Case 16346

Eurorad ••

Boy with sickle cell disease presenting with swelling bilateral fronto-parietal region

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DOI: 10.1594/EURORAD/CASE.16346 ISSN: 1563-4086 Section: Neuroradiology Area of Interest: Head and neck Haematologic Neuroradiology brain Procedure: Screening Procedure: Diagnostic procedure Procedure: Drainage Imaging Technique: Ultrasound Imaging Technique: Experimental Imaging Technique: CT Special Focus: Haemorrhage Blood Case Type: Clinical Cases Authors: Dr.Aman Gupta.Dr.HeeraRam.Dr.Chandrajeet Yadav. Patient: 16 years, male

Clinical History:

An 16 year old male child, a known case of sickle cell disease with swelling bilateral fronto-parietal region was referred for sonography local region with clinical suspicion of scalp abscess. On examination the swelling was tender, boggy, mobile and fluctuant. There was no history of head trauma or previous such episodes of scalp swellings.

Imaging Findings:

High resolution ultrasonography (HRUS) revealed anechoic collection 12 mm thick in subgaleal space in bilateral fronto-parietal region (Fig-1) .An USG guided aspirate from the swelling was performed which yielded hemorrhagic fluid (Fig-2).CT head revealed large extracalvareal subgaleal collection/hematoma along bilateral fronto-parietal convexity. The maximum thickness of collection was 28 mm.There was no epidural haematoma (Fig-3). His hematological profile was as follows: Hemoglobin 5.6 g/dL, hematocrit 26%, and peripheral blood smear shows microcytic hypochromic anaemia.His platelet count, PT and INR was within normal limit. **Discussion:**

Acute subgaleal hematoma/ Soft head syndrome is rare complications seen in children with sickle cell disease.Literature search has revealed only few cases describing the development of painful acute head swelling in patients with sickle cell disease [1, 2]. Pathophysiology of non-traumatic subgaleal hematomas in sickle cell disease is not yet fully understood, following hypothesis have been proposed:

1.An underlying bone infarction caused secondary to the veno-occlusive crisis disrupts the cortical bone, causing periosteal elevation resulting in bleeding in the subgaleal and epidural spaces [3, 4].

2. Another theory proposes that insufficient venous drainage is the inciting event that leads to oedema and hemorrhage [5].

3. Finally Dahdaleh [6] in 2009 proposed hyper-proliferative bone marrow disrupts the skull cortex and precipitates

extravasations of blood and marrow into the subgaleal and epidural spaces

Imaging plays important role not only in diagnosing subgaleal and epidural collection also in detecting mass effect and signs of bone infarction.High resolution sonography can detect sub galeal collection as in our case. MRI is much more sensitive than CT for detection of bone infarction [7].

Most cases of acute soft head syndrome in children with sickle cell disease resolve with conservative management. However epidural haematoma causing mass effect will require craniotomy and evacuation of EDH. The best treatment of such a rare neurosurgical crisis in sickle cell disease is prevention by preventing sickle cell crisis.

The differential diagnosis in a child with spontaneous sub galeal and epidural haematoma must include bleeding diathesis, vascular malformations of the

dura mater and metastasis to the dura or skull [8, 9].

'Written informed patient consent for publication has been obtained.'

Differential Diagnosis List: Soft Head Syndrome, Bleeding diathesis, Vascular malformations of the dura mater, Metastasis to the dura or skull

Final Diagnosis: Soft Head Syndrome

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Figure 1



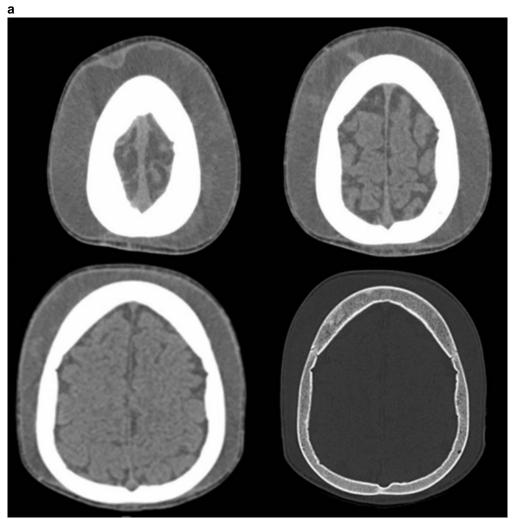
Description: High-resolution ultrasound image panoramic view revealing anechoic collection in subgaleal space in bilateral fronto-parietal region . **Origin:** Dept.Of Radiodiagnosis Sri.Aurobindo Medical College,Indore.

Figure 2



Description: An USG guided aspirate from the swelling yielded hemorrhagic fluid. **Origin:** Dept.Of Radiodiagnosis Sri Aurobindo Medical College.Indore

Figure 3



Description: Plain CT Head revealed large extracalvareal subgaleal collection/hematoma along bilateral fronto-parietal convexity with no epidural haematoma. **Origin:** Dept.Of Radiodiagnosis Sri Aurobindo Medical College,Indore