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Original Article

RADIOLOGICAL FINDINGS OF ACUTE SOFT HEAD SYNDROME IN AN ADOLESCENT: A RARE COMPLICATION OF SICKLE CELL ANAEMIA.

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ABSTRACT

Sickle cell anaemia predominantly affects individuals of African, Mediterranean, Middle Eastern, and Indian descent. Acute soft head syndrome is an uncommon complication linked to this condition. Herein, we report the case of a 17-year-old male patient with sickle cell disease who presented to our emergency department with generalized body pain, primarily localized to the abdomen, lower back, and hips, bilaterally. During his hospital stay, he developed a headache and progressive swelling in the scalp that subsequently extended to the forehead, finally involving the eyes. Radiological features and computed tomography (CT) of the brain demonstrated diffuse subgaleal soft tissue swelling of the scalp with subgaleal fluid collections. Magnetic resonance imaging (MRI) revealed findings consistent with acute osteonecrosis which were indicative of acute soft head syndrome. In light of these radiological findings, conservative management, including intravenous fluids and analgesics, was initiated. This case underscores the significance of considering acute soft head syndrome in the differential diagnoses for headache and scalp swelling in adolescents with a history of sickle cell anaemia. Furthermore, the purpose of this report is to highlight the significance of radiological evaluation in the proper management of acute soft head syndrome.

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يُعد فقر الدم المنجلي مرضًا وراثيًا ينتشر بشكل رئيسي بين الأفراد من أصول إفريقية، متوسطية، شرق .أوسطية، وهندية .تُعتبر متلازمة الرأس الطري الحادة إحدى المضاعفات النادرة المرتبطة بهذا المرض في هذا التقرير، نستعرض حالة مريض يبلغ من العمر 17 عامًا، يعاني من فقر الدم المنجلي، حيث حضر إلى قسم الطوارئ يشكو من آلام عامة في الجسم، تركزت بشكل رئيسي في البطن وأسفل الظهر والوركين .أثناء فترة إقامته بالمستشفى، ظهرت أعراض أخرى تمثلت في صداع وتورم تدريجي بفروة الرأس امتد لاحقًا إلى الجبهة، ثم العينين .وقد أظهرت نتائج الأشعة - بما في ذلك التصوير المقطعي المحوسب (CT) والتصوير بالرنين المغناطيسي (MRI) - تورمًا منتشرًا في الأنسجة الرخوة تحت فروة الرأس، بالإضافة إلى وجود نخر عظمي حاد، مما يدعم تشخيص الإصابة بمتلازمة الرأس الطري الحادة .اعتمدت خطة العلاج على التدابير التحفظية، مثل السوائل الوريدية ومسكنات الألم، دون الحاجة إلى أي تدخلات جراحية .تحسنت حالة المريض تدريجيًا، وتماثل للشفاء قبل مغادرته المستشفى دون أي مضاعفات. تؤكد هذه الحالة الدور الحاسم للتشخيص الشعاعي في تحديد متلازمة الرأس الطري الحادة وعلاجها بالشكل الصحيح، كما تسلط الضوء على أهمية الوعي بهذا النوع من المضاعفات النادرة لتجنب التشخيص الخاطئ أو اللجوء إلى إجراءات طبية غير ضرورية.

Keywords: Acute soft head syndrome; Sickle cell disease; Osteomyelitis; Case report.

1. INTRODUCTION

Sickle cell anaemia is a hereditary condition resulting from a mutation in the haemoglobin gene, leading to sickle-shaped cells that obstruct blood circulation, leading to pain, anaemia, and organ damage [1]. A rare consequence of sickle cell anaemia, referred to as acute soft head syndrome, presents with unexpected scalp swelling, pain, and tenderness; it is caused by sickle cell crises restricting blood flow, thereby causing localised infarctions. Appropriate evaluation is required to rule out haemorrhagic, infectious, or neoplastic conditions [2]. Magnetic resonance imaging (MRI) and computed tomography (CT) are crucial diagnostic tools for verifying the diagnosis of acute soft head syndrome [3]. Herein, we report the case of a 17-year-old male patient with a known history of sickle cell disease, admitted to our hospital's emergency department due to generalised body pain and shortness of breath.

2. CASE PRESENTATION

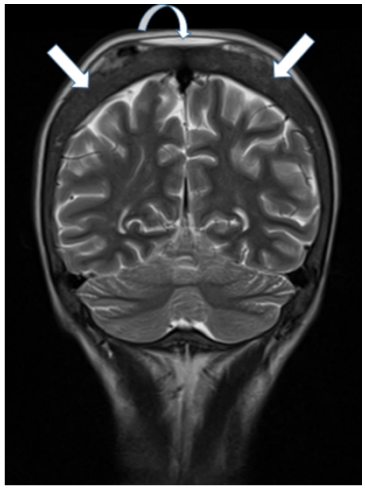
A 17-year-old boy with sickle cell disease was admitted to the emergency department

due to generalised body pain and pain-induced shortness of breath for a day. The patient had no recent history of head trauma; however, he had a history of vaso-occlusive episodes and had undergone a laparoscopic cholecystectomy at the age of 12.

On the first day after admission, the patient developed fever, prompting a septic workup followed by initial treatment with azithromycin and ceftriaxone. Three days after admission, he complained of a headache and swelling of the right forehead. The swelling was localised, soft, non-tender, non-fluctuant, and measured 3 × 3 cm, with no ecchymosis or discoloration, and no visual disturbances or swelling in other body parts. Five days after admission, the swelling had extended to the right eyelid and part of the nasal bridge.

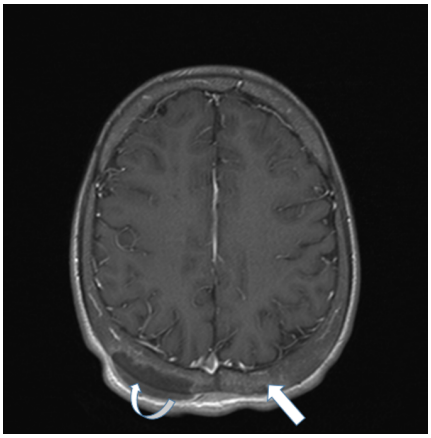
Initial examination revealed that the patient was conscious, alert, and oriented. He lay in bed and complained of diffuse abdominal pain. The vital signs were as follows: temperature: 36.8◦C; O2 saturation: 100%; blood pressure: 126/72 mmHg; chest examination: normal; abdominal examination: soft and lax, with widespread tenderness. Initial investigations showed leucocytosis, neutrophilic predominance, and normocytic normochromic anaemia, with a baseline of 9–10 Hb. The patient was administered intravenous fluid and narcotic analgesics.

The radiological findings were consistent with sickle cell disease, demonstrating multiple regions of acute osteonecrosis in the calvarium and skull base accompanied by adjacent reactive soft tissue collections indicative of acute soft head syndrome. Evidence of old lacunar infarctions was also present in the deep white matter of both the cerebral hemispheres.



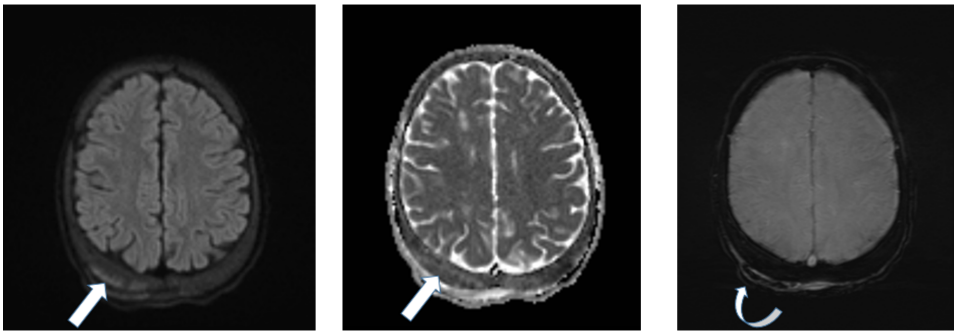
On coronal T2-weighted imaging, diffuse calvarial thickening is visible with altered signal intensity of the bone marrow; high signal intensity is observed for preserved subcutaneous fat (arrow).

Figure 1



Axial T1-weighted post-contrast image showing faint calvarial enhancement with no dural enhancement (arrow). Lack of enhancement of the subgaleal collection (curved arrow).

Figure 2



Axial diffusion-weighted image. ADC and SWI show dependent diffusion restriction (arrow) with internal susceptibility effects related to blood products within the subgaleal collection (curved arrow).

Figure 3

Following a thorough assessment of the patient's clinical symptoms and imaging findings, the diagnosis was confirmed as acute soft head syndrome. The selected course of treatment was conservative in nature, which included intravenous fluids and analgesics; the antibiotics were discontinued. The purpose of this treatment was to stabilise the condition of the patient and provide symptomatic relief. The swelling of the scalp subsided significantly. The patient was discharged without any complications and is currently being followed-up with haematology for sickle cell disease.

3. DISCUSSION

Sickle cell disease, a genetic disorder that impacts haemoglobin production, leads to the formation of abnormally shaped and inflexible red blood cells. This alteration in red blood cell morphology can impede vascular circulation, elevating the risk of infections, causing discomfort, and inflicting damage to various organs. This condition predominantly affects people of African, Mediterranean, Middle Eastern, and Indian ancestries [4]. Among the rare complications associated with sickle cell disease is acute soft head syndrome, defined by sudden, quickly advancing, localized scalp swelling. Failure to recognize this condition can result in misdiagnosis and unwarranted surgical procedures [5]. Although the precise pathophysiological mechanisms underlying acute soft head syndrome are not fully understood, proposed explanations include bone infarction, osteopenia leading to cortical bone thinning, periosteal disruption with subsequent blood extravasation into the subgaleal space, and extramedullary hematopoietic involvement of the cranial flat bones [6].

Radiological imaging is essential for identifying acute soft head syndrome, particularly within individuals affected by sickle cell disorder. For such cases, CT of the brain typically shows diffuse swelling of the subgaleal soft tissues and subgaleal fluid collections suggestive of haematomas. These findings, especially in the absence of trauma, indicate acute soft head syndrome, an established consequence of sickle cell condition [3]. Ultrasound (US) can help rule out fluid collections or abnormalities, as demonstrated by the absence of issues above the right eyebrow in our patient. MRI is the most sensitive diagnostic protocol, revealing diffuse calvarial thickening and altered bone marrow signal intensity characterised by the loss of normal T1-weighted hyperintensity. High signal intensity on T2-weighted/FLAIR sequences and heterogeneous signals on T1-weighted images indicate bone marrow oedema and infarction [3]. MRI also detects associated diffuse subgaleal fluid collections with heterogeneous signals and internal susceptibility effects from blood products. Additional findings may include smaller areas of altered signal intensity, subgaleal oedema, and focally enhancing bony lesions, such as haemangiomas or venous lakes. Importantly, MRI helps rule out acute brain infarctions, intracranial haemorrhages, and abnormal brain parenchyma, ensuring the comprehensive evaluation and accurate diagnosis of acute soft head syndrome [3].

A literature review of two similar cases revealed similar case presentations with specific MRI findings [5, 7]. MRI in these cases revealed abnormal signal intensities in the calvarium and associated fluid collection. The first case showed bilateral high signal intensities in the calvarium, accompanied by subgaleal collections and oedema. Similarly, our case demonstrated diffuse calvarial thickening with an altered bone marrow signal, along with subgaleal collections and blood products. The second case also involved fluid

collections, but these were subperiosteal with associated haemorrhagic content, comparable to the blood products observed in the first reported case, and in ours [5, 7].

Despite differences in the locations of the fluid collections, calvarial pathology, fluid collections, and haemorrhagic components were present in all three cases. These findings align with the data on the potential complications of sickle cell disease and support the recognition of acute soft head syndrome in the capacity of a rare but significant condition [5, 7].

Osteomyelitis is the closest differential diagnosis to acute soft head syndrome. Our case demonstrates the association between a history of sickle cell disease, skull symptoms, and radiological features such as subgaleal collections that lack enhancement, and the preservation of subcutaneous fat. The combination of these findings suggests a diagnosis of acute soft head syndrome rather than infection. Needless to add, the correct diagnosis leads to appropriate management and a shorter hospital stay. The management of acute soft head syndrome typically involves non-invasive symptomatic approaches such as intravenous hydration and pain relievers to address the pain and support the patient. In this case, the patient received supportive treatment for 6 days, and the swelling had resolved by then. The patient was discharged without any complications and is undergoing continued follow-up with haematology for sickle cell disease.

4. CONCLUSION

Acute soft head syndrome is an uncommon complication that occurs in patients who have sickle cell anaemia resulting in serious complications. Radiological imaging is crucial for its proper diagnosis, treatment, and management, as well as the prevention of further complications.

DATA AVAILABILITY

The data that support the findings of this study are available in our radiology information system, GE web-based, as well as the Cerner system at King Saud Medical City, Riyadh.

CONFLICTS OF INTEREST

The authors declare that there is no conflict of interest regarding the publication of this article.

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