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Case Report

Acute Soft Head Syndrome in Sickle Cell Anemia: Creating a Firm Approach

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ABSTRACT

Sickle Cell Disease (SCD) is the most common inherited disease worldwide and presents with a myriad of complications. A rare complication of SCD is Acute Soft Head Syndrome (ASHS). We present the case of a 14-year-old male with homozygous sickle cell anemia (HbSS) who presented with Vaso-Occlusive Crisis (VOC) complicated by the peculiar development of a scalp mass. Magnetic Resonance Imaging (MRI) showed calvarium changes consistent with ASHS. Current literature lacks standardized management for such a complication. As such, we outline recommendations for imaging modalities, therapeutic interventions, and ongoing management based on this patient's course over two years.

Keywords: Head/Scalp swelling; Bone infarction; Sickle cell anemia; Acute soft head syndrome; Hematoma; Pediatric/Child/Adolescent

Abbreviations: SCD: Sickle Cell Disease; HbSS: Homozygous Hemoglobin SS Genotype; VOC: Vaso-occlusive crisis; ASHS: Acute Soft Head Syndrome; EDH: Epidural Hematoma; SDH: Subdural Hematoma; SGH: Subgaleal Hematoma; SCI: Silent Cerebral Infarcts; MRI: Magnetic Resonance Imaging; HbSβ+: Sickle Cell Beta Thalassemia

INTRODUCTION

Sickle Cell Disease (SCD) includes a diverse number of hemoglobinopathies, the most severe being the homozygous HbSS form [1,2]. There are many complications of SCD with guidelines and strategies for practitioner's use [3-5]. However, none of these guidelines directly address the complication of ASHS. This uncommon finding is a cranio-vascular phenomenon with limited knowledge regarding pathophysiology and implications in pediatric patients. Our patient case represents a novel effort to accurately diagnose, effectively manage, and longitudinally observe patient outcomes in ASHS. Our aim is to increase awareness and recognition of this clinical presentation, and to provide practitioners with a structure for managing this rare complication. Written consent was obtained from the patient and his parents. A literature review was conducted utilizing database searches of MEDLINE, Embase, and Cochrane Central Register of Controlled Trials. Keywords included sickle cell disease, sickle cell anemia, osteonecrosis/bone infarction, pediatric/child/adolescent, head/scalp swelling, acute soft head syndrome. Studies were individually reviewed, including those reporting acute swelling and/or bony infarcts in patients 0-18 years. A total of 696 papers were found using these parameters. A manual review of abstracts was conducted to select relevant papers and 27 were included for full review. Approximately half of the fully reviewed papers focused on facial and orbital complications.

CASE PRESENTATION

A 14-year-old male of Nigerian descent with known HbSS genotype presented to a Canadian tertiary pediatric hospital with a suspected VOC. He presented to hospital six times with VOC in the preceeding two years despite medical therapy, including hydroxyurea, folate, and prophylactic antibiotics with confirmed compliance. One previous admission was complicated by osteomyelitis and septic arthritis to the left elbow, managed with surgical drainage and antibiotic therapy.

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At the time of this index presentation, he complained of refractory pain to the lower back and was admitted to the pediatric ward at a tertiary care pediatric hospital. He showed initial symptom improvement with intravenous fluids, acetaminophen, ibuprofen, and intravenous morphine. Forty-eight hours after admission, he had an evolution of pain to the shoulders and arms. At this time, a swelling appeared on the right parietal region of the scalp. The mass was soft and fluctuant, not causing significant pain. There were no dermatologic findings at the site of swelling.

An ultrasound of the scalp showed a superficial fluid collection consistent with a Subgaleal Hematoma (SGH). To differentiate infarction versus infection, an MRI brain was performed and showed evidence of three separate calvarium lesions that were non-enhancing, with decreased T1 and increased T2 signal. Each lesion was associated with surrounding edema and fluid. Each had a variable degree of intracranial extension, with no mass effect noted (Figure 1).

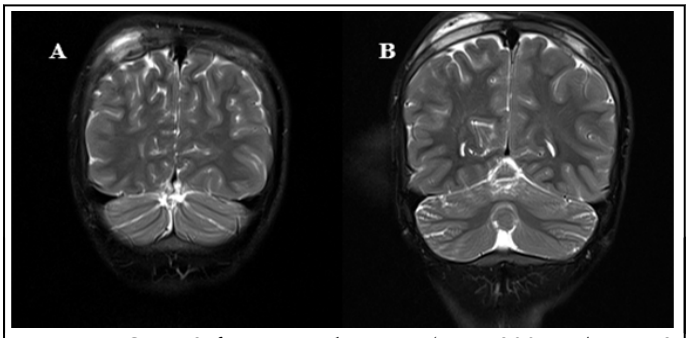

Figure 1: Cor T2 fat-saturated images (TR=7230 ms/TE=112 ms), 4.5 mm thickness. Image A shows a 3 mm depth intracranial fluid collection overlying right parietal lobe. Image B shows a 28 mm focus of increased T2 signal edema throughout the width of the right parietal bone. **Note:** Overlying superficial lenticular extracranial fluid collection measuring 34 mm in diameter and 6 mm in width.

The MRI report suggested that the findings were in keeping with ASHS. Additionally, several punctate areas of increased T2 signal were seen in the subcortical region of the left hemisphere consistent with remote lacunar infarction. Based on the significant intracranial and extracranial findings, the patient was given an exchange transfusion that same day, with rapid and complete resolution of clinical swelling and pain over the next 48 hours. During outpatient review of his clinical course and imaging, his hydroxyurea dosing was increased to the higher end of treatment range, and he began an ongoing exchange transfusion program every 4-6 weeks. The transfusions continued for over two years following the index presentation. The patient has had no further admissions to hospital for VOC and no additional episodes of swelling. Repeat MRI of the brain one year later showed resolution of the calvarium lesions, with overall stability of previously noted lacunar infarcts. Cerebral vasculature was otherwise normal.

DISCUSSION

Patients with SCD can experience complications that are numerous and severe. Particular concern are the effects of neurological complications [4]. This clinical case makes known a rare complication not previously reported in Canada. It confirmed that cerebral ischemic events can predate ASHS and thus need to be considered [6].

Reports of ASHS in pediatric patients in the literature are presented as case reports, and have included Epidural Hematomas (EDH), Subdural Hematomas (SDH) or SGH as associated findings [7-10]. These complications can involve any of the cranial bones [11]. Reported cases often occur in the context of a VOC. Only one case report described frontal bone infarction in a patient with Sickle Cell Beta Thalassemia (HbSβ +) genotype [11,12]. Most cases in the literature report extracranial or intracranial extension, but this case reported both simultaneously (Figure 2).

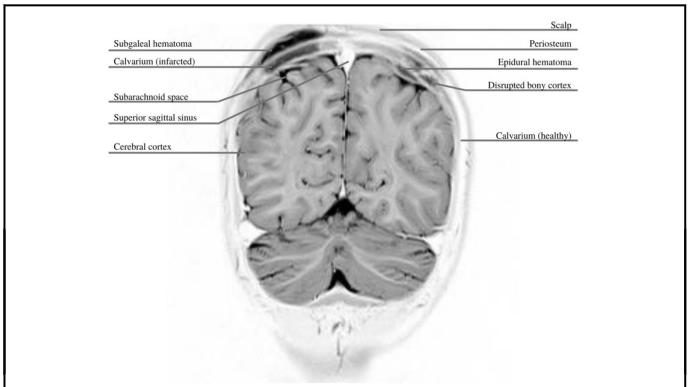

Figure 2: Inverted Cor T2 fat-saturated images (TR=7230 ms/TE=112 ms), 4.5 mm thickness, and is an identical coronal slice to Image B from Figure 1.
Note: Labels outline the anatomic structures of the head, and the multiple anatomic layers involved in fluid collections associated with ASHS.

The pathophysiologic mechanism for ASHS has been postulated in previous reports. Some have suggested cortical disruption and subsequent extravasation of blood and marrow contents into the extracranial and/or intracranial spaces leading to EDH, SDH and SGH formation. Our case shows that intracranial and extracranial complications occurred in the setting of the same infarct episode, which raises concern for inherent bone structure vulnerability in patients with the HbSS genotype. The severity and location of infarction and cortical disruption likely determines the anatomic layer affected by extravasation.

The literature review revealed a notable clustering of ASHS in adolescent males, and an impressive association with the HbSS [7-10,13-17]. Uncertainty exists as to why this occurs, but bone marrow hyperplasia [18], and chronic bony changes secondary to increased hematopoiesis [19] could promote susceptibility to calvarium infarcts with subsequent disruption of a poorly developed cortical bone structure. It is unknown if this phenomenon could be related to physiologic changes in males associated with pubertal and adolescent development.

Current guidelines for SCD outline that bone pain and infarcts can occur in most bones, but common locations are the long bones, ribs, sternum, spine, and pelvis [3]. Calvarium infarcts are

exceedingly rare findings, and the associated ASHS lacks an approach for how best to be investigated and managed.

Based on our experience, ASHS should be viewed as a harbinger for neurological complications. This patient had preceding Silent Cerebral Infarcts (SCI), previously undiagnosed. Evidence of scalp swelling, tenderness, or headache warrants imaging to assess for intracranial extension, as well as SCI.

This patient showed impressive resolution of clinical swelling and pain with the aid of exchange transfusion therapy. Previous reports noted a longer hematoma resolution time of 1-2 weeks with conservative management [7,9,11,13]. During conservative management of ASHS, extracranial to intracranial progression requiring surgical intervention has occurred [7]. This occurrence, contrasted with our case, highlights the potential role for exchange transfusion in mitigating intracranial progression.

Repeat MRI brain imaging one year later demonstrated resolution of calvarium infarcts, with stabilization and non-recurrence of SCI with exchange transfusion therapy. This case provides a unique perspective on the role exchange transfusion can play in the ongoing management of patients who experience ASHS, develop associated EDH, and have underlying SCI.

Recommendations for investigating and managing ASHS

1. Magnetic Resonance Imaging (MRI) brain is the preferred imaging modality for diagnosing ASHS detecting intracranial involvement, and discovering sites of silent cerebral infarction.
2. Episodes of ASHS can include intracranial bleeding and thus require urgent neurological assessment to detect neurologic emergencies.
3. Initial steps in managing Acute Soft Head Syndrome (ASHS) should follow those strategies for VOC including hydration, pain control, and simple transfusion. In addition, in facilities that have the capacity, exchange transfusion can rapidly and effectively resolve symptoms of ASHS, decreasing risk of intracranial progression.
4. All patients with episodes of ASHS should have their chronic sickle cell anemia management escalated (i.e. increasing hydroxyurea dosing, considering exchange transfusion program).

CONCLUSION

This case effectively illustrates the clinical presentation of Acute Soft Head Syndrome (ASHS), and sheds light on management strategies that can optimize outcomes and potentially reduce the likelihood of subsequent intracranial sequelae. Despite variation in regional health care resources, there is utility in establishing an approach to investigating and managing Acute Soft Head Syndrome and its associated complications. These recommendations can offer health care providers an outline for application to patients with sickle cell anemia who may present with this rare syndrome.

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