Acute Soft Head Syndrome in Children with Sick Cell Anaemia in Lagos, Nigeria

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Acute soft head syndrome is rare complications seen in children with sickle cell anaemia. A case report of a child with sickle cell anaemia who developed acute soft head syndrome. A 12-year old known sickle cell anaemia patient presented with acute, rapidly progressive skull pain and swelling, manifestations indicative of the rare complication of SCD which is called acute soft head syndrome. Conservative treatment with intravenous fluids and analgesics and empirical use of broad-spectrum antibiotics resulted in recovery. Acute soft head syndrome is a rare complication in children with sickle cell anaemia probably related to skull infarction. It further draws attention to the importance of acute soft head syndrome as a differential to be considered for pains in the head and skull swellings in children with sickle cell anaemia.

Keywords Children · Sick cell anaemia · Soft head · Acute · Syndrome

Introduction

Sickle cell anaemia is one of the commonest single gene disorders in man with variable geographical distribution and variable clinical manifestations [1]. Acute soft head syndrome is a rare complication seen in children with sickle cell anaemia [2]. Acute soft head syndrome is also known as cephal-haematoma or skull haematomas. To the best of the authors' knowledge, there is paucity of report of soft head syndrome in

Nigerian children with sickle cell anaemia. We report a case of soft head syndrome in a child with sickle cell anaemia. It is believed that this information will add to available data and rekindle awareness as far as this rare complication in children with sickle cell anaemia is concerned.

Case Report

AT is a 12 year-old boy with sickle cell anaemia diagnosed at age 2 years of life. He was referred to our facility with 4 days history of fever and generalised body pains which started from the back before spreading to other body parts including upper and lower limbs. 3 days after the onset of fever and generalised body pains he developed scalp swellings. The swellings located in both parietal regions and also the frontal region started initially as a pinhead-sized lesion and later increased progressively in size to about an apple-size. There was no history of prior head injury. Also, there were no swellings in any other body parts. This was the first episode of scalp swellings reported by the parents and patient.

Examination showed sickle cell stigmata, pallor and jaundice. The scalp evaluation revealed multiple swellings involving the frontal and parietal regions (Fig. 1). The swellings measured 5.0 × 6.0 cm each and were tender, mobile and fluctuant. The skin overlying the swellings was normal. The examination of the central nervous system was essentially normal. An initial diagnosis of multiple scalp abscesses with vaso-occlusive crisis in a child with sickle cell anaemia was made and he was admitted and placed on systemic antibiotics (erythromycin and chloramphenicol) with analgesic (ibuprofen) and intravenous fluid infusion.

The skull radiography showed normal findings in the bones. Brain computerized tomographic scan to exclude

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Fig. 1 Showing the head on admission

intracranial extension of the lesion was not done due to financial constraints. The clotting profile was normal. The packed cell volume was 21.4 % while other components of the complete blood count results were within normal limits. An aspirate from the swellings was haemorrhagic and did not clot in the syringe after 5–10 min. The aspirate did not yield any pathogen. At this point, a diagnosis of acute soft head syndrome was made.

While on admission, the scalp swellings progressively reduced in size. He was discharged home after 11 days of hospital admission to come back for follow-up. Physical examination during the follow-up visit 2 weeks after discharged home revealed that the scalp swellings had completely resolved (Fig. 2).

Discussion

Sickle cell anaemia, characterized by homozygous presence of haemoglobin S, is a common cause of chronic anaemia among children of African descent [1]. In this condition, chronic anaemia leads to an increased need for blood production and results in intra-medullary or extra-medullary haematopoiesis [2]. Because of this increased need for blood, chronic anaemia is associated with increased cardiac output and high blood flow velocity, and it may lead to arterial tortuosity [3].

Acute soft head syndrome is an uncommon manifestation of sickle cell disease but there are isolated case reports of children who presented with acute, rapidly progressive skull pain and swelling [2, 4]. The pathogenesis of this condition is not fully understood, but probably it is related to the skull infarction. It is thought that the expansion of intramedullary haematopoietic tissue results in the disruption of the inner and outer skull margins which may lead to

Fig. 2 Showing the head 2 weeks after discharge

thinning of cortical bones. These altered bone and periosteal structures as well as local vessel wall necrosis due to vaso-occlusion precipitate non-traumatic extravasation of blood and haematopoietic tissue into the subgaleal and epidural spaces [5].

The differential diagnosis in a child with haematoma must include bleeding diathesis. In the index case however, platelet count, prothrombin time and partial thromboplastin test were all within normal limits. The steady state packed cell volume of the patient herein reported was not known but is likely to have been higher than 21 % observed on admission. However, the patient remained stable even without blood transfusion.

Computed tomography (CT) scan and magnetic resonance imaging (MRI) scan have significant roles in the differential diagnosis of the sickle cell anaemia patient with skull swellings. The CT and MRI findings in bone infarction are limited to soft tissue swelling and fluid collections. In bone infarction, disruption of normal trabecular architecture is initially seen at CT but difficult to detect [6]. MRI is much more sensitive than CT for detection of bone infarction [6]. At T2-weighted and short inversion time inversion-recovery MRI, areas of high signal intensity represent bone marrow edema and infarction [6]. At contrast material-enhanced MRI, areas of infarction demonstrate heterogeneous, rim-like enhancement [6]. However, neither investigation could be done in the reported case due to financial constraints.

Most cases of acute soft head syndrome in children with sickle cell disease resolve with conservative management that includes treatment of the vaso-occlusive crises with intravenous fluids and analgesics [6]. Our patient received this conservative management and did well. Empiric uses of broad-spectrum antibiotics as in the reported case is advisable because patients with sickle cell disease are susceptible to infection and it is often difficult to clinically differentiate infarction from infection [7].

In conclusion, acute soft head syndrome is a rare complication in children with sickle cell anaemia. Our case and others emphasize the need to recognize skull infarction, possibly resulting in subperiosteal haematomas, as possible causes of headache and skull swellings in sickle cell anaemia patients.

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