ACUTE SOFT HEAD SYNDROME IN AN ADULT WITH SICKLE CELL ANAEMIA IN ABAKALIKI, SOUTH EASTERN NIGERIA.

UGWU Ngozi Immaculata[1], Emmanuel O. Ezeokoli[2], Anene N. Ekwe[2] Collins N. Ugwu[3], Ozoemena S. Oboke[4], Obalum I. Onyenweife[4] Samuel N. Esonwunne[4], Emmanuel Uchendu[4]

ABSTRACT

Acute soft head syndrome is a rare complication seen in patients with sickle cell anaemia (SCA). We report the case of a young male adult Nigerian and a known sickle cell patient who presented with complaints of headache, fever and multiple progressive swellings on his scalp, which were unrelated to any trauma events. Full clinical recovery, including complete resolution of the swellings on the scalp, occurred within a period of two weeks without any active intervention other than provision of analgesics and intravenous fluid for a few days at presentation. The pathophysiology of this syndrome is not fully understood but is thought to be related to vaso-occlusive crisis, leading to skull bone marrow infarction, and tearing of small vessels, with extravasation of blood into the subgaleal and epidural spaces. Acute soft head syndrome should be considered in the differential diagnosis of headache and scalp swellings in patients with sickle cell anemia.

Key words: Acute soft head syndrome, Cephalhaematoma, Nigeria, Sickle Cell Anaemia

INTRODUCTION

Sickle cell disease (SCD) is a hereditary hemoglobin disorder characterized by sickling of red blood cells under hypoxic conditions.[1] This is as a result of the presence of abnormal sickle haemoglobin known as haemoglobin S (HbS) which can be inherited in combination with other abnormal haemoglobin such as HbSC.[1] Haemoglobin S can also be inherited in homozygous state (HbSS) known as sickle cell anaemia and it is the commonest form of sickle cell disease.[2] High prevalence of SCD is found in Africa, Mediterranean region, South America, and East Asia, with Nigeria as the country with the highest burden of SCD in the world.[3]

Sickle cell anaemia is one of the most common single gene disorder in man with variable clinical manifestation. Acute soft head syndrome is a rare complication seen in patients with sickle cell anaemia. It consists of the triad of headache, fever and multiple swellings on the scalp.[4] There is paucity of report on acute soft head syndrome in adults with sickle cell anaemia in the literature. We report a case of an adult Nigerian male who is a known sickle cell anaemia patient and presented with headache, fever and painful scalp swellings due to cephalhematoma.

Case Report

GG is a 22 years old male, known sickle cell anaemia patient presented to our center with complaint of 2 day history of persistent headache, fever and generalized body pain. The patient described the headache as constant and not preceded by an aura. It was not aggravated by any known factor but partially relieved by paracetamol tablets. There was no associated history of visual changes, altered level of consciousness, vomiting or neck pain/ stiffness.

UGWU Ngozi Immaculata[1], Emmanuel O. Ezeokoli[2], Anene N. Ekwe[2] Collins N. Ugwu[3], Ozoemena S. Oboke[4], Obalum I. Onyenweife[4] Samuel N. Esonwunne[4], Emmanuel Uchendu[4]

Authors Affiliation:

[1]Department of Haematology and Immunology, Faculty of Basic Clinical Sciences, Ebonyi State University, Abakaliki, Nigeria. [2]Department of Haematology Alex Ekweme Federal University Teaching Hospital, Abakaliki, Nigeria. [3]Department of Internal Medicine, Faculty of Clinical Medicine, Ebonyi State University. [4]Department of Radiology, Alex Ekweme Federal University Teaching Hospital, Abakaliki, Nigeria.

Corresponding Author:
UGWU Ngozi Immaculata
(Corresponding author).
Department of Haematology & Immunology,
Faculty of Basic Clinical Sciences,
Ebonyi State University, Abakaliki, Nigeria.
Email: ngoziugwu5@gmail.com
Phone number: +2348061177100

Examination showed a young man in mild painful distress, warm to touch, mildly icteric, moderately pale, with no pedal edema and no peripheral lymphadenopathy. Evaluation of the scalp showed multiple swellings involving the parietal and frontal regions (Figure 1). The swellings, four in number with each located on both parietal and both frontal regions, measured about 8cm by 6cm by 3cm, tender and fluctuant. The skin overlying the swellings was normal. Central nervous system examination was unremarkable and no other abnormality was found.

A provisional diagnosis of malaria with vaso-occlusive crisis in a known SCA patient to rule out sepsis was made and he was admitted and placed on with analgesic (paracetamol infusion 1 g/ 6 hourly and intramuscular pentazosine 30mg 8 hourly), antimalaria (E-mal), systemic antibiotics (ciprofloxacin infusion 200 mg intravenously every 12 hours) and intravenous fluid (normal saline, 1liter/ 8hrly).

On the 3rd day while patient was on admission in the hospital, he also developed multiple scalp swellings. The swellings started on the left side of the head where one appeared on the left parietal region while the second one appeared on the left frontal region. They started regressing progressively over the next one week while another two swellings appeared on the right half of the scalp, one on the right parietal and frontal region each. Each of the swellings started initially as a pinhead sized lesion and later increased progressively in size to about an apple size. There was no associated history of trauma or head injury prior to the onset of the scalp swellings. There were no other swellings in any other part of the body. This was the first episode of scalp swellings ever reported by the patient.

Computed Tomography (CT) Scan of the head done initially due to complaint of persistent severe headache showed no abnormality except for an incidental finding of a polyp in the left maxillary antrum (Figure 2). Ultrasound scan of the scalp done later when the multiple swellings developed reported soft tissue swellings with blood collection in the swellings (Figure 3). Full blood count result showed haemoglobin value of 8.4g/dl, white cell count of 5.6 X 10^9/l and platelet count of 616 X10^9/l. Prothrombin time and partial thromboplastin time were normal. Based on his clinical presentation and results of radiological investigations, the diagnosis was subsequently changed to acute soft head syndrome.

While on admission, the scalp swellings progressively reduced in size. He was discharged home after 10 days of hospital admission to come back for follow-up. Physical examination during the follow-up visit 2 weeks after he was discharged revealed that the scalp swellings had completely resolved (Figure 4).

Figures

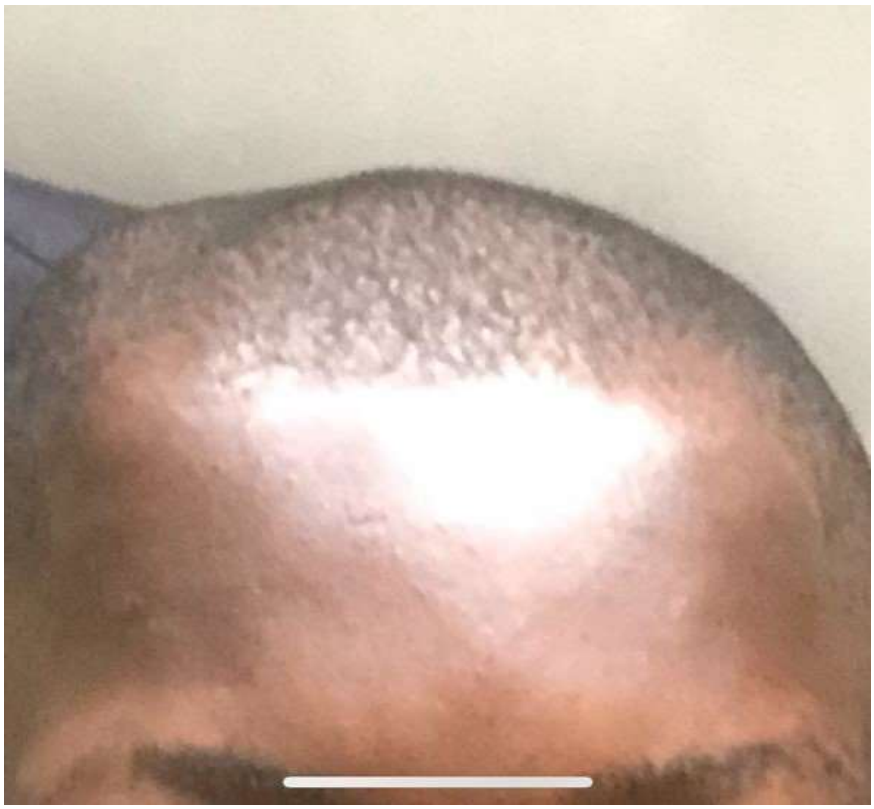

Figure 1: Swellings on the parietal and frontal regions bilaterally

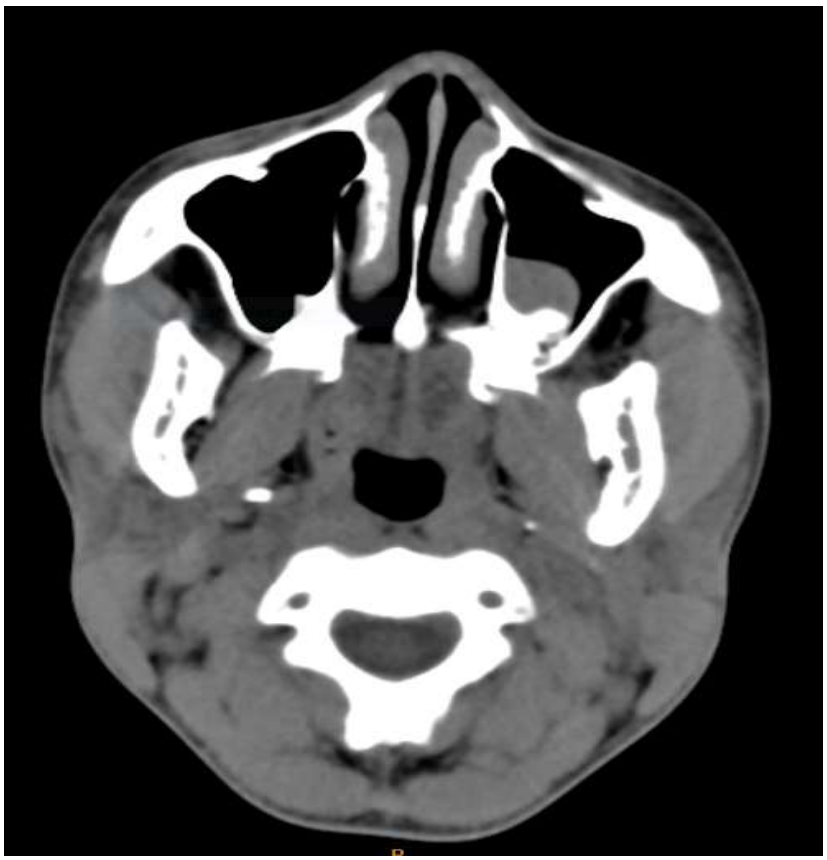

Figure 2: Polyp on the left maxillary antrum on CT Scan of the head

Figure 3: Ultrasound finding of soft tissue swelling and blood collection within the scalp swellings

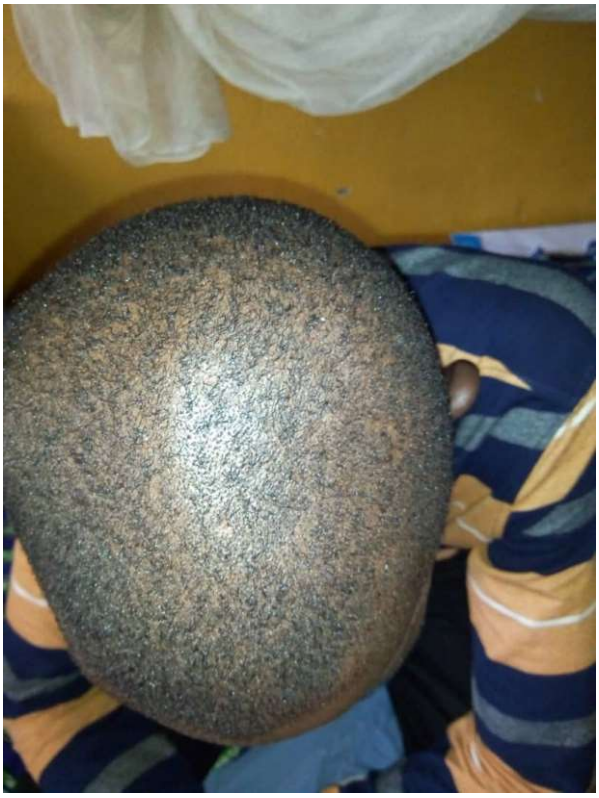

Figure 4: Scalp swellings completely resolved when patient came for follow up two weeks after discharge from the hospital

DISCUSSION

Sickle cell anaemia, characterized by homozygous presence of haemoglobin S, is a common cause of chronic anaemia particularly among people of African descent.[3] Chronic anaemia results to an increased need for haemaopoiesis which occurs both intra-medullary and extra-medullary. Due to increased need for blood, chronic anaemia is associated with increased cardiac output and high blood flow velocity, which may result to arterial tortuosity.[5]

Acute soft head syndrome is a rare manifestation of SCA but there are isolated case reports of patients who presented with headache and acute, rapidly progressive skull swelling.[6,7] It is made of a triad of headache, fever and scalp swelling (sickle cell cephalohematoma, or skull hematoma).[4] Scalp swelling can present as diffused or multiple localized swellings. Our patient presented with headache and fever initially and subsequently developed multiple swellings on the scalp involving the parietal

and frontal regions. The pathogenesis of this condition is not fully understood, but most likely related to vaso-occlusive crisis, leading to skull bone marrow infarction, and tearing of small vessels.[8] The process of cortical bone thinning is due to expansion of intramedullary hematopoietic tissue which results in disruption of inner and outer skull margins.[8] These altered bone and periosteal structures as well as local vessel wall necrosis due to vaso-occlusion precipitate non-traumatic extravasation of blood into the subgaleal and epidural spaces.

Manifestation of unexplained skull haematomas or sickle cell cephalohaematoma is a call for investigation for other causes of haematoma such as bleeding diathesis, trauma or raised intracranial pressure. The index case had normal prothrombin time and activated partial thromboplastin time. Patient also had thrombocytosis, which ordinarily would have been expected to be in favour of abnormal thrombus formation rather than abnormal bleeding. Patient had no associated history of trauma nor history suggestive of raised intracranial pressure such as altered level of consciousness, vomiting or visual changes.

A major cause of worry for patients with sickle cell anaemia presenting with severe, acute onset headache is cerebral infarction or hemorrhage. Parenchymal and subarachnoid hemorrhages due to aneurysmal rupture have also reported.[9] Stroke in patients with sickle cell disease has been associated with headache or even seizure.[9] Osteomyelitis with overlying abscesses is also a differential.[10] Computed Tomography (CT) scans of the brain and cerebrospinal fluid analysis are usually important to confirm the diagnosis of stroke or hemorrhage in the affected patients. The presence of scalp swellings, as seen in this patient, indicates involvement of extraparenchymal tissues, though both can rarely co-exist.[8] Computed Tomography (CT) scan done for the index patient on initial presentation with persistent headache without scalp swellings revealed no abnormality except an incidental finding of polyp in the left maxillary antrum. Whether this finding has any association with acute head syndrome is not known. However, subsequent ultrasound of the skull done when the swellings were noticed reported soft tissue swelling with blood collection within the swellings.

Previous studies have shown that most cases of acute soft head syndrome in patients with sickle cell anaemia resolve with conservative management which includes treatment of the vaso-occlusive crises with analgesics and intravenous fluids.[11,12] The index patient was managed conservatively and he completely recovered with resolution of the scalp swellings. Empiric use of broad-spectrum antibiotics as in this case is advisable because patients with sickle cell anaemia are susceptible to infection and it is often challenging to differentiate infection from infarction clinically.

Acute soft head syndrome can be managed conservatively and should be considered in the differential diagnosis of headache and scalp swellings in patients with sickle cell anemia.

Ethics approval: Not applicable.

Consent for publication: Written informed consent was obtained from the patient for publication of this article and any accompanying images.

Conflict of interest: Nil

REFERENCES

1. Heish MM, Tisdale JF, Rodgers GP. Thalassaemias and Sickle cell Disorders. In: Rodgers GP, Young NS (editors). Bethsda handbook of Clinical Hematology. 2nd edition. Philadelphia, Lippincott Williams and Wilkins 2010: p 35 – 55.

2. Isa HA, Adegoke SA, Madu AJ, Abdul-Aziz H, Ohiaeri CN, Chianumba R, et al. Sickle Cell Disease Clinical Phenotypes in Nigeria: A preliminary analysis of the Sickle pan Africa Research consortium (SPARCO) Nigeria Database. Blood cells, Molecules and

Diseases 2020; 84: 102438.

3. World Health Organisation. Sickle cell anaemia report by the secretariat. Fifty ninth world health assembly A59/9, 2006.

4. Okunade MA, Okpala IE, Olatunji PO, Shokumbi WA, Aken'nova YA, Ogunsanwo BA. A review of cephalhaematoma in adult sickle cell disease patients. Afr J Med Med Sci 1994 Sep;23(3):253-255.

5. Gladwin MT, Sachdev V. Cardiovascular abnormalities in sickle cell disease. J Am Coll Cardiol. 2012; 59(13):1123-1133.

6. Akodu SO, Njokanma OF, Diaku-Akinwumi, Ubuane PO, Adejiji UO. Acute Soft Head Syndrome in Children with Sickle Cell Anaemia in Lagos, Nigeria. Indian J Hematol Blood Transfus 2014; 30(Suppl 1):S67–S69.

7. Zedah C, Rameh V, Atweh LA. Acute soft head syndrome in a sickle cell disease patient. Journal of Radiology Case Reports 2021; 15(4):1-6.

8. Ejindu VC, Hine AL, Mashayekhi M, Shorvon PJ, Misra RR. Musculoskeletal manifestations of sickle cell disease. Radiographics 2007; 27(4): 1005–1021.

9. Kamath SD, Pai MG. A case series of hemorrhagic neurological complications of sickle cell disease: Multiple faces of an underestimated problem! Int J Med Sci Public Health 2020;9(6):386-391.

10. Yolande D, Tufong K, Jules T, Mayah A, Charlotte E, Njinkui D, et al. (2021) Osteomyelitis in Children with Sickle Cell Disease: A Challenging Diagnosis: Case Report from Cameroon. Open Journal of Pediatrics 2021; 11: 208-214.

11. Taha ZI, Mohammed SE, Essa MEA, Elsid WM, Hussein MMA, Osman SME, et al Acute Soft Skull Syndrome in an Adult Male with Sickle Cell Anemia in Sudan: A Case Report. Exploratory Research and Hypothesis in Medicine 2019; 4(4): 90–93.

12. Al-Zahir ZZ, Al- Hajjaj A. Acute Soft Head Syndrome in Sickle Cell Disease in Qatif Central Hospital, Saudi Arabia—Case Report and Review of the Literature. Open Journal of Internal Medicine 2020; 10: 135-140.