Case Report

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Skull Bone Osteomyelitis in A Sickler With Acute Soft Head Syndrome Managed Surgically; A Rare Case Report

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Abstract

Introduction: *Acute soft head syndrome is an extremely rare complication of sickle cell disease that mostly affects children and young persons. It occurs due to focal necrosis of skull bone secondary to occlusion of blood supply to that region by micro-thrombi. Upon reviewing the literature, only 3 cases were reported and the three of them were managed conservatively. In this paper we would like to present a rare case with acute soft head syndrome as the 1st case with acute soft head syndrome that is been managed surgically.*

Case Description: *A 17 years old young male, known case of sickle anemia presented with spontaneous forehead swelling for about 10 days duration and pus discharge from the forehead swelling. CT-brain reported Left frontal scalp soft tissue mass with infiltration and destructive changes to the underlying frontal bone. There was no intracranial extension. CT with contrast showed septations with enhancement of the walls of the subgaleal collection and enhancement of the underlying dura. The abscess was evacuated, the bone was looking infected and the periosteum also was unhealthy looking, therefore both were excised. The inner table and outer table were involved at different levels, the abnormal bone was nibbled away until normal edges were encountered. Histopathology revealed XANTHOGRANULOMATOUS INFLAMMATION WITH FIBROPLASTIC REACTION AND CONGESTION COMPATIBLE WITH ORGANIZING ABSCESS. The patient was discharged in a good shape, with intact neurological function, GCS 15/15 afebrile and healthy-looking surgical wound. He was scheduled for elective cranioplasty with custom-made bone flap later on on elective basis.*

Conclusion: *This case represents a unique presentation of these conditions with osteomyelitis other than the usual presentation with necrosis. This draws our attention to the fact that not all of these cases can be managed conservatively. Brain imaging with contrast is an important diagnostic tool that can pick up the cases that might need surgical intervention. The early surgical intervention fasten patient's recovery and prevents the spread of infection if any. More studies need to be carried out to establish international guidelines for managing this rare condition.*

Keywords: Acute soft head syndrome, sickle cell anemia, osteomyelitis, bone necrosis, early surgical intervention.

1. Introduction

Osteomyelitis of skull bone can develop either in acute or chronic forms. Acute osteomyelitis occurs secondary to scalp infection when the abscess is in close contact with skull bone [1]. If the patient is immunocompromised or was not managed properly, it can spread deeper to affect the underlying brain tissue[1] and if it is close to the dural venous sinuses it can be complicated with sinus thrombosis [2]. Also it can occur posttraumatic even if it was an old trauma specially when the patient has subgaleal hematoma and was not covered adequately with antibiotics [3-6]. Another source for skull osteomyelitis can be from the nearby sinusitis, mastoiditis as well as otitis. Organisms isolated can be Staphylococcus aureus, Streptococcus Pneumoniae, Corynebacterium, Candida and Escherichia coli [7-9]. During COVID-19 pandemic, some cases of skull osteomyelitis were discovered secondary to Post-COVID mucormycosis and they required aggressive combined

medical and surgical management [10]. On the other hand, chronic osteomyelitis usually affects immunocompromised patients and is usually caused by Cryptococcus neoformans which was reported in about 10% of the cases with disseminated cryptococcosis [11].

When this affects the frontal bone it is called Pott's Puffy Tumor; a rare entity characterized by frontal skull osteomyelitis with underlying subperiosteal collection. It is usually caused by untreated sinusitis, trauma or odotogenic disease. The disease is usually polymicrobial and requires multidisciplinary approach to treat the patient [12].

Acute soft head syndrome is an extremely rare complication of sickle cell disease that mostly affects children and young persons [13]. It occurs due to focal necrosis of skull bone secondary to occlusion of blood supply to that region by micro-thrombi. This is usually followed by a chemical reaction mediated by the inflammatory cells secondary to bone necrosis. Patients with this condition usually present with headache, low grade fever, skull bone-ache and acute progressive subgaleal scalp swelling [13-15]. This condition can be investigated with skull x-ray CT Brain including 3D reconstruction and MRI [16, 17]. Due to the scarcity of cases reported with this condition, up to the moment still there are no existing guidelines to manage this condition [14].

Upon reviewing the literature, only 3 cases were reported [13-15] and the three of them were managed conservatively. In this paper we would like to present a rare case with acute soft head syndrome as the 1st case with acute soft head syndrome that is been managed surgically.

2. Case Description:

A 17 years old young male, known case of sickle cell anemia was following in hematology clinic. He presented to the clinic with spontaneous forehead swelling for about 10 days duration. Empirical antibiotic was commenced, 10 days later he developed pus discharge from the forehead swelling. So, he was referred from the clinic to the ER. Neurosurgery team was contacted as his CT-brain reported Left frontal scalp soft tissue mass with infiltration and destructive changes to the underlying frontal bone. There was no intracranial extension.

On examination the patient was conscious, oriented, GCS was 15/15, no motor or sensory deficit and no cranial nerves abnormalities were detected.

Temperature was 37C, pulse rate was 69 bpm, BP was 110/70 and O2 saturation was 98% in room air.
Local examination revealed about 6-7cm x 3 cm forehead swelling not tender, small area of hear loss seen around the lesion (alopecia) although no reported history of trauma to this area. (Fig.2C)
Labs showed: WBCs 8430 – HG 10.1 – plats 601.000 - INR 1.14 - PT 15.4 - PTT 30.5
Echocardiogram was performed as part of investigation and was reported as normal
Hemoglobin electrophoresis: Hb A 1.6% \ Hb A2 1.9% \ Hb F 9.4% \ Hb S 41.1% \ Hb C 46%
CT-brain with IV contrast was done on the next day (OCT 13, 2022) and showed a multiloculated frontal subgaleal collection with surrounding enhancing walls and internal septations associated with erosion to the underlying bone and extension to the superior sagittal sinus with a filling defect worrisome for sinus thrombosis. Mild meningeal thickening was noticed at that area that could represent meningitis.
The subgaleal collection was measuring approximately 7 x 2 x 1.5 cm (AP X TR X CC) associated with surrounding edematous changes and soft tissue fat stranding.

The reminder of the major intracranial sinuses were patent including straight, bilateral transverse and sigmoid sinuses as well as vein of Galen, and visualized parts of the internal jugular were also patent. (Fig.1, 2A&B).

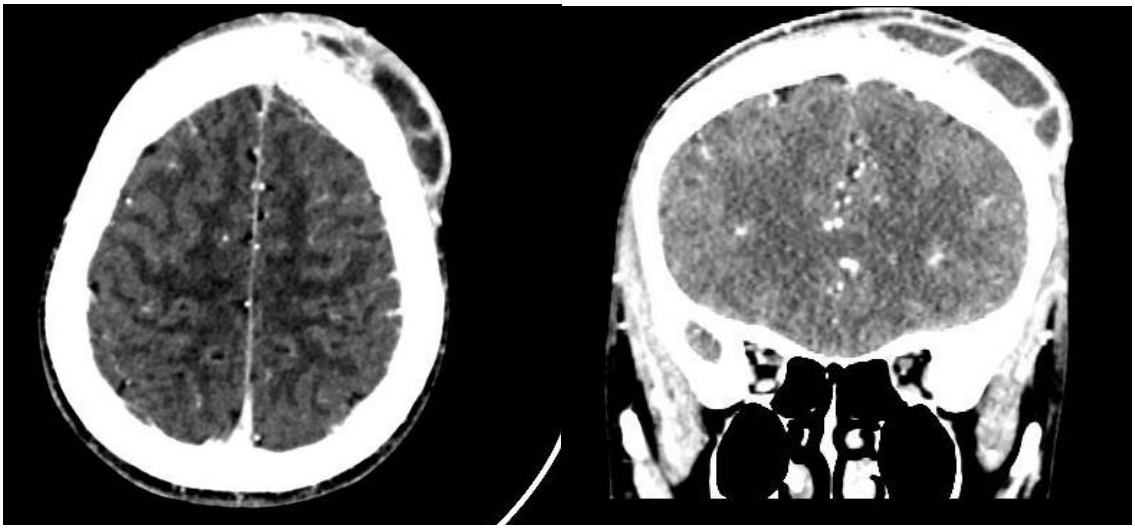
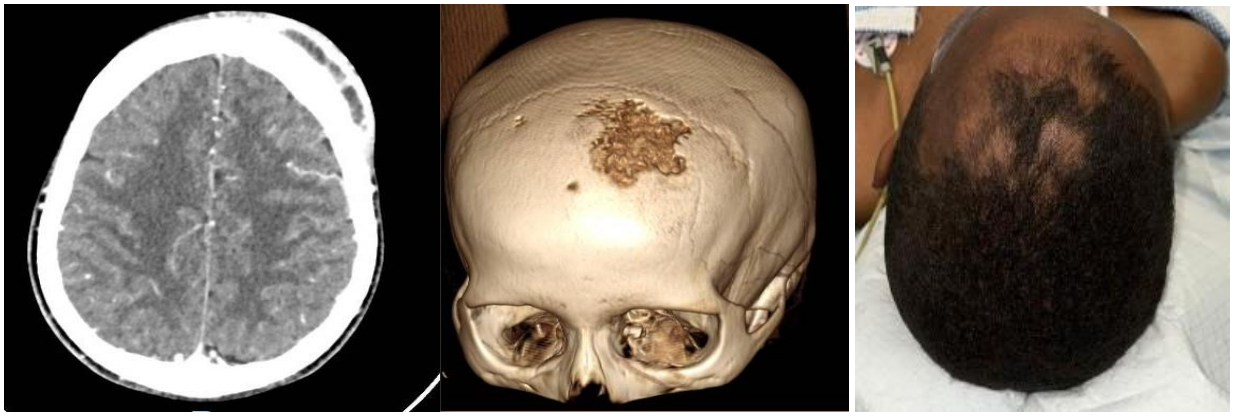


Figure 1: *preoperative CT-brain with intra-veinous contrast showing multiloculated collection with septations at left frontal area (entirely extra-cranial with no intra-cranial extension)*



(A) (B) (C)

Figure 2: *preoperative CT-brain W/contrast showing multiloculated collection with septations at left frontal area (A) CT-brain 3D image showing area of osteomyelitis in frontal bone to ward left side (B), area of alopecia around the collection site, left frontal region (C).*

On Oct 23, 2022 the patient was operated for subgaleal abscess evacuation and excision of the underlying possibly infected frontal skull bone.

Intra-operatively, the abscess was evacuated, the bone was looking infected and the periosteum was also unhealthy looking, therefore both were excised. The inner table and outer table were involved at different levels, the abnormal bone was nibbled away until normal edges were encountered.

Post-operatively, the patient was kept in ICU for close observation, next day he was discharged from the ICU to general ward.
Post-operative MRI brain with gadolinium showed no evidence of osteomyelitis. (Fig.3).

Tissue cultures (abscess fluid and excised bone) were negative
Histopathology report revealed:
- XANTHOGRANULOMATOUS INFLAMMATION WITH FIBROPLASTIC REACTION AND CONGESTION COMPATIBLE WITH ORGANIZING ABSCESS.
-GMS STAIN NEGATIVE FOR FUNGI
It worth mentioning that the patient was already on antibiotics (vancomycin and ceftriaxone) before surgery.

He was been followed by infectious disease (I.D team) and later on, they advise to discontinue antibiotics as the patient is afebrile, clinically stable and tissue cultures were negative.

The patient was discharged in a good shape, with intact neurological function, GCS 15/15 afebrile and healthy-looking surgical wound. He was scheduled for elective cranioplasty with custom-made bone flap later on on elective basis.

3. Conclusion

This case represents a unique presentation of these conditions with osteomyelitis other than the usual presentation with necrosis. This draws our attention to the fact that not all of these cases can be managed conservatively. Brain imaging with contrast is an important diagnostic tool that can pick up the cases that might need surgical intervention. The extent of craniectomy should be based on carefully studying pre-operative images and how does the bone looks like intraoperatively. It is important to trim out all unhealthy bone until the healthy edges are encountered. The early surgical intervention fasten patient's recovery and prevents the spread of infection if any. More studies need to be carried out to establish international guidelines for managing this rare condition.

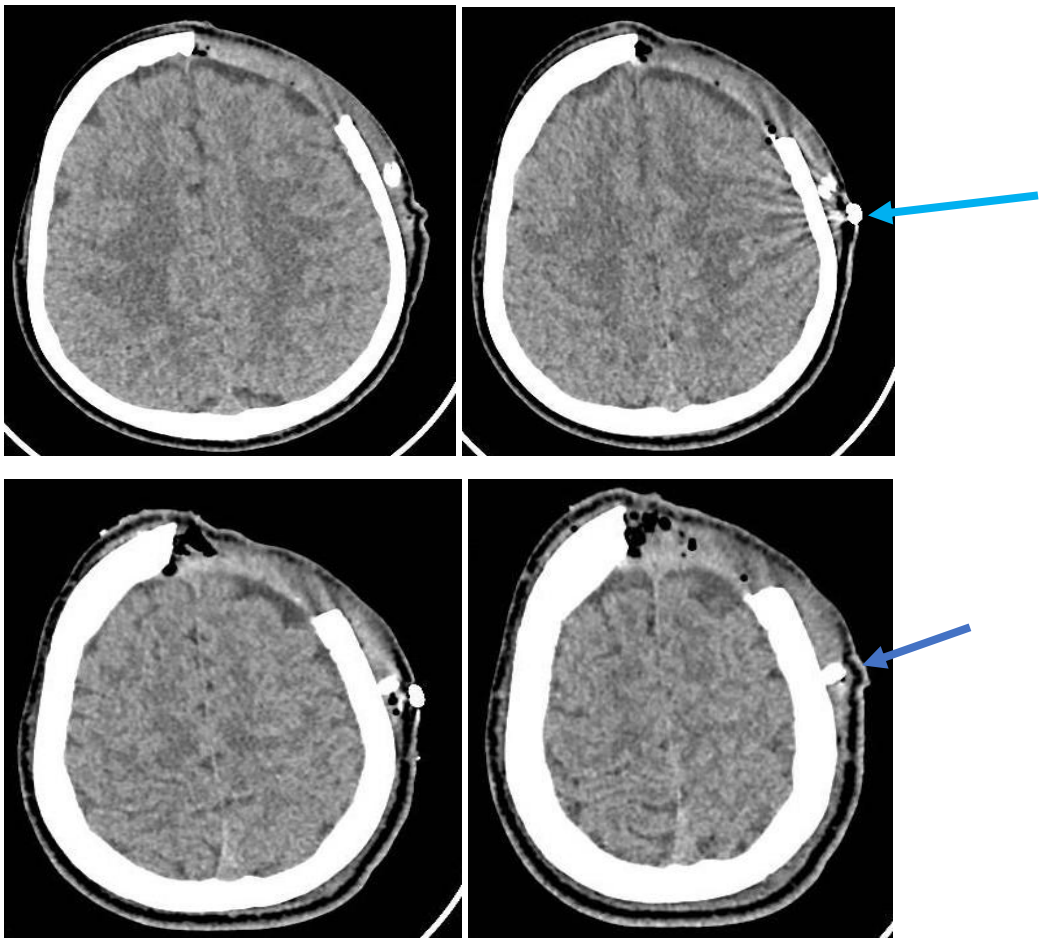


Figure 3: *showing post operative CT-brain with drain inserted in subcutaneous space (blue arrows)*

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