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Acute soft head syndrome –A rare complication of sickle cell disease

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Abstract

Background

Acute soft head syndrome is a rare complication of sickle cell disease. This case report describes an 18-year-old male with sickle cell disease who presented with right-sided periorbital and scalp swelling, indicative of acute soft head syndrome, one of the rarest complications of this condition. The patient was treated without aspiration of the scalp swelling, highlighting that acute soft head syndrome should be considered a differential diagnosis for scalp pain and swelling in sickle cell disease patients.

Key Words : Osteomyelitis, Vaso-occlusive, Subgaleal haematoma

Introduction

Sickle cell disease is a prevalent inherited blood disorder in India, with varying prevalence across different regions. Among tribal groups, the carrier rate ranges from 1 to 40 percent, with Madhya Pradesh having the highest prevalence. In Maharashtra, particularly in the Vidarbha region, the sickle gene is widespread. In Gujarat, tribes such as the Dhodia, Dubla, Gamit, and Naika exhibit a high prevalence of HbS

Sickle cell disease is an autosomal recessive disorder caused by the production of abnormal HbS due to the substitution of valine for glutamic acid. This results in unstable red blood cells with shortened survival, assuming a sickle shape. Sickle cell disease can present acutely as vaso-occlusive phenomena, infections, or severe anemia, and may lead to chronic complications

involving multiple organ systems. In our region, sickle cell disease typically manifests in a milder form.

Bone and joint complications are the most common manifestations of sickle cell disease, affecting bones through infarction, bone marrow hyperplasia, or osteomyelitis, which can involve long bones and flat bones, including the skull. Acute soft head syndrome is an uncommon complication of sickle cell disease.

Case Report

We present the case of an 18-year-old male with known sickle cell disease admitted to New Civil Hospital, Surat. He presented with a severe right-sided headache and swelling over the right scalp and periorbital region for three days. There was no history of joint swelling, fever, dizziness, or prior head trauma. He had a history of recurrent vaso-occlusive crises. Physical examination revealed multiple tender swellings on the right parietal and temporal regions extending to the cheek, with no signs of inflammation. Other systemic examinations were unremarkable. An initial diagnosis of multiple scalp abscesses versus cerebral venous sinus involvement was made, and the patient was treated with broad-spectrum antibiotics (third-generation cephalosporins), adequate analgesia, and hydration.

An MRI on the third day of admission showed subgaleal hematomas with T2 predominant signals in the pre-septal, periorbital, and premaxillary regions, with edema over the right fronto-temporo-parieto-occipital region. Altered signals were seen in the right temporo-parietal bones on STIR images, suggestive of bone infarcts.

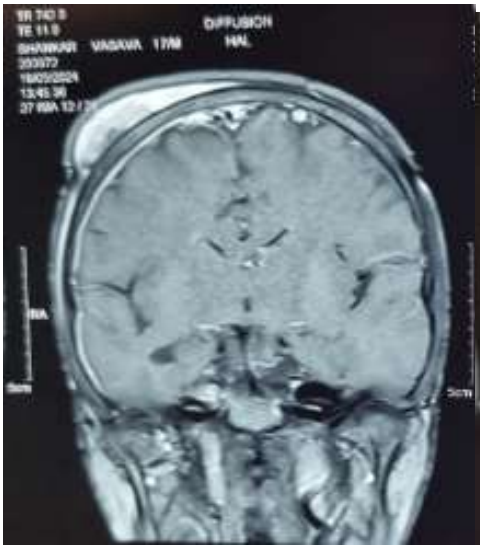

Fig 1 shows MRI BRAIN with subgaleal haematoma

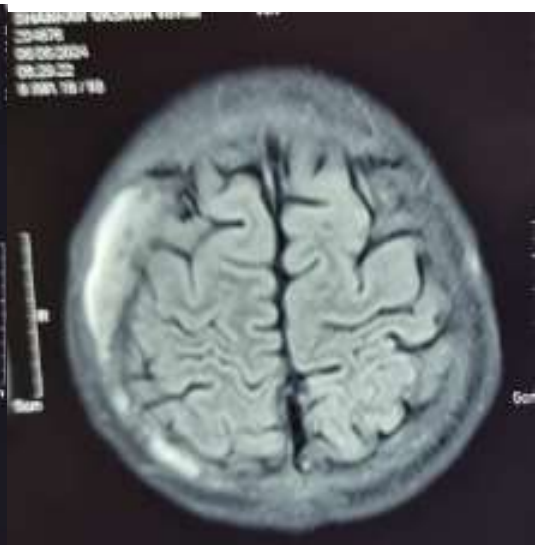

Fig 2 showsaltered signals were seen in the right temporo-parietal bones on STIR images

Variable	Reference Range	On Admission Day 1	On Admission Day 3	On Discharge
HbG (gm/dl)	13-17	9.1	7.8	8.7
WBC (cmm)	4000-10000	10200	8400	8800
Hct (%)	40-50	30.8	23.6	27.1
MCV (fl)	77-93	69	73	75
MCH (pg)	27-32	21	22.8	23.9
MCHC (gm/dl)	31.5-34.5	30.5	31.1	31.9
Plt (cmm)	1.5-4 lakh	401000	316000	247000
Total Bilirubin	0-1.2 mg/dl	0.9	0.6	0.6
Direct Bilirubin	0-0.3 mg/dl	0.3	0.2	0.2
Indirect Bilirubin	0-0.9 mg/dl	0.6	0.4	0.4
ALT (U/L)	30-45	27	12	15
LDH (U/L)	81-360	841	550	314

Based on clinical presentation and imaging studies, the diagnosis was acute soft head syndrome. The patient was treated conservatively with cefoperazone-sulbactam combination, intravenous fluids, and analgesics. A senior neurologist confirmed the management plan. The headache gradually resolved, and the swelling subsided over two weeks. The patient was discharged in satisfactory condition after 14 days.

Discussion

Acute soft head syndrome is a rare manifestation of sickle cell disease, typically presenting as diffuse or localized swelling. Our patient exhibited diffuse swelling of the entire scalp and forehead without periorbital involvement. The pathogenesis involves vaso-occlusive crises leading to skull bone marrow infarction and small vessel tearing. Thinning of cortical bones due to intramedullary hematopoietic tissue expansion disrupts skull margins. Vascular damage results from vasoconstriction, leading to obstruction by sickled red blood cells and platelet fibrin thrombi formation due to decreased fibrinolysis and increased factor VIII activity. Severe vascular damage during crises can cause vessel rupture.

Differential diagnosis of sickle cell-related skull hematoma includes bleeding diathesis and skull bone osteomyelitis, necessitating thorough history and examination to exclude osteomyelitis. Our patient's coagulation studies were normal. Most cases of acute soft head syndrome in sickle cell disease are managed conservatively with hydration and analgesia. Our patient responded well to conservative management and broad-spectrum antibiotics to differentiate between infarction and infection. Aspiration is generally not indicated unless infection is suspected, as it may lead to secondary infection.

Conclusion

Acute soft head syndrome is a rare complication of sickle cell disease associated with repeated vaso-occlusive crises. Recognizing this syndrome is crucial for proper management, which can often be achieved conservatively without the need for aspiration.

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