Unusual presentation of more common disease/injury

Progressive seizures in a patient with congenital coagulopathies

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Dural arteriovenous fistula (DAVF) is a rare complication of cerebral venous sinus thrombosis (CVST). DAVFs develop as a result of direct arterial to venous sinus communications evolving in response to an occluded sinus. The authors present a patient with Down's syndrome who developed progressive, uncontrolled seizures and chronic CVST secondary to factor V Leiden deficiency. Brain MRI and computerised tomographic venography revealed a complex secondary DAVF, which once embolised resulted in a dramatic reduction in seizures and improvement in the clinical state. DAVFs should be considered as a potential complication in patients with persistent features of CVST.

BACKGROUND

Cerebral venous sinus thrombosis (CVST) may present with any of three major syndromes: isolated intracranial hypertension resulting in headache, a focal syndrome such as seizures, and/or as an encephalopathy. Although the presentation and course are variable, the prognosis following treatment is usually favourable. However, in patients who continue to show neurological decline despite treatment, transformation to an acquired dural arteriovenous fistula (DAVF) should be considered as the management of this has a potentially favourable outcome.

CASE PRESENTATION

Our patient is a 46-year-old woman, with Down's syndrome, who was referred with a 12-month history of gradual onset progressive headache associated with

seizures and cognitive decline. Brain MRI revealed extensive venous collaterals on the surface and parenchyma of the brain (figure 1A). These changes were thought to be secondary to chronic CVST, subsequently confirmed on CT venogram (figure 1B). The patient was investigated for coagulopathies and was found to have both factor V Leiden and protein S deficiencies. The patient was commenced on carbamazepine, and anticoagulated with warfarin.

Despite optimal anticoagulation therapy, the patient continued to have persistent seizures, and progressive cognitive and functional decline. Repeat MRI of the brain revealed new high signal change in the white matter of the right cerebral hemisphere and a multiplicity of dilated blood vessels suggestive of a DAVF (figure 2A). A digital subtraction angiogram was performed to characterise the

Figure 1 (A) Axial T2-weighted MRI demonstrating prominent cortical veins in the right cerebral hemisphere consistent with chronic venous sinus thrombosis. (B) Coronal reconstruction CT venogram confirming the above changes consistent with CVST.

Figure 2 (A) Axial T2-weighted MRI and (B) axial CT venogram reconstruction showing multiple dilated veins. There is also oedema in the right temporal and occipital white matter.

Figure 3 Pre- and postembolisation angiograms. Lateral view from a right common carotid artery angiogram demonstrating an occluded right sigmoid sinus. There is a dural arteriovenous fistula involving the right sigmoid sinus supplied by the branches of the right external carotid artery. There is retrograde drainage into the inferior anastomotic vein and into several cortical veins. Postembolisation AP right common carotid angiogram showing coils occluding the right sigmoid sinus with complete obliteration of the fistula. Lateral view from a left common carotid angiogram showing that the patient has a small dural arteriovenous fistula involving the superior sagittal sinus antegradely into the superior sagittal sinus.

Table 1 Predictors of poor long-term prognosis in CVST²

Central nervous system infection
Any malignancy
Thrombosis of the deep venous system
Haemorrhage on head CT or MRI
Glasgow Coma Scale Score <9 on admission
Mental status abnormality
Age >37 years
Male gender

abnormality. This revealed an occluded sigmoid sinus with an adjacent DAVF. Cortical venous reflux was evident, indicating this to be a lesion associated with a high risk of spontaneous intracranial haemorrhage. The DAVF was successfully embolised with coils using a direct transcranial approach to the transverse sinus after failed transfemoral approaches (figure 3A–C).

OUTCOME AND FOLLOW-UP

Following treatment, her focal neurology and seizure frequency improved, which has significantly enhanced her quality of life.

DISCUSSION

The overall incidence of CVST is <1/100 000.¹ This patient presented with headache and seizures, which occur in 89% and 37% of patients with CVST, respectively.² These symptoms are thought to be due to the increased venous pressure caused by the obstruction of the venous structures. Women are more likely to have headache than men,³ and seizures are frequently recurrent.

The major risk factors for CVST in adults include prothrombotic conditions, oral contraceptives, pregnancy, malignancy and infection. In one cohort study, a prothrombotic condition, either genetic or acquired, was found in 34% of patients.² Screening for a thrombophilic state is, therefore, recommended in all patients unless a clear risk factor is identified. Both factor V Leiden and protein S deficiency were found in our patient, for which anticoagulation therapy was commenced.

About 23% of patients with CVST deteriorate after presentation.⁴ This is in terms of a depressed level of consciousness, new seizures or a worsening of, or development of, new focal deficits. Neuroimaging at this stage shows new parenchymal lesions in one-third of patients as in our case.⁴ A number of predictors of poor long-term prognosis were identified in our patient, including altered mental state, a depressed level of consciousness on admission and her age² (table 1).

Catheter angiography is the gold standard for diagnosis, treatment planning and follow-up of patients with DAVF.⁵

The commonest presentation of a DAVF is with pulsatile tinnitus. However, a more aggressive DAVF may manifest as focal neurological deficits.⁶ The angioarchitecture of a DAVF is direct arterial to venous connections without an intervening capillary network. These channels appear to open as an acquired feature following a venous sinus thrombosis. An intraluminal thrombosis causes ‘growth’ of the dural arteries normally present in the walls of the sinuses. This abnormal collateral pathway may result in a direct communication between artery and vein, resulting in a fistula formation.

The risk of an intracranial bleed from a DAVF is determined by the angioarchitecture. DAVFs are treated differently from CVST. Higher grade DAVFs, which drain retrogradely into cortical veins, have a significant risk of haemorrhage. The mainstay of treatment is surgical or endovascular therapy. Lower grade lesions, which have a low haemorrhage risk, are often managed conservatively. Radiosurgery is a useful adjunctive therapy in selected cases.^{5 7}

Learning points

- ▶ For the vast majority of patients, prognosis in CVST is favourable.
- ▶ Although rare, CVST can transform to a DAVF.
- ▶ Once aetiology for the thrombosis has been found and treated, further neurological deterioration in a patient should raise the suspicion of secondary DAVF.

Competing interests None.

Patient consent Obtained.

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