CASE REPORT

Dural arteriovenous fistula (D-AVF): a rare cause of symmetrical bithalamic changes with free diffusivity changes

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SUMMARY

We report a rare case of an encephalopathic presentation of a posterior fossa dural arteriovenous fistula with concomitant florid symmetrical bithalamic free diffusivity changes on MRI in a previously healthy individual. We describe the structural and functional imaging findings and the role of catheter angiography in diagnosis, prognostication and the timing and strategy in the management of this challenging vascular entity.

BACKGROUND

Symmetrical bithalamic hyperintensity on MRI is an unusual appearance with a limited but well-described differential diagnostic list including terminal basilar artery thrombosis, deep venous thrombosis, infiltrative tumor and certain encephalitides (including encephalitis lethargica). Imaging findings of dural arteriovenous fistulae (D-AVF) causing changes restricted to the thalamus are extremely rare. Two previous reports have described similar MRI findings^{1 2} and a further case report demonstrated CT imaging of changes isolated to the thalamus and attributed to a D-AVF.³

We report a rare case of a posterior fossa D-AVF presenting with an encephalopathic picture, with florid symmetrical bithalamic T2 high signal change on MRI, with free diffusivity on diffusion-weighted imaging/apparent diffusion coefficient (DWI/ADC) maps. The imaging appearances were attributable to engorgement and hypertension of the deep venous system as a result of a D-AVF and concomitant occlusion of the mid-portion of the straight sinus. We describe the subtle structural changes on the initial presenting MRI and the subsequent findings on the intracranial CT angiogram (CTA).

CASE PRESENTATION

A right-handed middle-aged patient presented with a shuffling gait, somnolence, hiccoughing and diplopia; the latter resulted in a road traffic accident and his subsequent admission. Travel history included a recent holiday to Europe. There was no significant past medical history.

The initial examination showed mildly dysfluent but appropriate speech. Higher cerebral functions were normal, however the patient had a slow and dyspraxic gait. Power, sensation and cranial nerve examination were entirely normal.

A structural MRI study showed symmetrical bilateral T2 hyperintensity limited to the thalami (figure 1A), with no abnormal parenchymal enhancement evident. The spinal fluid was acellular with an elevated protein value of 1.39 mg/mL and glucose of 3.6 mg/mL, but no xanthochromia, organisms or malignant cells were identified. Oligoclonal bands were negative. Blood investigation was normal. EEG showed no background abnormal activity and no periodic discharges.

Over the following days there were periodic exacerbations in cognitive decline and a drop in the mental test score from 30/30 to 18/30. The patient also developed extrapyramidal signs with limb rigidity, bradykinesia, and increasingly dysarthric speech and a new left facial palsy. The gait further deteriorated, in keeping with thalamic astasia, and developed frontal release reflexes, utilization behavior and extensor plantar responses.

Further MRI brain imaging showed progression of symmetrical thalamic increased T2 signal, with corresponding free diffusivity on the DWI/ADC maps in keeping with vasogenic (interstitial) edema. On review of the imaging at the Regional Neurosciences Centre, a prominent vein within the left ambient cistern and possible straight sinus occlusion were noted. The possibility of a D-AVF was raised. A CTA and subsequent catheter cerebral angiogram were performed, confirming the diagnosis of a posterior fossa D-AVF with accompanying mid-straight sinus occlusion (figure 2). Surgery was planned but, in the interim period, the patient had a left basal ganglia hemorrhage. Despite emergency surgical occlusion of the D-AVF and removal of the hematoma via a posterior fossa craniotomy, the patient had a further fatal left basal ganglia hemorrhage.

Post-mortem examination showed the recent basal ganglia hemorrhage and a complex, predominantly left-sided, extensive posterior fossa vascular abnormality involving the cerebellum and left thalamus with a complex branching architecture and notably irregular muscle wall thickness, presumed to be a result of the arterIALIZATION of the venous system secondary to the D-AVF.

DISCUSSION

The finding of symmetrical bilateral thalamic T2-weighted hyperintensity on MRI has a relatively small differential diagnosis which can be divided into vascular and non-vascular etiologies. The major vascular entities include terminal basilar

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Figure 1 (A) Axial gradient T2-weighted image (FLIP 90°, TR 5035 ms, TE1000 ms) and (B) apparent diffusion coefficient (ADC) map (b1000) at the level of the diencephalon showing symmetrical bithalamic T2 hyperintensity and corresponding high signal on ADC mapping, in keeping with free diffusion.

thrombosis and deep venous system occlusive disease; non-vascular causes include intrinsic infiltrative tumor and, rarely, diencephalic encephalitis.

We report a rare cause of bilateral thalamic hyperintensity on MRI, in which the underlying cause was a D-AVF with associated straight sinus occlusion. After a Medline search and, to the best of the authors' knowledge, there is only a single report of CT findings of D-AVF resulting in isolated thalamic changes³ and two case reports with MRI features of a D-AVF causing diffuse changes within the thalami, but not restricted to the diencephalic structures.^{1 2}

The venous drainage of the thalami has been previously studied by Stephens and Stillwell⁴ who demonstrated several groups of veins draining these structures. The superior and medial portions of the thalami drain into the internal cerebral vein or great cerebral vein and the inferior and lateral portions of the thalamus drain into the basal vein or the posterior mesencephalic vein. Four groups of veins have been described: superior, inferior, anterior and posterior.⁵ The superior thalamic vein is the largest of the thalamic veins and is formed by the convergence of several smaller vessels before exiting from the medial surface of the thalamus and draining into either the internal or great cerebral vein. The inferior thalamic veins accompany the thalamoperforating arteries and traverse the posterior perforated substance before draining into the peduncular branch of the basal vein or the posterior mesencephalic vein. The anterior

thalamic vein is a constant vessel that drains into the internal cerebral vein or the septal vein at the level of the foramen of Monro, while the small posterior thalamic veins, which empty the posterior inferolateral portion of the thalamus, drain into the basal or posterior mesencephalic veins.

Venous drainage of the striatal structures is complex and follows two pathways, one involving the internal cerebral veins via the superior striatal veins and caudate veins and the other involving the basal vein of Rosenthal via the inferior striatal veins. In our case it is likely that the venous outflow from the striatum found an alternative pathway while that from the thalami did not, with the resulting imaging appearances.

Conventional angiography remains the gold standard for the definitive diagnosis and full characterization of D-AVFs.⁶ The classification system for D-AVFs is well documented and, in our case, we recognized this as a high-grade Cognard IV, Borden 3 D-AVF. Structural imaging with MRI and intracranial CTA can be complementary in the initial diagnostic investigation, as in this case.

There are a number of endovascular, surgical or combined management strategies in the treatment of D-AVFs.^{7–11} For all patients, long-term follow-up with angiography is recommended due to the risk of fistulous reconstitution.¹²

Radiologists should be cognizant of the potential diagnosis of a D-AVF in a previously healthy, acutely presenting 'encephalopathic' patient with abnormal thalamic imaging findings and/or

Figure 2 Selective left internal carotid artery digital subtraction angiogram (DSA) mid and late arterial phase (A and B, respectively) showing early opacification of a prominent left lateral pontomesencephalic vein (black arrow), marginal tentorial artery (white arrow) supplied via the lateral clival artery and fistulous point (black arrow). There is associated retrograde opacification of the vein of Galen (black arrowheads) and internal cerebral veins (double black arrows), and failure of opacification of the straight sinus. The DSA findings are consistent with a type IV dural arteriovenous fistula.

prominent flow voids on structural imaging. Consideration of a posterior fossa dural fistula is warranted as a potential underlying pathological process in all cases of thalamic signal change, especially in patients with free diffusivity within the thalami but also in patients with more widespread changes involving other deep ganglion structures. Isolated occlusion of the straight sinus should also prompt clinicoradiological review of imaging for a possible underlying dural fistula, albeit not necessarily with a catheter angiogram.

Learning points

- ▶ Dural arteriovenous fistulae (D-AVF) often present with vague and non-specific symptoms and are therefore a challenge to diagnose.
- ▶ Radiologists should consider the potential diagnosis of a D-AVF in a previously healthy, acutely presenting 'encephalopathic' patient with abnormal thalamic imaging findings.
- ▶ Isolated occlusion of the straight sinus should also prompt consideration of a possible underlying dural fistula.

Competing interests None.

Patient consent None.

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