LESSON OF THE MONTH

Myelopathy from intracranial dural arteriovenous fistula

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Dural arteriovenous fistulas arising intracranially are an uncommon cause of progressive myelopathy. This report is of a patient in whom the diagnosis of the condition was confounded by coexisting small vessel cerebrovascular disease.

A 68 year old right handed man was admitted to his referring hospital in March 2001 with a three month history of deteriorating mobility, lower limb weakness, and recurrent falls. He had initially presented to his general practitioner with a sudden onset of unsteadiness, and a stroke was diagnosed. He failed to improve, and following a gradual but marked deterioration he was admitted to hospital. Over the course of a month in hospital his condition worsened, with recurrent collapses caused by a postural fall in blood pressure, increasing leg weakness and unsteadiness, loss of sphincter control, and difficulty in swallowing.

His past medical history consisted of a previous right sided stroke 10 years before, from which he had made a complete recovery. He had suffered two myocardial infarcts. He had essential hypertension, diet controlled hypercholesterolaemia, and was an ex-smoker. Drug treatment on admission to hospital was aspirin, dipyridamole, and various antihypertensive agents. The latter were discontinued when his hypotensive episodes were noticed, and fludrocortisone was added.

Magnetic resonance imaging (MRI) of the brain and cervical spine at the referring hospital suggested florid diffuse small vessel cerebrovascular disease involving the deep white matter of the hemispheres and brain stem, with extension to the upper cervical spinal cord. The radiological changes, in the context of his known significant vascular history, were initially thought sufficient to explain his clinical syndrome.

Examination on transfer to our unit revealed a cogent, articulate man. A significant postural fall in systemic blood pressure (an average difference of 50/30 mm Hg between the supine and sitting positions) was noted; general examination was otherwise unremarkable. Fundoscopy and eye movements were normal. Facial movements were full and symmetrical. His cough was weak, with a bovine character, but palatal movement appeared symmetrical and the gag reflex was intact. A formal swallowing assessment was satisfactory. The upper limbs were neurologically normal. The lower limbs revealed mild wasting with a moderate pyramidal weakness, more noticeable proximally. All deep tendon reflexes were brisk without clonus, and the plantar responses were flexor. A urinary catheter was in situ. There was no sensory disturbance. He was unable to sit unsupported because of truncal weakness.

Neuroradiological review of his original cranial MRI confirmed widespread, confluent areas of T2 signal prolongation in the hemispheric white matter (fig 1A); these were unusually extensive in the brain stem (fig 1B). The latter changes were noted to extend into the cervical cord on both sagittal and axial T2 sequences (not shown). A sagittal T1 weighted view of this region (fig 2A) revealed an irregular contour of the medulla and cervical cord, suggestive of vascular flow voids. These were confirmed on a contrast enhanced examination (fig 2B), which revealed a network of enlarged vessels enmeshing the lower brain stem and cervical cord. Angiography of the carotid and vertebral arteries was performed. This revealed a dural arteriovenous fistula of the clivus fed by clival and meningeal branches of the intracranial left internal carotid artery (fig 3). There was venous drainage into enlarged and tortuous perimedullary veins on the anterior aspect of the brain stem, and thence to the anterior and posterior perimedullary venous plexuses of the cervical spinal cord (not shown). The fine calibre of the feeding vessels precluded endovascular treatment of the arteriovenous fistula.

Following neurosurgical referral, the arterialised draining vein was clipped just beyond the arterial feeders. The patient
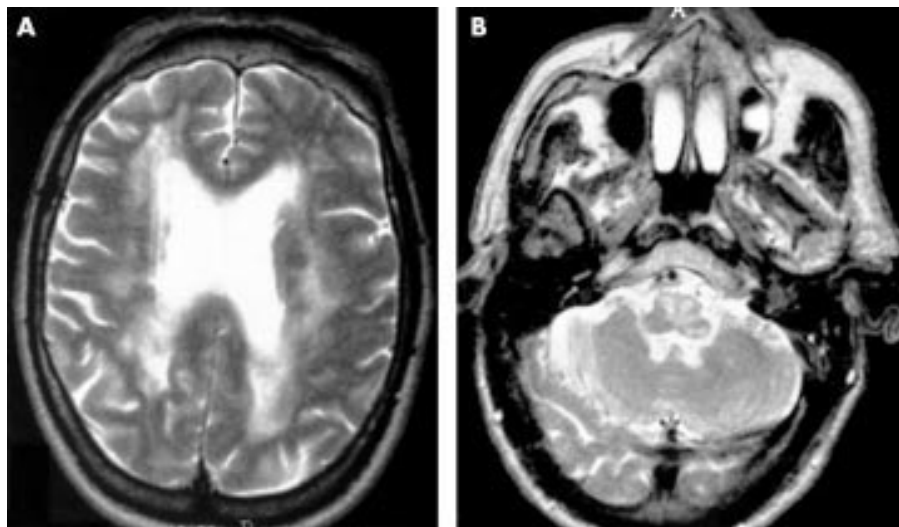

Figure 1 T2 weighted axial sequences through the hemispheres (A) and medulla (B) showing confluent areas of high signal change. The florid abnormality in the latter (which was continuous with similar appearances in the pons and cervical cord) is unusual for small vessel cerebrovascular disease.
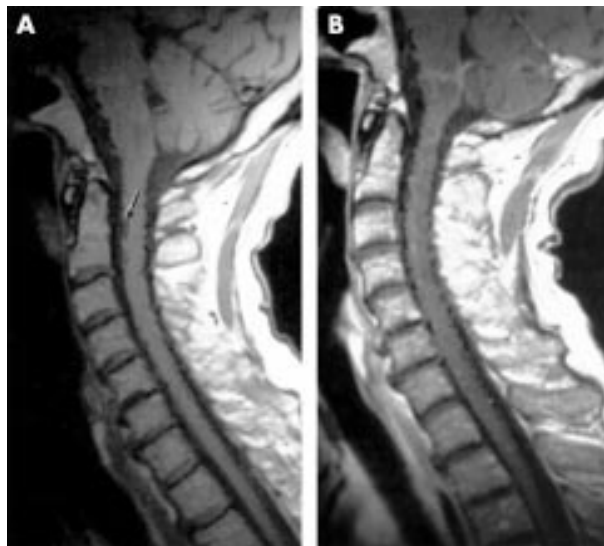
Figure 2 T1 weighted sagittal sequence (A) through the posterior fossa and cervical cord. Note the irregular appearance of the anterior surface (arrow), suggesting vascular flow voids. Contrast enhanced examination (B) confirmed these; a network of enlarged vessels is seen enveloping the lower brain stem and cervical cord.
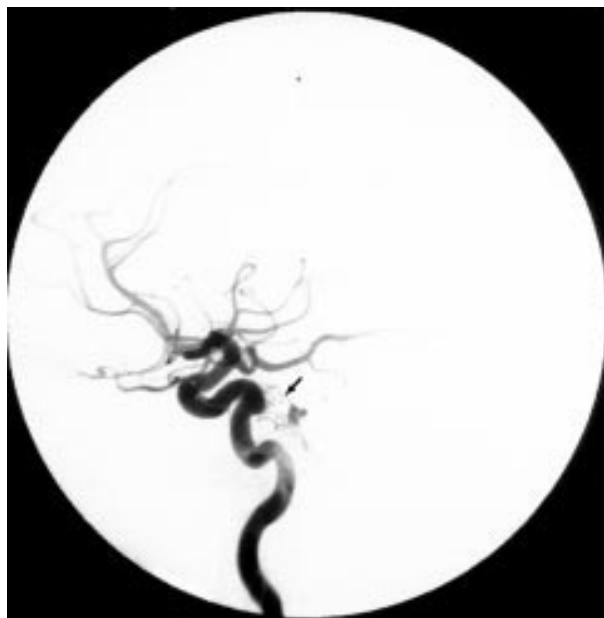

Figure 3 Cerebral angiography: left internal carotid injection. A cluster of fine calibre abnormal vessels (arrow) is seen arising from the terminal portion of the carotid in the early phase. Late phase (not shown) revealed drainage caudally from veins on the anterior brain stem into the anterior and posterior perimedullary venous plexuses.

was subsequently discharged to a rehabilitation unit, where his initial progress was promising: four months after operation (October 2001) he was able to sit unaided and was starting to walk with support. The ferromagnetic character of the surgical clip has precluded postoperative MRI.

DISCUSSION

Progressive myelopathy from an intracranial arteriovenous fistula with spinal perimedullary venous drainage is a rare entity—our patient appears to be only the 15th case to be reported. Anatomically, the condition seems to arise as an exaggeration of the normal anastamoses between the pontomesencephalic and the anterior and posterior spinal veins.[1] This may be provoked by the thrombotic occlusion of a draining intracranial venous sinus, although there was no history of this in our patient, nor any suggestive angiographic evidence. Thus, while a dural spinal arteriovenous fistula would usually obtain its arterial supply from that of the spinal cord itself, it is occasionally possible for the nidus to arise intracranially. The arterialised spinal veins lead to congestive oedema and subsequent ischaemia of the spinal cord, causing the symptoms.

Fourteen other patients with myelopathy caused by intracranial dural arteriovenous fistulae have been reported.[2][7] There is a male preponderance (male to female ratio, 13:2), and the mean age of onset is 55.5 years. In most previous cases there was considerable difficulty and delay in establishing a definitive diagnosis before contrast enhanced MRI and cerebral angiography were undertaken. Cranial MRI was either not performed or was normal in all cases. Endovascular embolisation, either alone or followed by surgery, was the treatment used. The delay before diagnosis has perhaps skewed the prognosis of the condition—only four patients are documented as having made a recovery to independent ambulation. Two patients died: one from the series of five reported by Gobin et al,[5] who died postoperatively, and one from a series of three reported by Partington et al,[4] who died of unrelated causes. The remaining eight are reported to have been left disabled, though treatment presumably halted any further decline.

Patients with a suspected spinal arteriovenous malformation will usually have spinal angiography. However, as the authors of the above case series emphasise, angiographic studies must include the cranial vasculature when spinal studies are normal or if the abnormality on MRI is maximal in the upper spinal cord. Apart from reiterating this diagnostic pitfall, our main objective has been to highlight the potential for neurologists to be led astray by the common MRI appearances of "diffuse small vessel disease." While this label was appropriate for the supratentorial imaging changes in our patient, reflecting his age, background vascular history, and risk factors, it was clearly not appropriate for the brain stem and cervical cord ischaemia. Such a radiological distinction is admittedly challenging for a clinician to make. In this case it was the atypical, and rapidly progressive, nature of the clinical picture that prompted us (GPK, AMT) to seek the expert neuroradiological assistance that finally led to his correct diagnosis.

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NEUROLOGICAL PICTURE...

Gadolinium-enhanced magnetic resonance imaging demonstration of a mycotic aneurysm

A 64 year old woman with one week of low grade fever and malaise was found unconscious at home. On arrival at hospital, she was obtunded but had intact brainstem reflexes. She was febrile and a heart murmur was auscultated. Head computed tomography revealed subarachnoid haemorrhage in the left sylvian fissure. Transoesophageal echocardiography demonstrated multiple mitral valve vegetations (fig 1, arrows) with bileaflet prolapse and severe mitral regurgitation (fig 1A). Moderate posterior mitral annular calcification was evident with an associated mobile vegetation (fig 1B, arrowhead). Multiple blood cultures grew *Staphylococcus aureus*. While magnetic resonance angiography (MRA) was unremarkable, gadolinium-enhanced T1 weighted axial magnetic resonance imaging (MRI) revealed an enhancing lesion in continuity with distal parietal branches of the left middle cerebral artery (fig 2A, arrow). Cerebral angiography corroborated the diagnosis of mycotic aneurysm (fig 2 B, arrow).

Subarachnoid haemorrhage is an infrequent yet devastating complication of bacterial endocarditis, typically resulting from rupture of a mycotic aneurysm. The distal location and small size characteristic of mycotic aneurysms may preclude detection with MRA.[1] Gadolinium-enhanced MRI may identify such lesions despite normal MRA. Enhancement may be indicative of the highly inflammatory nature of these lesions.

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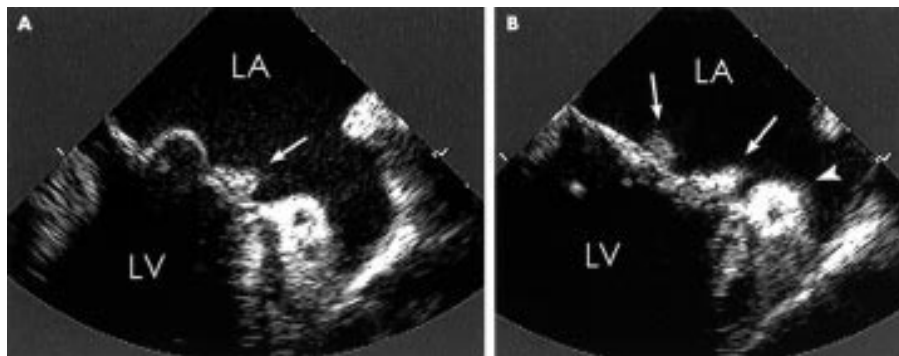
Figure 1 Transoesophageal echocardiogram demonstrating multiple mitral valve vegetations (arrows). Associated findings included mitral valve prolapse with severe mitral valve regurgitation (A) and moderate posterior mitral annular calcification with mobile vegetation (arrowhead, B).
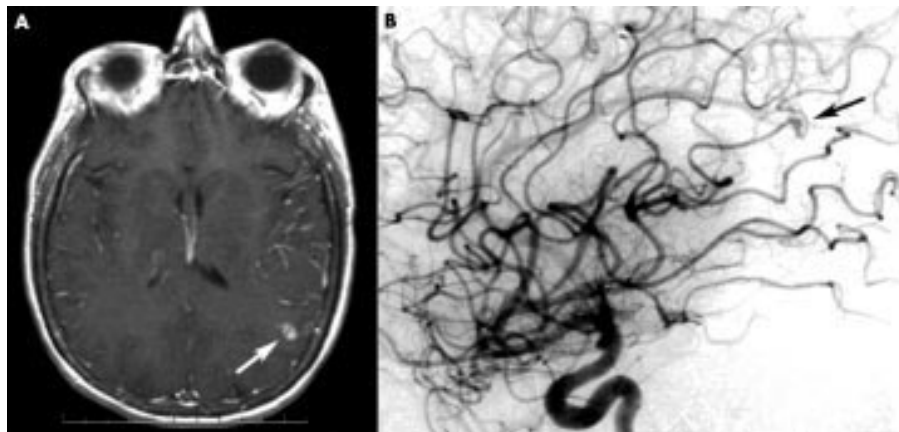

Figure 2 (A) Gadolinium-enhanced T1 weighted axial MRI and (B) left internal carotid angiogram showing a mycotic aneurysm in a distal branch of the left middle cerebral artery (arrows).