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/mm³, platelet count of 219/mm³ 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.
Epilogue

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. About 25% of patients with SCD will have a neurological complication, with ischaemic stroke being the most commonly reported. An urgent non-contrast CT head scan is important to rule out intracranial blood. The presence of any enhancement of the EDHs indicates infection. Our case responded well to conservative management. However, exchange transfusion remains the mainstay in management of acute stroke in SCD with an aim to reduce HbS % to ≤30%. 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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REFERENCES

7 Nevitt SJ, Jones AP, Howard J. Hydroxyurea (hydroxycarbamide) for sickle cell disease. Cochrane Database Syst Rev 2017;106.
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