Answer

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The answer to this Diagnostic Challenge is C: Straight sinus thrombosis.

Cerebral venous sinus thrombosis (CVST) was first described in 1825. *Primary* CVST is a rare disorder that may occur at any age,[1] often associated with some form of coagulation disorder. Our patient was subsequently diagnosed with two coagulation disorders: prothrombin variant G20210-->A and factor V Leiden, both of which have been reported previously.[2,3] Sex hormones may also play a role, as primary CVST has been reported in association with oral contraceptive use,[4] during pregnancy and in the postpartum period. No explanation is found in approximately 30% of primary cases. *Secondary* CVST has been reported with infections such as meningitis and mastoiditis, as well as with head trauma, metastasis, severe dehydration and inflammatory diseases such as ulcerative colitis.[5]

The clinical presentation can vary. Headache is usual, but may occasionally be absent.[5] Neurological findings such as unilateral weakness or sensory loss, cranial nerve palsies, cognitive impairment, amnesia, confusion and mutism have all been described. More severe cases may have papilledema, altered consciousness, seizures, coma and death.

Diagnosis is often first made by CT (computed tomography).[6] A non-contrast study may show the classic *cord sign* that represents a high-density clot in a cortical vein, as seen in Figure 1. Contrast enhanced scanning may reveal the *empty delta sign*, a filling defect due to clot within the sagittal sinus. Other CT findings are described, but CT alone may occasionally miss the diagnosis.[6]

Either MRI (magnetic resonance imaging) or MR angiography (MRA) is the optimal diagnostic modality.[7] MRI visualizes clot and may reveal filling defects in the sinuses. MRI is superior to CT for detection of brain edema and infarction, allowing differentiation between venous and arterial infarcts. In this case a 2-dimensional time-of-flight MRA (Fig. 2) showed complete thrombosis of the superior sagittal sinus, straight sinus and transverse sigmoid sinuses bilaterally.

Treatment for CVST has traditionally included anticoagulation and, more recently, thrombolyis.[8] An attempt at lysis was made in this case with a direct infusion of urokinase into the right transverse sigmoid sinus, without success. Nonetheless, the patient eventually improved and was discharged on warfarin. Other treatments for the complications of CVST may include corticosteroids for edema, shunting and acetazolamide for hydrocephalus, and anticonvulsants.

The prognosis of CVST is variable, and outcomes may range from complete recovery to death.[1] Long-term seque-

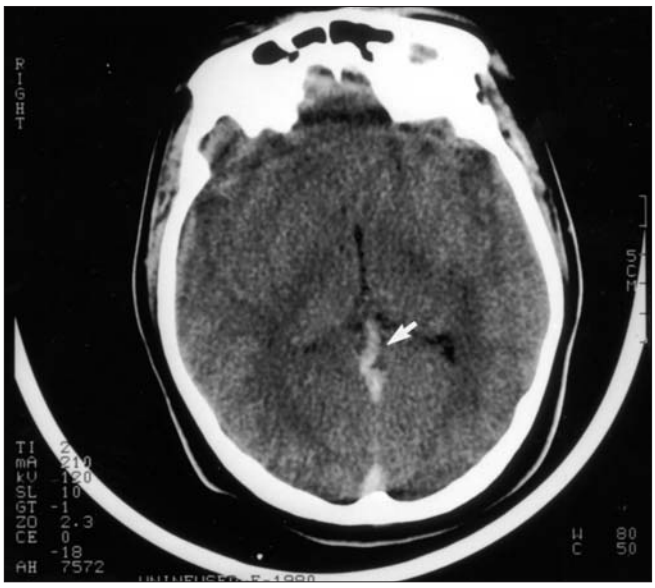

Fig. 1. Non-enhanced computed axial tomography showing high density clot in the sagittal sinus (arrow).

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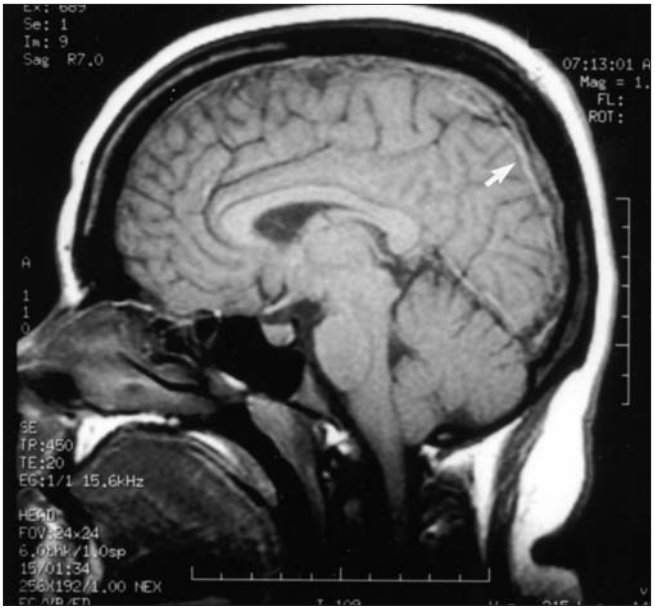

Fig. 2. Sagittal magnetic resonance imaging study showing clot within the sagittal sinus (arrow).

lae may include cognitive impairment, seizures, and intracranial hypertension. Factors related to poor outcome include presence of papilledema, altered consciousness or coma, extremes of age, a delay in diagnosis, intracerebral hemorrhage, and involvement of the straight sinus or cerebral veins.

Competing interests: None declared.

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