Skull infarction in patients with Sickle Cell Disease.

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Introduction

Osteonecrosis is a well-described complication of sickle cell disease (SCD) that occurs secondary to vaso-occlusion[1]. Skull infarction is rare and presents with unique clinical challenges and complex anatomy involving vital structures that can be life-threatening[2].

Objective

This study aims to explore the main clinical and imaging findings in patients with SCD who developed skull infarction.

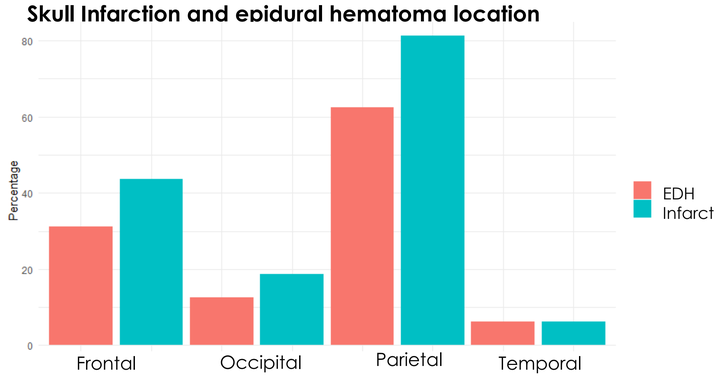
Materials and methods

We searched the Pubmed database for case reports on skull bone infarction in pediatric patients with SCD. We identified 67 articles and included 15 for analysis. A 16th case reported by senior author was included. We extracted the most relevant clinical data for each case and analyzed it.

Results

Seventy-five percent of the cases were males aged 7 to 21 years. The most common symptom at onset was headache (88%); in the remaining 12%, headache appeared as a secondary symptom. Epidural hematoma developed in 65% of the cases, and subgaleal hematoma appeared in 29%. The location of the skull infarction corresponded with the location of the hematoma in 70% of the cases. Bilateral skull infarction (50%) and parietal bone involvement (82%) were the most common imaging findings. No fatal outcomes were reported. 30% required drainage, and conservative measures were the only treatment in 24% of the cases, and exchange infusion was reported in 18%.

Demographic and clinical characteristics of cases	
Age (Median, IQR)	17, 7-21
Sex (Male)	12 (75%)
Haplotype SS SC	 9 (81.8%) 2 (18.2%)
Ongoing treatment Hydroxyurea Penicillin Folic acid	 6 (75%) 2 (25%) 4 (50%)
Onset duration- days (Median, IQR)	3.88, 1-9
Headache at presentation	14 (88%)
Scalp swelling	7 (43.8%)
Treatment: Drainage Exchange transfusion Conservative	 5 (45.5%) 3 (27.3%) 4 (36.4%)



Conclusions

We identified headaches as the most common presenting symptom in skull bone infarction.

Persistent headaches in patients with SCD, should raise suspicion of for skull bone infarction and warrants additional investigation and possibly follow up imaging. The association between skull infarction and epidural hemorrhage was considerable, even though none of the cases reported fatal outcomes.

In the emergency department, clinicians should consider skull infarction in pediatric patients with SCD presenting with acute headache. We attempt to increase awareness of this entity, highlight the significance of early detection and management of skull infarction in SCD in a collaborative team-based approach.

References

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