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Case Report

Beyond the Usual Crises: Acute Soft Head Syndrome in Paediatric Sickle Cell Anaemia

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ABSTRACT

There is very little information on the global occurrence and clinical features of acute soft head syndrome, as it is an incredibly rare condition. Subgaleal hematoma (SGH) represents an uncommon, yet significant complication in adolescents with sickle cell anaemia (SCA).

Here, we present a case of a 16-year-old with sickle cell disease who developed a spontaneous SGH. This report highlights the diagnostic challenges and management strategies associated with this rare occurrence and reviews recent literature in order to enhance understanding of SGH within the context of SCA.

Keywords: Acute soft head syndrome, subgaleal haematoma, sickle cell anaemia, periorbital swelling

Introduction

Subgaleal hematoma (SGH) represents a rare, yet significant complication in adolescents with sickle cell anaemia (SCA). This condition is marked by bleeding into the subgaleal space between the galea aponeurotica and the periosteum of the skull (1). Adolescents with SCA are particularly susceptible due to their frequent vaso-occlusive crises and bone infarctions, predisposing them to spontaneous bleeding events (2).

There is very little information on the global occurrence and clinical features of acute soft head syndrome (ASHS), as it is an incredibly rare condition. The lack of literature and the co-occurrence or overlap with other sickle cell disease (SCD)-related diseases, such as extramedullary haematopoiesis, make diagnosis especially difficult (3).

Here, we present the case of a 16-year-old with SCA who developed spontaneous SGH. This report highlights the diagnostic challenges and management strategies associated with this rare occurrence and reviews the recent literature in order to enhance understanding of SGH within the context of SCA.

Case Report

A 16-year-old male, the fifth child of a non-consanguineous marriage, presented with intermittent fever spikes, and a yellowish discoloration of the eyes lasting for 15 days. He also had had intermittent pain in his knee joint for a week (pain scale 4/10). Low grade fever was present accompanied with inter-febrile normal periods. The alarming symptom was the sudden onset of a painless swelling on the left side of his scalp over the

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temporoparietal area noticed by the child over 3-4 days which then progressed to right-sided periorbital swelling 2 days later. There was no history of loss or blurring of vision. There were no complaints of redness, pain in the eyes, floaters or flashes of light. There was no history of recent trauma to the head.

He had been taking hydroxyurea and folic acid for the management of SCA. His past medical history included a single episode of vaso-occlusive crisis requiring admission to the hospital. He had received erythrocyte transfusion only once 8 prior. The patient had poor compliance and had not gone for any regular follow-ups.

On examination he was conscious and well-oriented to time, place and person with a Glasgow Coma Scale of 15/15. He had severe pallor and deep icterus, vitally, a fever of 39 °C, tachycardia (112 beats/min.), and blood pressure of 104/64 mm Hg with an oxygen saturation of 99% in room air. His bilateral pupils were normal size and reacting to light normally. There were no signs of increased intracranial pressure or meningeal irritation.

Systemic examination revealed a mild splenomegaly (soft and non-tender) and a hemic murmur (a sign of severe anaemia). Central nervous system examination was within the normal limits. Local examination revealed a boggy swelling on the left parietotemporal region of the scalp, with ill-defined margins, fluctuant, transillumination negative, non-tender with normal overlying skin. There was also right-sided periorbital swelling with no signs of inflammation.

The initial differential diagnoses were retinal haemorrhage, central retinal arterial occlusion, or carotid artery infarcts/stroke.

Laboratory investigations revealed anaemia [haemoglobin (Hb): 7.00 g/dL, mean corpuscular volume 98 femtolitre with thrombocytopenia 73,000 x109/L, absolute neutrophil count: 3,600 x109/L with bilirubin (total bilirubin: 5.1 mg/dL; indirect bilirubin: 3.6 mg/dL with serum glutamic oxaloacetic transaminase: 190 IU/L, serum glutamic pyruvic transaminase: 76 IU/L). The Gruber-Widal test was positive. Blood culture was negative. Serum lactate dehydrogenase was 2,487 U/L, serum homocysteine was 48.74 µmol/L, erythrocyte sedimentation rate was 38 mm/hour, and coagulation profile was normal. Hydroxyurea was discontinued due to pancytopenia.

An ophthalmology opinion was taken regarding the periorbital and scalp swelling in order to rule out central retinal arterial occlusion. Fundus examination, bilateral pupillary reflexes and extraocular movements were all normal. A carotid Doppler was performed to rule out any atherosclerotic plaque disrupting the flow to the retinal vessels which were revealed to be normal. Two-dimensional echocardiography was carried out in order to check pulmonary pressures and thrombus, and these revealed normal findings.

Magnetic resonance imaging (MRI) brain with contrast (Figure 1) revealed skull bone infarcts in the bilateral parietal bone, haematoma in the right orbital (extraconal) along with a temporal area measuring 1.4x0.5 cm and right-sided subperiosteal haematoma (Figure 2). The haematoma had displaced the superior rectus muscle inferomedially with a size of 6.5x1.1 cm in the right parietal region.

A neurosurgery opinion was taken and conservative management of the scalp and periorbital swelling including cold fomentation, compression, and anti-inflammatory medications were recommended by the neurosurgeon in order to provide relief without the need for invasive interventions.

During hospitalization, the patient received symptomatic treatment for fever and joint pain along with ceftriaxone. He became symptomatically better over a week and there was a remarkable reduction in the periorbital swelling but the scalp swelling showed only mild reduction. The neurosurgeon suggested that it would take time to decrease in size. The patient was discharged and called for follow-up

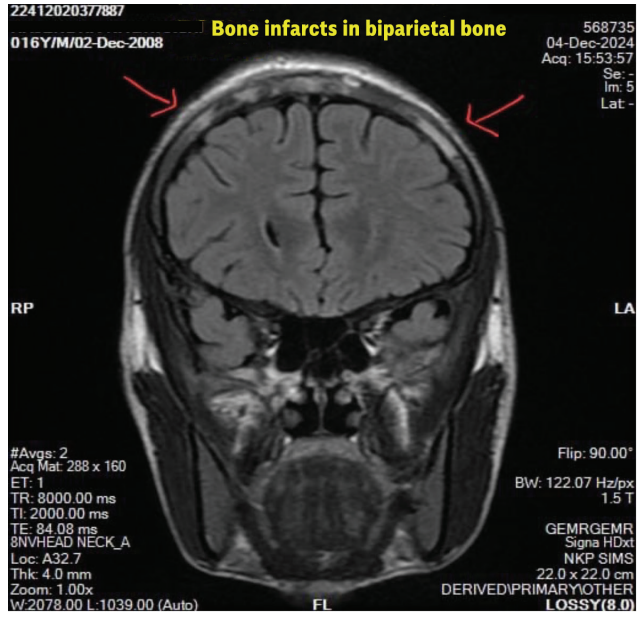

Figure 1. MRI brain with contrast showing bone infarcts in bilateral parietal bone
MRI: Magnetic resonance imaging

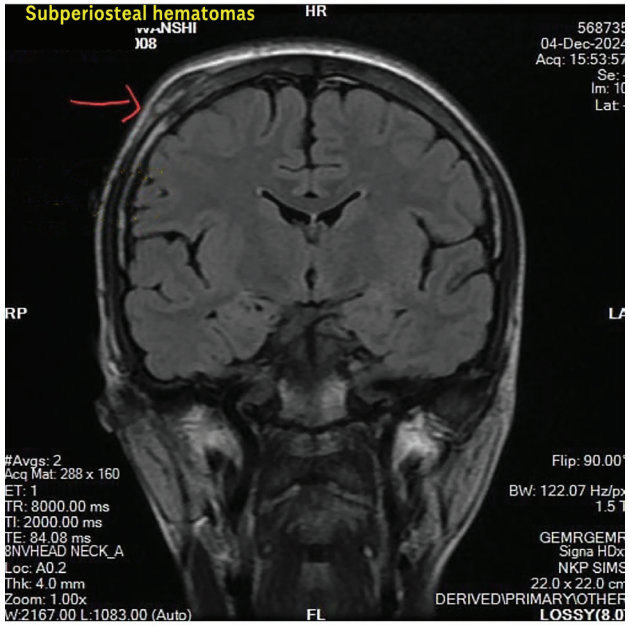

Figure 2. MRI brain with contrast showing right sided subperiosteal haematoma
MRI: Magnetic resonance imaging

after one week, where it was seen that the scalp swelling was significantly reduced.

This case underscores the challenges of managing SCD and the importance of a tailored, holistic approach. With appropriate treatment and regular monitoring, such patients are able to achieve better health outcomes and improved quality of life.

Discussion

SGH is a rare but serious complication in adolescents with SCD. The pathophysiology of SGH in SCD is thought to be related to bone infarctions and vaso-occlusive crises, which can lead to spontaneous bleeding into the subgaleal space.

An uncommon side effect of SCA is a ASHS, which presents as a widespread or localized swelling of the head due to edema and hematoma formation beneath the scalp's galea aponeurotica layer. Epidural hematoma frequently coexists with ASHS in most individuals with a number of hypotheses. The pathophysiology of ASHS is still only partially understood.

Firstly, hypoxia response in SCA is thought to cause prolonged extramedullary hematopoiesis in the skull bones, which weakens and thins the cortical matrices and increases their vulnerability (2). Secondly, angiogenic reactions brought on by hypoxia lead to the development of delicate local vascular beds. These beds work in concert with elevated cardiac output to cause bone fracture and blood leakage into the subgaleal area (3). Thirdly, repeated veno-occlusive crises (VOCs) may cause multiple, mild micro infarctions which, over time, cause bone thinning, local artery wall necrosis, and changed bone and periosteal structures, which can culminate in non-traumatic blood extravasation into subgaleal and epidural areas (4).

The patient's presentation with joint pain, fever spikes, and jaundice is indicative of an acute sickle cell crisis, which can predispose to complications such as SGH. Recent studies have highlighted the importance of recognizing SGH in SCD patients. Over a decade, there have been cases reported from Africa, Arabia, the USA. and recently India (2,5). There was a male preponderance which is similar to our report.

Alqurashi et al. (1) reported a case of a 17-year-old male with SCD who developed a spontaneous SGH, emphasizing the need for prompt medical intervention. In addition to these studies, Foula et al. (2) reported a case of spontaneous SGH in a patient with SCD, highlighting the need for comprehensive care and close follow-up in order to prevent further complications. The presence of jaundice and systemic symptoms suggests the need for evaluation in order to rule out additional complications such as infection or hematologic crisis (2). Recently, a systematic review carried out by Perez et al. (6) stated that headache was the most prevalent complaint at onset (88%). Imaging results frequently showed parietal bone involvement (82%) and bilateral skull infarction (50%) as well. In 65% of instances, an epidural hematoma formed with drainage necessary in 30% of cases and exchange infusion was noted in 18% of cases (6). There were no reported fatalities.

Pathognomonic radiologic characteristics are absent in ASHS. The most sensitive diagnostic method, MRI, usually shows several non-enhancing calvarial lesions which show up as hyperintense on T2-weighted imaging and hypointense on T1-weighted imaging. These lesions may show variable degrees of cerebral expansion without a noticeable mass effect, and they frequently accompany surrounding edema (6). Additionally, MRI is crucial in order to identify related intracranial abnormalities such as extramedullary haematopoiesis and extra-axial collections. Conservative therapy was associated with a prolonged hematoma resolution time of one to two weeks, according to earlier publications (2,4,7)

Drew et al. (8) screened 786 reports of ASHS in paediatric patients, and among these, there were descriptions of

epidural hematomas, subdural hematomas, or SGH. Any of the cranial bones may be affected by these issues (2). A VOC is frequently the setting for reported cases which was similar to our case. Frontal bone infarction in a patient with Hb sickle cell beta thalassemia genotype was reported in just one case (2). Extracranial or intracranial extensions have been reported in the majority of instances in the literature (8). There are many complications of SCD with the following guidelines and strategies for practitioners recommended by Drew et al. (8,9).

1. Cranial MRI is the preferred imaging modality for diagnosing ASHS detecting intracranial involvement and silent cerebral infarct areas.

2. Episodes of ASHS can include intracranial bleeding and thus require urgent neurological assessment in order to detect neurological emergencies.

3. Initial steps in managing ASHS should follow those strategies for VOC including hydration, pain control, and simple transfusion. In addition, exchange transfusion can rapidly and effectively resolve symptoms of ASHS, decreasing the risk of intracranial progression.

4. All patients with episodes of ASHS should have their chronic SCA management escalated (i.e., an increase in hydroxyurea dosing, and a consideration of an exchange transfusion program).

Conclusion

Skull infarction is a potentially serious side effect of SCD which poses particular clinical difficulties. However, although ASHS is usually treated conservatively, a precise diagnosis is essential, as a misdiagnosis could result in unnecessary surgical procedure. Comprehensive care and close follow-up are essential in managing such patients in order to prevent further complications and ensure optimal outcomes. The patient's SGH was managed conservatively with cold compression and anti-inflammatory drugs.

Ethics

Informed Consent: Informed written consent was obtained from one of parent.

Footnotes

Authorship Contributions

Concept: G.C.R., R.A.S., G.N., Design: G.C.R., R.A.S., Data Collection or Processing: G.C.R., R.A.S., D.A., Analysis or Interpretation: G.C.R., R.A.S., D.A., G.N., Literature Search: G.C.R., R.A.S., Writing: G.C.R., R.A.S.

Conflict of Interest: No conflict of interest was declared by the authors.

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References

1. Alqurashi MM, Raslan OM, Gmati GE. Spontaneous subgaleal hematoma in a sickle cell disease patient: a case report. J Med Cases. 2020; 11:46-8.
2. Foula MS, Hassan A, AlQurashi A, Alsaihati A, Sharroufna M. Spontaneous subgaleal hematoma in a patient with sickle cell disease: a case report and literature review. Clin Case Rep. 2019; 7:2220-4.
3. Dahdaleh NS, Lindley TE, Kirby PA, Oya H, Howard MA 3rd. A "neurosurgical crisis" of sickle cell disease: case report. J Neurosurg Pediatr. 2009; 4:532-35.
4. Nelson CE, Scarfone RJ. A teenager with sickle cell disease and scalp swelling. Pediatr Emerg Care. 2018; 34:e168-70.
5. Magitta NF, Komanya FB, Alphonce BO, Bitesigilwe MD, Sindato EM, Meda JR. Acute soft head syndrome in a teenager with sickle cell anemia: a case report. Clin Case Rep. 2023; 11:e8174.
6. Perez AM, Garcia-Guaqueta DP, Setty BN, Neri C, Torres AR. Children and adolescents with sickle cell disease and skull infarction: a systematic review. J Child Neurol. 2024; 39:433-9.
7. Al-Ansari RY, Al Harbi M, Al-Jubair N, Abdalla L. Acute soft head syndrome (subgaleal haematoma) with periorbital oedema as a rare presentation in sickle cell disease. Eur J Case Rep Intern Med. 2020; 7:001766.
8. Drew M, Fladeland D, Sinha R. Acute soft head syndrome in sickle cell anaemia: creating a firm approach. J Hematol Thrombo Dis. 2023; 11:1-4.
9. Brandow AM, Carroll CP, Creary S, et al. American Society of Hematology 2020 guidelines for sickle cell disease: management of acute and chronic pain. Blood Adv. 2020; 4:2656-701.