

## Non-traumatic head swellings in a child with a sickle cell disease presented to a tertiary hospital North of Tanzania: A case report

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### Abstract

Sickle cell is the commonest inherited haemoglobin disorder in Sub-Saharan Africa. Often patients present with painful crises in the extremities. But rarely non-traumatic Sub-galeal hematoma whose pathogenesis is not clearly defined, presenting with scalp swellings can occur. We present a case, nine years old male, with this uncommon complication which is also known as Acute Soft Head Syndrome and was managed conventionally at a tertiary hospital in Tanzania with hydration, analgesics, blood transfusion and antibiotics with marked improvement within a week.

### Keywords:

Sickle Cell Disease, Sub-galeal Hematoma, Acute Soft Head Syndrome, Bone Infarction

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### Background

Sickle cell disease is a group of inherited blood disorders which occur due to a mutation affecting the gene encoding for a  $\beta$ -globulin chain of haemoglobin and it makes red blood cells change into crescent shapes in hypoxic conditions due to polymerisation. It is a disease that is more prevalent in Africa and among African American descent (Ware *et al.*, 2017). It is the main cause of chronic anaemia in children and it is associated with other chronic or recurrent complications like pain due to acute vaso-occlusive crises, infections and organ damage (Fortin-Boudreault and Story, 2016). Sub-galeal hematoma is a rare complication to occur in Sickle Cell Disease (SCD), presenting with soft and boggy swellings on the head. This condition is also termed Acute Soft Head Syndrome with little known and documented about it. In this report, we present a patient who was admitted with such a rare complication to the paediatric unit of Kilimanjaro Christian Medical Centre, a zonal Referral Hospital, in northern Tanzania. We believe that this will add to the current knowledge available and create awareness of the condition. Ethical approval to conduct and organize the case report was granted by the Kilimanjaro Christian Medical University college's clinical research and ethics committee. Both assent and consent were obtained from the child and parent, respectively.

### Case report

ARO, the 9-year-old male, known patient with SCD since the age of 2 years, only on regular Folic acid medication since then, presented to the hospital as a referral case with the complaints of painful swelling on the left forearm and left ankle for one week and head swellings for four days. Initial symptoms started spontaneously and gradually progressed, with no history of any trauma, and was no associated fever. The patient was not able to use the affected limbs due to pain. Four days before admission, spontaneous gradual onset of head swellings ensued and was progressive. The swellings were not associated with pain and the patient reported no history of headache, convulsions, or loss of

consciousness. During the illness, the patient was given various oral and Intravenous (I/V) medications to relieve the pain and swelling at the nearby hospital until the head swellings started, rendering a referral to our facility. Medically in the past, the patient has been admitted several times due to complaints of pain and swellings on the limbs and signs and symptoms of anaemia. He has been transfused several times as well. This is the only child in the family.

On examination, he was alert with jaundice and severe pallor with stable vitals. On the scalp, two biparietal swellings, measuring 6 cm × 7 cm on the right and 4 cm × 5 cm on the left were noted (Figure 1). These were soft, boggy, non-tender and fluctuant with normal skin overlaying them. No neurological deficit was noted on Central Nervous System (CNS) examination. There was tender swelling on the left forearm, with normal skin temperature over the area, just as the swelling on the lateral malleolus of the left ankle. Provisionally we had the impression of SCD with vaso-occlusive crisis to rule out Osteomyelitis.

Figure 1:



*Right head parietal swelling, 3 days post admission with the left one subsided almost completely*

*Right head parietal swelling, 3 days post admission with the left one subsided almost completely*

The patient underwent skull X-ray which had no abnormality and an Ultrasound Scan of the head swellings showed extra periosteal fluid collections, anechoic with minimal floating strands. A computed Tomography Scan or Magnetic Resonance Imaging was not done due to financial constraints. The full blood count showed mild leucocytosis with neutrophilia with an Erythrocyte Sedimentation Rate (ESR) of 21mm/hr. The patient had a haemoglobin level of 5.9 g/dl with a Haematocrit of 20.8%. Bleeding indices were within the normal range.

The patient was hydrated with I/V and oral fluids and was given Ibuprofen, together with a single blood transfusion. Antibiotics were administered for five days due to suspicion of a possible infection. The patient stayed in the ward for six days by which time left head swellings had disappeared

completely while the right one had subsided remarkably. Control Haemoglobin at the time of discharge was 7.8 g/dl.

### Discussion

Africa has the highest burden of sickle cell disease, with up to 75% of worldwide births with SCD occurring per year with Tanzania among the top five countries with the highest number of annual births with SCD in the world. The majority of the patients presenting at the hospital are between the ages of 5 - 17 years with various complications (Makani *et al.*, 2018). Pain is the main presenting symptom necessitating a hospital visit. Head swellings due to Sub-galeal hematoma also known as Acute Soft Head Syndrome is one of the rare complications in SCD patients presenting with single or multiple boggy swellings on the head (Hanafy *et al.*, 2019). Not so much of the condition is known and the majority of the literature has been limited to case reports.

Pathogenesis of Acute Soft Head Syndrome is not well understood but several theories have been postulated. Mainly this is secondary to thinning of the skull bone cortex due to chronic infarction occurring after repeated occlusion of its vessels. In addition, vessel wall necrosis along this area can cause non-traumatic bleeding to occur (Akodu *et al.*, 2014). Another postulated mechanism is the extravasation of the overactive marrow of SCD patients through the weakened bone cortex to the outside (Hanafy *et al.*, 2019).

If the bleeding/extravasation occurs on the outer side of the skull in the Sub-galeal space, the presenting feature will be scalp swellings, like in this case and when the bleeding is on the inner side of the skull in the epidural space, the patient may present with various neurological symptoms of epidural hematoma which is another rare, non-traumatic, life-threatening secondary complication. MRI is the best radiological investigation compared to CT-scan, for recognition of bone infarction in these conditions (Saito *et al.*, 2010). But unfortunately, due to financial constraints, this was not the case. Ultrasonography of the swellings instead, was very helpful, showing the extra-periosteal fluid collection, a report which was consistent with our diagnosis of non-traumatic Sub-galeal hematoma due to Acute Soft Head Syndrome. Aspiration of the fluid was discouraged and was not done due to the fear of introducing infections.

One of the important differential diagnoses which should be considered is osteomyelitis. It is difficult to distinguish the two entities clinically because they can both present the same swelling, pain and fever. In a low-resource setting this can be ruled out by x-ray of the affected site or ultrasonography since it has a sensitivity of up to 74% when showing periosteal elevation with the accumulated subperiosteal fluid of 4mm or more. Our patient skull x-ray did not show any feature of osteomyelitis neither did the ultrasonography of the swelling on the scalp. Bleeding diathesis is another important differential diagnosis to consider. The platelet count was within the normal range on a full blood picture in our patient (Fortin-Boudreault and Story, 2016).

It has been reported that acute soft head syndrome is managed conservatively with no specific treatment (Fortin-Boudreault and Story, 2016). We hydrated our patient adequately and gave analgesics. Transfusion was given once since the Haemoglobin level was very low (5.5 g/dl). Antibiotics are given upon proving or suspecting an infective process from the history, physical findings or investigations as SCD patients are prone to get bacterial infections due to functional asplenia. The management of this complication can be done successfully even in a primary health care facility with complete remission occurring within a few days using minimal resources as in this case. Awareness among clinicians and the community at large about early diagnosis and management is of paramount importance.

Acute soft head syndrome is one of the uncommon complications in children with SCD, presenting with single or multiple boggy swellings on the head. It is a condition which is managed conservatively. Osteomyelitis is an important differential diagnosis which needs to be ruled out.

### **Conflict of interest**

None declared

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### **References**

- Akodu, S. O., Njokanma, O. F., Diaku-Akinwumi, I. N., Ubuane, P. O. and Adediji, U. O. (2014) Acute soft head syndrome in children with sickle cell anaemia in Lagos, Nigeria., *Indian Journal of Hematology & Blood Transfusion : An Official Journal of Indian Society of Hematology and Blood Transfusion*, 30 (Suppl 1), pp. 67–9. DOI:10.1007/s12288-013-0251-6.
- Fortin-Boudreault, R.-P. and Story, E. (2016) Spontaneous Subgaleal Hematoma: An Unusual Complication of Sickle Cell Disease, *Journal of Clinical Case Reports*, 06 (01), pp. 1–2. DOI:10.4172/2165-7920.1000681.
- Hanafy, E., Amri, S. Al, Alenazi, A., Balawi, A. Al and Abdullah, N. (2019) Acute soft head syndrome and a mini review of bone and neurologic complications in patients with sickle cell disease, *International Journal of Case Reports*, pp. 1–9. DOI:10.28933/ijcr-2019-06-2606.
- Makani, J., Tluway, F., Makubi, A., Soka, D., Nkya, S., Sangeda, R., et al. (2018) A ten year review of the sickle cell program in Muhimbili National Hospital, Tanzania., *BMC Hematology*, 18 (1), pp. 33. DOI:10.1186/s12878-018-0125-0.
- Saito, N., Nadgir, R. N., Flower, E. N. and Sakai, O. (2010) Clinical and radiologic manifestations of sickle cell disease in the head and neck., *Radiographics : A Review Publication of the Radiological Society of North America, Inc*, 30 (4), pp. 1021–34. DOI:10.1148/rg.304095171.
- Ware, R. E., de Montalembert, M., Tshilolo, L. and Abboud, M. R. (2017) Sickle cell disease., *Lancet (London, England)*, 390 (10091), pp. 311–323. DOI:10.1016/S0140-6736(17)30193-9.