Cerebral venous sinus thrombosis: a diagnosis not to be missed

Introduction

Cerebral venous sinus thrombosis usually affects young to middle-aged people and has potentially serious consequences with mortality between 5.5 and 30%.

Discussion

The most common predisposing risk factors for cerebral venous sinus thrombosis (Renowden, 2004) are pregnancy and the puerperium, oral contraceptives and coagulopathies; approximately 25% of cases remain idiopathic. This patient was taking black cohosh (a source of plant oestrogens) and had a family history of thromboembolism and systemic lupus erythematosus. Further investigations revealed heterozygosity for the prothrombin gene variant.

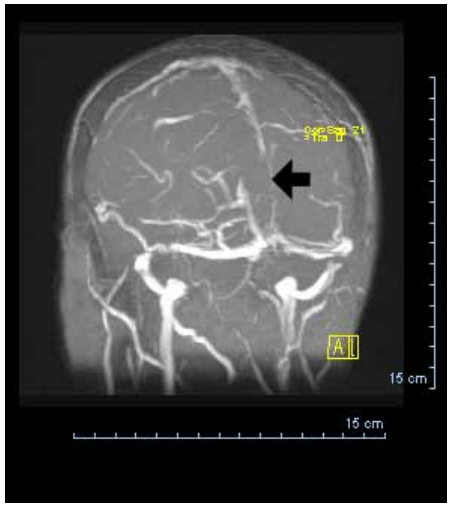
The most commonly affected dural sinuses are the superior sagittal sinuses (70–80%), transverse and sigmoid sinuses (70%) and, less frequently, the cavernous and straight sinuses. Presentation may be acute (28%, usually in infectious or obstetric conditions), subacute (41%) or chronic (31%, usually in inflammatory diseases), depending on the extent of the venous collateralization.

Headache is often the first and most frequent symptom occurring in 74–90% of all patients (Tessitore et al, 2001). Other reported presenting symptoms and signs include papilloedema (41%), nausea and emesis (61%), seizures (37–47%), impaired consciousness (19–39%), coma (15%) (Benveniste et al, 2004) and cranial nerve palsies.

The prevalence increases in inherited primary thrombophilia that can trigger prothrombotic events in any vascular bed. Cerebral venous sinus thrombosis has been associated with prothrombin gene mutation; G20210A variant (Martinelli et al, 1998) is the second most frequent prothrombotic polymorphism in Caucasians and has been reported in association with myocardial infarction, cerebral infarction in children (Gurgey et al, 2005), multiple intracardiac thrombosis (Cinar et al, 2005), dural sinus thrombosis 3 months post-partum, pulmonary embolism and multi-organ failure caused by mesenteric and inferior vena cava thrombosis, and central retinal vein occlusion.

Heparin is a first-line treatment for cerebral venous sinus thrombosis because of its efficacy, safety and feasibility, fol-

Figure 1. Deep venous thrombus in the superior sagittal sinus.



Dr Komal E Imtiaz *is Consultant Physician in the Department of Endocrinology/ Diabetes, Lancashire Teaching Hospital NHS Foundation Trust, Chorley Hospital, Chorley PR7 1PP,* **Dr Sanjay Rajpara** *is Consultant Dermatologist in the Department of Dermatology, Aberdeen Royal Infirmary, Aberdeen and* **Dr Tahir Majeed** *is Consultant Neurologist in the Department of Neurology, Lancashire Teaching Hospital NHS Foundation Trust, Chorley Hospital, Chorley*

Correspondence to: Dr KE Imtiaz

Case Report

A 47-year-old woman was admitted with a severe headache of 5 days' duration, vomiting and collapse with transient loss of consciousness in December 2002. She had been taking black cohosh, a herbal preparation containing plant oestrogen for menopausal symptoms, for 2 years. Her father had a history of pulmonary embolism and her sister had systemic lupus erythematosus. Clinical examination was normal except for slight neck stiffness and photophobia. Brain computed tomography was normal and lumbar puncture revealed normal CSF; CSF pressure was not recorded. Her symptoms improved slowly and she was discharged home with an appointment with the neurologist arranged as an outpatient.

Five days post-discharge from the hospital, she was readmitted because of the persisting headache and was found to have papilloedema on neurological examination. Chest X-ray, full blood count, erythrocyte sedimentation rate, clotting screen, urea and electrolytes, liver function tests, serum glucose, immunoglobulins, complement and autoantibody screening were normal or negative. Magnetic resonance imaging and venogram showed deep venous thrombosis in the superior sagittal sinus (*Figure 1*). She was anticoagulated with heparin followed by warfarin. Her headaches settled and she was discharged home 8 days later.

Further investigations confirmed heterozygosity for the prothrombin gene variant (G20210A). She continued to describe 'pressure headaches' with tinnitus. Repeat brain magnetic resonance imaging and venogram was normal in 2004. Her headaches recurred and the diagnosis of secondary intracranial hypertension was made and acetazolamide and bendrofluazide were commenced. Further magnetic resonance imaging of the brain with venogram in September 2007 revealed a hypoplastic left transverse sinus and patent superior sagittal sinus. She was referred to the neurosurgeons and had a lumbo-thecal peritoneal shunt insertion in November 2007 with improvement in her symptoms, despite a small frontal subdural collection on brain computed tomography postoperatively, which had resolved on repeat magnetic resonance imaging in March 2008. She is regularly followed up in the neurology and ophthalmology clinics for residual bilateral papilloedema and left amblyopia and continues on ramipril, bendrofluazide and warfarin.

lowed by warfarin therapy (lifelong in cases of inherited coagulopathies). Cerebral venous sinus thrombosis may present with intra-cranial haemorrhage and anticoagulation is still considered in these patients, however, more clinical trials are required to evaluate long-term outcomes in this sub-group of patients.

Local thrombolysis may be indicated in cases where deterioration occurs despite adequate heparinization, management of raised intracranial pressure and seizure prophylaxis. Neurosurgical management may involve CSF drainage techniques, decompressing craniotomy, endovascular local infusion of thrombolytics or mechanical thrombectomy to treat the venous sinus thrombosis. Early diagnosis is crucial since anticoagulation may reduce the risk of a fatal outcome and severe disability without promoting intracranial haemorrhage. **BJHM**

Benveniste RJ, Patel AB, Post KD (2004) Management of cerebral venous sinus thrombosis. *Neurosurg Q* **14**(1): 27–35

Cinar A, Dilber E, Karagoz T, Bahadir N, Gucer S, Tekinalp G, Gurgey A (2005) Association of prothrombin gene mutation with sepsis in a preterm with multiple intracardiac thrombi. *Echocardiography* **22**(4): 340–4

Gurgey A, Unal S, Okur H, Duru F, Gumruk F (2005) Prothrombin G20210A mutation in Turkish children with thrombosis and the frequency of prothrombin C20209T. *Pediatr Hematol Oncol* **22**(4): 309–14

Martinelli I, Sacchi E, Landi G et al (1998) High risk of cerebral vein thrombosis in carriers of a prothrombin gene mutation and in users of oral contraceptives. *N Engl J Med* **338:** 1793–7

Renowden S (2004) Cerebral venous sinus thrombosis. *Eur Radiol* **14:** 215–26

Tessitore E, Schonauer C, Fera F et al (2001) Superior sagittal sinus thrombosis as unusual cause of headache: case report. *J Headache Pain* **2:** 97–9