

Case report

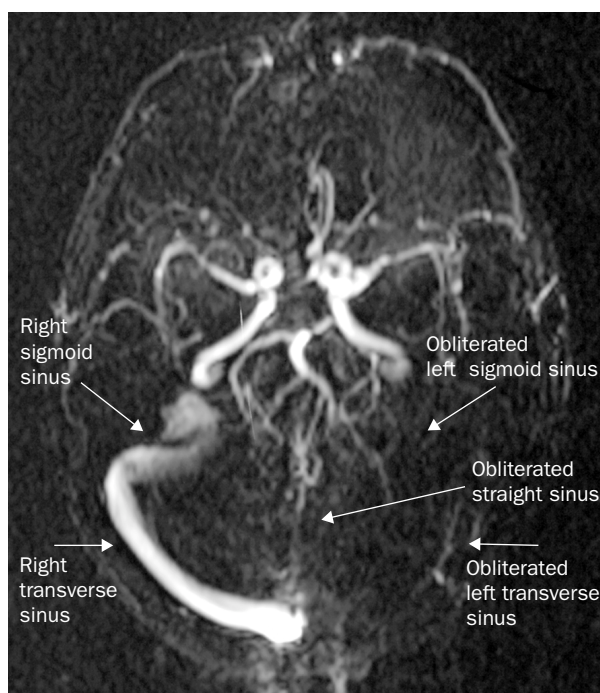
An unconscious girl with sickle-cell disease

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A 12-year-old girl, who had been adopted from Colombia, with homozygous sickle-cell disease was admitted in November, 2001, to the department of haematology for the evaluation of severe pain in her thoracic spine. Physical and radiological examination showed no abnormalities. Her haemoglobin was 6.6 g/dL and HbS content was 90.9%. She was treated with rehydration, morphine, and a blood transfusion (post-transfusion Hb 9.0 g/dL) to relieve a presumed sickle-cell crisis. Two weeks after admission, she complained of progressive headaches and nausea, accompanied by vomiting and drowsiness. Two days later, she had two short generalised convulsions with deviation of the eyes to the right. She became increasingly unconscious and could not move her right arm. A computed tomogram showed a small cortical haemorrhagic infarction in her right frontal lobe. Magnetic resonance imaging showed a second area of cortical ischaemia and haemorrhage in her left parietal lobe.

Magnetic resonance venography (figure) showed complete thrombosis of the straight, left transverse, and sigmoid sinus and an irregular, partial obliteration of the frontal part of the superior sagittal sinus. Later that day she was found to have mild bilateral papilloedema and a left-sided VI cranial nerve palsy. Exchange transfusion was started to prevent further sickling and nadroparin calcium was given for three weeks. Two days after the first seizure the patient had made a full neurological recovery. She was treated with warfarin for another three months and with blood transfusions to guarantee an HbS content below 30%. She is reviewed monthly by a haematologist.

We report an unusual cause of neurological deterioration in a patient with sickle-cell disease, in which occlusive or haemorrhagic stroke occurs in up to 11% of patients before the age of 20 years.¹ Seizures and hemiparesis in our patient could have been due to a stroke. The alteration of consciousness, however, together with signs of raised intracranial pressure suggested a multifocal disorder and, taking into account her previous headache and vomiting, led to the suspicion of sinus thrombosis.² Although rare in children, sinus thrombosis has been associated with ear, nose, and throat infections, dehydration, connective tissue disease, cancer, cardiac disease and prothrombotic state,² but only once has been described in sickle-cell disease.³ When erythrocytes sickle, they adhere to the vessel wall, resulting in tissue ischaemia.⁴ Beside erythrocyte abnormalities, activation of



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the coagulation system such as increased antiphospholipids and decreased protein C and S contribute to thrombogenesis and subsequent vascular occlusion.⁵ Tests for prothrombotic disorders in our patient did not find any abnormalities except for increased factor VIII activity (>400%), which may have contributed to venous thrombosis. Cerebral sinus thrombosis is a difficult clinical diagnosis, which requires a high degree of suspicion and specific neuroimaging techniques. Although rare, sinus thrombosis should be considered in the differential diagnosis of patients with sickle-cell disease and neurological symptoms.

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