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Sickle cell crisis: A crisis of a different sort?

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A 9-year-old boy with sickle cell disease (SCD) was presented to the emergency department with acute headache and swelling over his bilateral temporoparietal region. There was no history of antecedent trauma, fever, vomiting or other features of an intercurrent illness. On arrival, his blood pressure was 112/62 mm Hg, heart rate was 98/min and his Glasgow Coma Scale score was 15/15. There was evidence of significant scalp tenderness over the bilateral temporoparietal region. A complete neurological examination including direct and consensual pupillary response was unremarkable. Initial investigations revealed haemoglobin of 9.6 g/dL, leucocyte count of 6.8/mm^3, platelet count of 219/mm^3 and a normal coagulation profile. His current medications included hydroxyurea and penicillin prophylaxis. He underwent an urgent CT of the head followed by MRI of the brain, which revealed abnormalities as depicted in figures 1,2.

QUESTION 1
Is this one of the most common neurological presentation seen in sickle cell crisis?

QUESTION 2
How common is this presentation in paediatric SCD?

QUESTION 3
What is the best way to manage this child?

Answers to the questions are on page 2

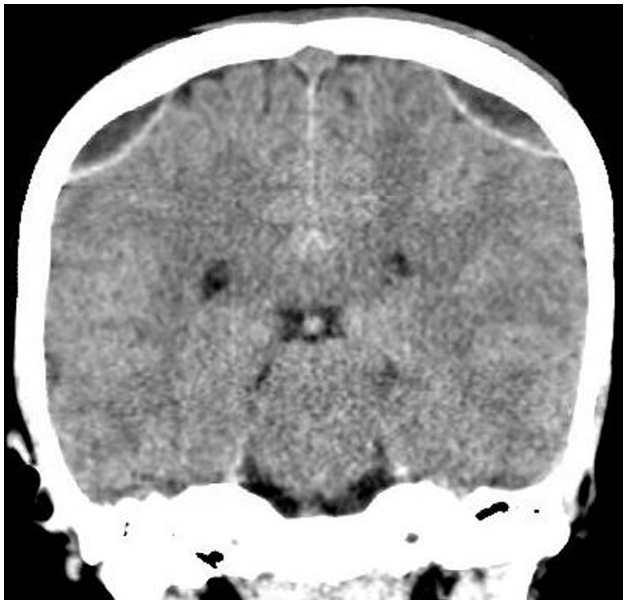

Figure 1 CT head.

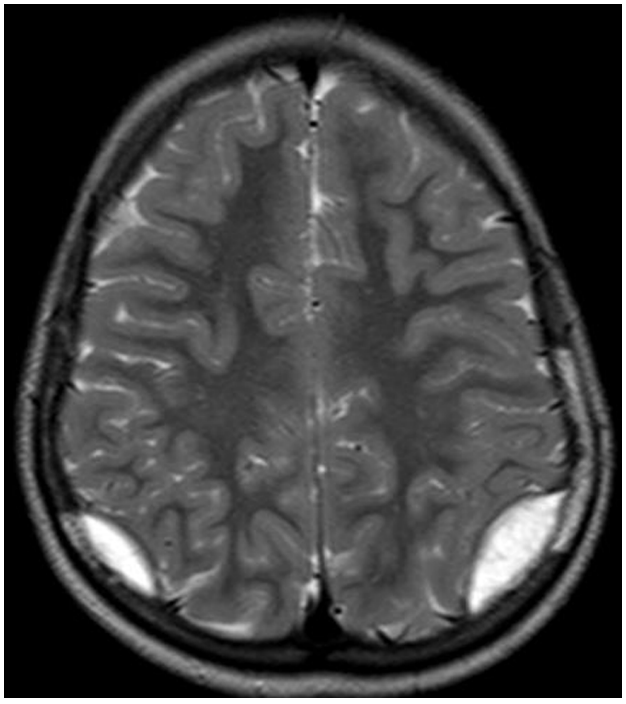

Figure 2 MRI brain.

ANSWERS TO THE QUESTIONS ON PAGE 1

ANSWER TO QUESTION 1

SCD is a very common autosomal recessive genetic disorder of red blood cells commonly affecting people of Afro-Caribbean descent. Neurological manifestations of SCD include ischaemic or haemorrhagic stroke, epilepsy, headache, myonecrosis, cognitive impairment due to chronic anaemia and silent infarcts.[1 2] About 25% of patients with SCD will have a neurological complication, with ischaemic stroke being the most commonly reported.[3] An urgent non-contrast CT head of our patient revealed bilateral extradural collection showing hypodense fluid with peripheral rim of hyperdensity suggesting subacute extradural haematomas (EDHs). Non-traumatic EDHs are extremely rare and are most commonly attributed to infectious causes and coagulopathy. A literature review of 22 cases with SCD and non-traumatic EDHs revealed acute presentation at a mean age of 15 years (range 2–35 years), with frontal and parietal areas being most commonly affected. Fifty per cent of these patients needed acute surgical evacuations of EDHs to prevent further neurological complications.[4]

ANSWER TO QUESTION 2

Skull infarction and EDHs are extremely rare complications with only a handful of such cases reported in the literature. Various pathophysiological mechanisms of the non-traumatic EDHs in SCD have been hypothesised as below; however, sequence of events or direct causation is poorly explained[5]:

- Infarction of the bone causing elevation of the periosteum and disruption of the cortical bone margin and bleeding into the epidural space.
- Insufficient venous drainage which may lead to oedema and haemorrhage.
- Chronic medullary haematopoiesis culminating in abnormal anatomy of the skull, thereby disrupting the inner and outer table leading to extravasations of blood into the subgaleal and epidural space.

The axial T2-weighted MRI brain in our patient revealed bilateral EDHs measuring 0.9×2.5×1 cm on the right and 3.7×1.5×1.2 cm on the left side. The overlying parietal bones on both sides and a small foci in the left frontal bone showed abnormal signal on T2-weighted imaging with mild restricted diffusion on diffusion-weighted imaging suggestive of bone infarction.

ANSWER TO QUESTION 3

Most of the patients respond well to conservative management.[4 6] However, a minority of patients may require urgent neurosurgical intervention if there are signs of raised or worsening intracranial pressure.[4] Our case responded well to conservative management with intravenous fluids, oxygen and opioid analgesics, and repeat neuroimaging after 48 hours did not show any enhancement of the EDHs. He was continued on hydroxyurea, analgesics and discharged uneventfully. Hydroxyurea is useful in the long-term management of SCD as it prevents stroke in children by maintaining transcranial Doppler velocities, but is not helpful in acute management of arterial ischaemic stroke or EDHs. Exchange transfusion remains the mainstay in management of acute stroke in SCD with an aim to reduce H*bS* % to ≤30%.[7] Paediatricians must be aware of this rare but serious presentation, as early recognition and management may prevent potentially serious complications.

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Contributors GK was involved in writing the manuscript. AAAM was involved in managing the patient and data collection. APSS finalised the manuscript and is guarantor of the paper.

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