with a 1-day history of headache and joint pain. The complaint continued after admission, with increasing headache severity and development of rapid skull swelling, which indicated the rare sickle cell disease complication known as an acute soft head syndrome. Conservative management resulted in good response and rapid recovery of this case of acute soft skull syndrome with sickle cell anemia mainly related to skull infraction.

Introduction

Sickle cell disease (SCD)¹ includes a group of related genetic diseases affecting hemoglobin structure,² and is considered the most common genetic blood disorder among children.³ SCD frequently occurs in individuals whose ethnic origin is Africa,⁴ Middle East,⁵ Asia,⁶ Southern Europe,⁷ Central or South America,⁸ or the Caribbean.⁹ Sudan includes mosaic ethnic diversity that ranges from Afro-Arab and Arab to African tribal.⁴ This ethnic diversity includes groups with Negroid genetic features, with a recognized history in

areas such as Nilotes and Nuba.¹⁰

Acute skull soft syndrome is an uncommon complication of sickle cell anemia.¹¹ The pathogenesis and mechanism underlying the complication of acute skull soft syndrome is not fully understood. In addition, no information has been gained from the previous studies of acute skull soft syndrome for suffers in Sudan. The case of such presented herein is thus novel and of particular importance to Sudanese clinicians,¹² and we hope it will serve to increase knowledge and kindle awareness of this complication in Sudan.

Case Report

A 20-year old male with a known history of sickle cell anemia, diagnosed at the age of 2 years-old, came to our clinic at AlMawada (a private hospital) on May 18, 2019. The patient complained of headache and generalized joint pain that had begun 1 day prior to clinic presentation. The patient described the headache as constant, with mild frontal pain and not preceded by an aura nor associated or aggravated by anything, and partially relieved by paracetamol tablets. He denied any visual changes, vomiting, altered level of consciousness, neck pain, or variation of headache with positional changes or during the hours of the day. He also reported isolated joint pain involving wrists, knees and hips. The pain had severity score of 6/10 (according to self-judgment) and was dull in quality but not interfering with movement. There was moderate grade fever and the pain had intermittent association with nausea, but no

Keywords: Adult; Acute soft skull syndrome; Sickle cell anemia; Conservative management.

Abbreviations: CT, computed tomography; MRI, magnetic resonance imaging; SCD, sickle cell disease.

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Table 1. Lab investigations and findings for the patient

Date	Investigation	Result	Reference value
18/5/2019	Blood urea nitrogen	141 mg/dL	10–50 mg/dL
	Serum creatinine	18.5 mg/dL	0.7–1.4 mg/dL
	Serum Na+	139 nm/L	131–149 nm/L
	Serum K+	3.6 nm/L	3.1–5 nm/L
	Random blood glucose	141 mg/dL	79–140mg/dL
	Immunochromatographic (ICT) for malaria	Negative	
20/5/2019	Serum Na+	137 nm/L	131–149 nm/L
	Serum K+	3.6 nm/L	3.1–5 nm/L
21/5/2019	White blood cells	12.7 cells/cumm	4–10 cells/cumm
	Red blood cells	2.01 mill cells/cumm	3.8–5 mill cells/cumm
	Blood hemoglobin	6.5 g/dL	12.5–15.5 g/dL
	Mean corpuscular volume	98 fl	78–98 fl
	Platelets	98,000 cells/cumm	150,000–450,000 cells/cumm
	Blood urea nitrogen	25 mg/dL	15–45 mg/dL
	Creatinine	1.5 mg/dL	0.7–1.4 mg/dL

shivering or rigger. The patient also reported a burning sensation during micturition. There were no complaints involving any other organ systems.

The patient had a history of multiple previous hospital admissions, due to sickle cell crises. He had no history of diabetes mellitus, hypertension or any similar conditions, but he had been diagnosed 1 month previous with gallstone (“not operated”). The patient was on regular folic acid tablets, 5 mg daily, and not known to be allergic to any type of medication. On examination, the patient looked ill and in pain, and was pale and jaundiced. Vital signs were all within normal parameters, except for the body temperature at 38 °C. Blood pressure was 120/80 and heart rate was 70 beats/m. The patient was fully conscious and oriented when the central nervous system tests were performed. The Mini-Mental State Examination (commonly referred to as the ‘MMSE’) score was 30/30. All cranial nerves were intact, and Kernig’s and Brudzinski’s signs were negative.

Upper limbs showed no muscle wasting or fasciculations, and there was intact normal tone sensation; power was grade 5, and deep tendon reflex was +2 bilaterally. The lower limbs showed no muscle wasting or fasciculations, normal tone and intact sensation; power grade was 5, deep tendon reflex was +2 bilaterally, and gait was normal. Musculoskeletal examination of hands showed no swelling, tenderness, deformities, muscle wasting, skin rash or signs of dactylitis, and normal temperature and intact radial pulses. Phalen’s test was negative, with both knees and hips showing no swelling nor hotness but tenderness was present; the range of movement was normal. The cardiovascular examination also showed normal first and second heart sounds, with no murmurs or signs of heart failure. Clear breath sounds were heard bilaterally in chest examination. Results from blood sample testing for complete blood count, renal function and blood glucose are summarized in Table 1. Upon admission, the patient was immediately begun on intravenous normal saline (1 L/8 hours) with ciprofloxacin infusion (200 mg intravenously every 12 hours), tramadol tablets (once per day), paracetamol infusion (1 g/ 6 hours), and 4 U of packed red blood cells. On the third day of admission, the headache reached its maximum, being associated with scalp swelling and tenderness (Fig. 1). There was no loss of consciousness, visual change, projectile vomiting or neck stiffness. The patient denied history of head trauma, and stated he had never experienced such symptoms before. Again, the patient was negative for Kernig’s and Brudzinski’s signs. Brain

magnetic resonance imaging (MRI) was conducted and revealed no abnormalities (Fig. 2). The diagnosis was made clear as soft skull syndrome. The patient was treated with conservative management of intravenous normal saline (1 L/8 hours) and pethidine injection (50 mg twice every day). After two days, the patient fully recovered and was discharged.

Discussion

SCD is considered to be one of the most common causes of chronic anemia among pediatric patients of African descent.¹³ Chronic anemia, as in our case, leads to an increase in the demands of blood production and development of an extramedullary or intramedullary hematopoiesis.¹⁴ As such, chronic anemia is linked with elevated cardiac output and increased blood flow velocity, resulting in arterial tortuosity in many cases.^{15,16}

Acute soft skull syndrome is a rare manifestation of SCD, and the pathogenesis is not clearly understood, although it is believed to be related to infraction of the skull due to expansion of intramedullary hematopoietic tissues leading to disruption of the outer and inner skull margins and in turn resulting in softening of cortical bones.¹⁷ The differential diagnosis of a patient with acute soft head syndrome must include other causes of hematoma, such as trauma (which our patient had no history of), bleeding diathesis (in the present case, the platelet count and bleeding profile were within the normal range), and intracranial pressure (we excluded by the absence of any related features, such as altered level of consciousness, vomiting or visual changes). Clinical examination will also exclude any relative causes of skull swelling, such as lymph node enlargement due to any reason.

MRI and computed tomography (CT) scans of the skull have an important role in the differential diagnosis of sicker patients with acute soft swelling, as they are both able to detect bone infraction.¹⁸ MRI, However, is more sensitive than CT in detection of bone infraction.¹⁸ The presented patient underwent MRI and showed diploic space marrow signal changes consistent with known disease, which overall indicated an unremarkable finding. A contrast-enhanced MRI will demonstrate an area of heterogeneous and rim-like enhancement,¹⁸ and T2-weighted and short-inversion time inversion recovery MRI can detect areas of the high-



Fig. 1. During and after recovery from the acute attack of soft skull syndrome. (a) and (b) shows the patient with acute soft skull syndrome. (c) Shows the patient after recovery.

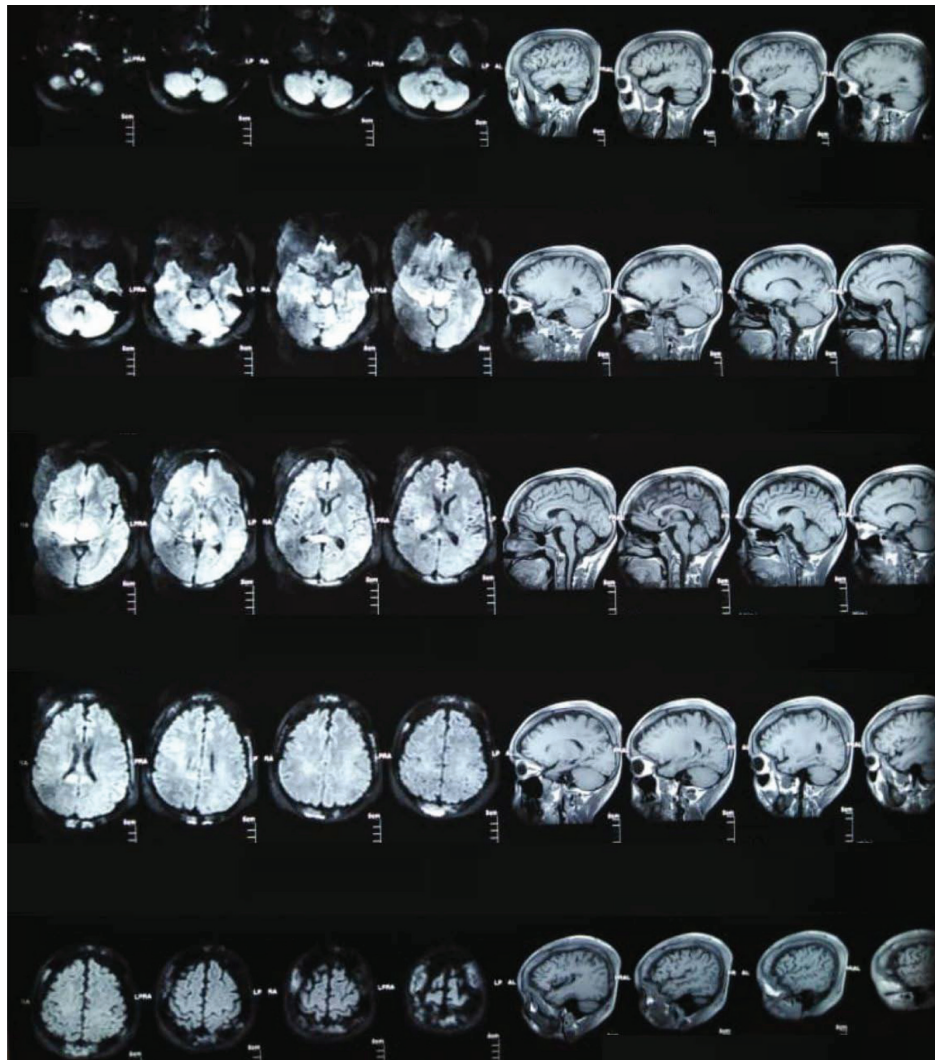


Fig. 2. Brain magnetic resonance imaging scans of the patient, indicating left maxillary polypoid sinusitis and diploic space marrow signal changes consistent with known disease. In conclusion: unremarkable study.

intensity bone marrow infarction and edema.¹⁸ Although, neither one of those investigations could be afforded by our patient.

Acute soft head syndrome due to sickle cell anemia is well known to be resolved by the same treatment of other sickle cell crises, that being conservative management (intravenous fluids and analgesics).¹⁸ Our patient did very well after receiving the intravenous fluids and tramadol.

In conclusion, this complication of SCD is uncommon but other reported cases along with ours serve to emphasize the need and desire for recognizing skull infarction, perhaps resulting in subperiosteal hematomas as causes of skull swellings and headache in sickle cell patients.

Patient consent

Consent was obtained from the patient.

Ethical approval

Ethical approval was obtained from Sudan Federal Ministry of health.

Financial disclosure

No funds or financial aid were received for this study.

Conflict of interest

All authors declare there are no conflicts of interest.

Author contributions

Case diagnosed and treatment supervision (ZIT), follow up (SEA, WME, SMEO), drafting of the manuscript (MEAE), analysis and interpretation (MMAH, HOA), critical revision of the manuscript for important intellectual content (MDY), study supervision (AAA).

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