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Case Report

Spontaneous subgaleal haematoma with severe headache in two children during acute painful sickle cell crisis

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Abstract

Sickle cell disease is an inherited disorder of haemoglobin formation, predominantly affecting individuals of African, Indian, or Mediterranean descent. Acute painful episodes frequently occur in these patients due to the blockage of small blood vessels by sickled cells, leading to tissue infarction. Two children of Nigerian descent, presented with severe headaches during a painful crisis episode with associated swelling on their scalps. CT imaging in both cases showed subgaleal haematoma which was managed conservatively in addition to managing the painful crisis with a good outcome. This report highlights the clinical presentation, diagnostic approach, and management of this uncommon complication of sickle cell anaemia.

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Quick Response Code:

Introduction

Sickle cell disease is a genetic disorder of haemoglobin formation, mainly affecting people of African or African-Caribbean, Indian, Arabian Peninsula, and Mediterranean origin.[1,2] Acute painful sickle cell episodes (also known as painful crises) are a common complication/presentation in these children and it is caused by blockage of the small blood vessels. Changes in any of these conditions may cause the cells to block small blood vessels and cause tissue infarction.

Subgaleal haematoma, a complication of sickle cell disease is a little-reported/known presentation in children with Sickle Cell Disease.[3] The cause of this is not fully elucidated but is thought to be due to altered bone and vessel-wall structure following the skull bone infarction as part of the sickling crisis. Although it is said to be an uncommon complication of sickle cell disease it is increasingly being reported in the literature. [1-5]

In this report, we present two patients of Nigerian origin with sickle cell disease who developed spontaneous subgaleal hematomas during a painful crisis episode.

Case presentation:

Case 1

A 9-year-old girl with sickle cell disease from Nigeria was on summer vacation in the UK. She was on prophylactic penicillin V and folic acid supplements. She suddenly fell ill with a history of vomiting and headaches. On admission, she was in painful distress, febrile, and hemodynamically stable with no abnormalities detected on physical examination. Laboratory values on admission were: Haemoglobin (Hb) 71 g/L (usual baseline Hb 100 g/L), haematocrit 0.21, white cell count 16.3 x 10^9/L, platelets 311 x 10^9/L. Clotting profile, urea and electrolytes, and liver function tests were within normal reference range. The malaria parasite test was negative.

She was admitted with a sickle cell painful crisis and sepsis in view of the unexplained fever. She started on IV ceftriaxone, paracetamol/ibuprofen, and IV maintenance fluid (5% dextrose in 0.9% normal saline) in line with guidelines for treating painful crises in sickle cell patients [6]. Inflammatory markers continued to rise during her stay, with a peak CRP (C-reactive protein) at 164 on day four of admission. She developed scalp tenderness with a boggy swelling in the right parietal area, measuring approximately 11 cm x 10 cm. The scalp was intact. Further history revealed no trauma to the head. She had a similar head swelling 4 years ago in Nigeria which was resolved with conservative management. During that episode, she had a sickle cell crisis and was found to have swelling in the right parietal area, which was suspected to be a bleed despite no head injury. She did not have any imaging of the head then but was advised that the swelling would resolve with time, and it did.

Two days into the present illness, she developed a severe right-sided headache, which she scored at 9-10 on pain score. There was little relief with paracetamol and non-steroidal anti-inflammatory pain killers necessitating the use of morphine. The headaches persisted for two days, prompting a CT head scan. The scan revealed a 10 cm swelling in the right parietal region, corresponding to the boggy area on examination, with no evidence of acute intracranial pathology. There was no underlying skull infarction or skull fracture reported. The right parietal subgaleal swelling was likely due to a hematoma. This swelling had not been noted previously. A neurosurgical consult at our specialist tertiary centre advised that no intervention was required for the subgaleal hematoma. She had to return to Nigeria due to school resumption. Her hematoma resolved within 4 weeks of discharge according to her mother during a telephone consultation.

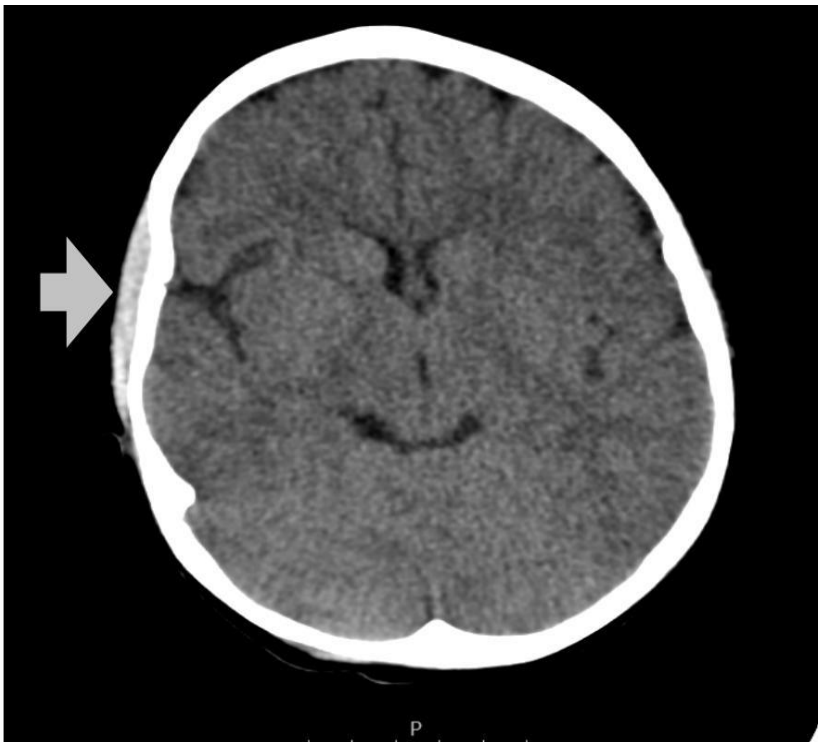

Figure 1: Computed tomography scan of head (sagittal view) showing subgaleal haematoma in right parietal region (arrowed) in 9-year-old SCA girl.

Case 2

A 14-year-old boy with known sickle cell anaemia presented to the emergency department with a 4-day history of a painless, soft swelling on the left side of his scalp. There was no prior history of falls or trauma to the head. He was on prophylactic penicillin V and folic acid supplements. A week earlier, he was managed for a painful crisis as per NICE guidelines and discharged home after two days. He notes that he had a severe headache then on the same side as the current swelling. He did not have any changes in behaviour, speech, or gait. On examination, he looked well other than a soft, cystic, non-tender 10 cm x 5 cm swelling on the left parietal region of the head. The nervous system examination did not reveal any neurological deficits.

Laboratory values on admission were: Haemoglobin (Hb) 85 g/L (usual baseline Hb 110 g/L), haematocrit 0.21, white cell count 15 x 10^9/L, and platelets 580 x 10^9/L. CRP was 260 on presentation. Clotting profile, urea and electrolytes, and liver function tests were within normal reference range.

He underwent a CT scan which showed a normal appearance of the neuro-parenchyma and ventricular system, with no acute ischemic changes, intracranial haemorrhage, or extra-axial collection seen. A left-sided scalp hematoma was noted. No skull fracture or underlying skull infarction was reported. Neurosurgical advice was sought from our specialist tertiary centre which advised conservative management. He was discharged home as he remained stable and asymptomatic. He had weekly review in the paediatric assessment unit and the swelling resolved within 4 weeks.

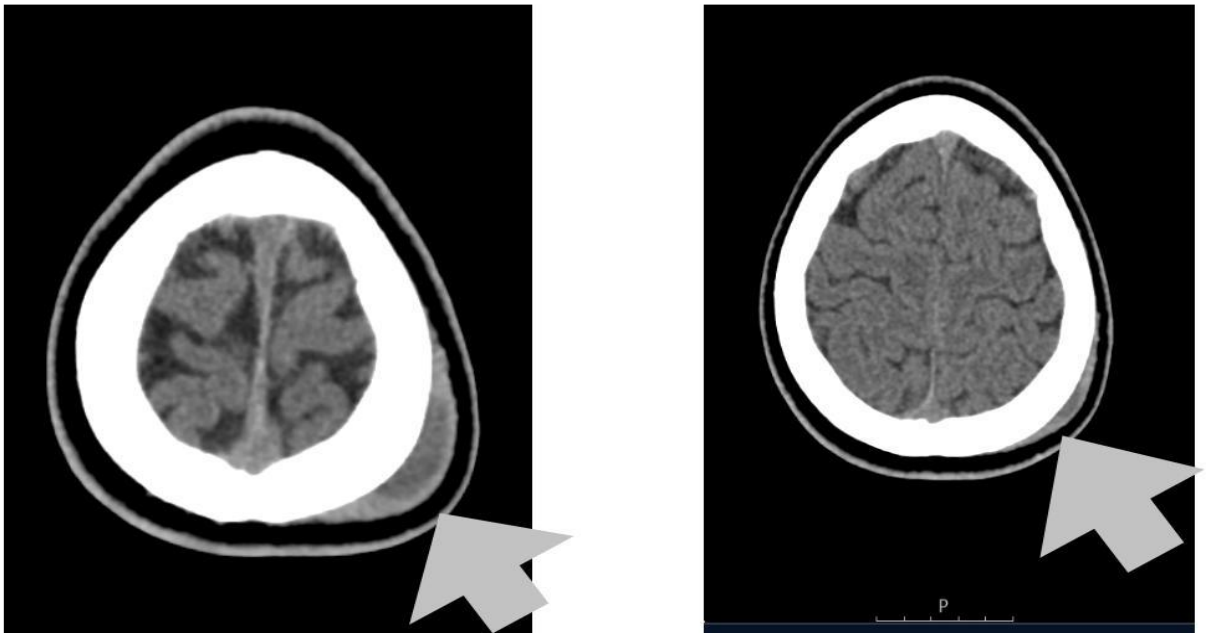

Figure 2: Computed tomography scan of the head (sagittal view) showing subgaleal haematoma (arrowed) in the left parietal region of male adolescent sickle cell anaemia patient.

Discussion:

We report two cases of spontaneous subgaleal haematoma in two sickle cell anaemia patients of African origin in a space of three months. Subgaleal haematoma is an unusual and uncommon complication of sickle cell anaemia with unclear pathophysiology. [8,9]

There are very few case reports describing the occurrence of spontaneous (non-traumatic) extradural hematomas as a complication of sickle cell disease.[4] As reported in other cases in the literature, there was no history of preceding head trauma in our cases. While the first case occurred during admission in the ward the latter case was noted a few days after being discharged from hospital following treatment for a painful sickle cell crisis. This is similar to the case report by Gonçalves et al. [7] The clotting profile in both cases were normal so, the haematoma formation was unlikely to be due to clotting derangement. Subgaleal haematoma is postulated to be caused by skull bone infarction, altered skull bone anatomy due to extramedullary haematopoiesis, and venous congestion due to sluggish blood flow in diploic veins.[8]

Both cases presented with severe headaches with attendant boggy scalp swelling during a painful sickle cell crisis. There were no neurological deficits in both patients. A fall in haemoglobin level from a steady state and raised inflammatory markers requiring antibiotics may suggest the same underlying cause of the subgaleal haematoma. Both cases had a good outcome with conservative management of the scalp haematoma, management of painful crisis as per national guidelines and complete resolution of the scalp swelling within 4 weeks. This is in keeping with other documented cases in the literature and management of scalp haematoma in sickle cell anaemia. [9,10,11]

Timely brain imaging is crucial in patients with severe headaches during a sickle cell crisis to distinguish subgaleal hematoma from other serious intracranial pathologies that complicate sickle cell anaemia [5, 12]

Conclusion:

Spontaneous (non-traumatic) subgaleal haematoma is a rare but not uncommon complication of sickle cell anaemia in children. Clinicians should maintain a low threshold for brain imaging in patients presenting with severe headaches during a sickle cell crisis to identify this condition and differentiate it from other potentially sinister intracranial causes of headaches. Early recognition and conservative management do lead to favourable outcomes, as demonstrated by our cases.

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