

SHORT REPORT

Intracranial dural fistula as a cause of diffuse MR enhancement of the cervical spinal cord

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Abstract

Spinal MR findings are reported in a patient with progressive myelopathy and intracranial dural arteriovenous fistula draining into spinal veins. Associated with previously reported abnormalities on T1 weighted and T2 weighted images, post-contrast T1 weighted images disclosed diffuse intense enhancement of the cervical cord itself. This enhancement decreased after endovascular treatment.

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Intracranial dural arteriovenous fistulas (DAVFs) are abnormal arteriovenous connections located within the dura mater. They account for 10%-15% of all intracranial arteriovenous malformations.¹ The clinical presentation, management strategy, and clinical outcome is highly variable and depends on the location of the DAVF and type of venous drainage.²⁻³

Intracranial DAVF with venous drainage into spinal veins is a rare event and may be associated with myelopathy.⁴⁻¹³ Knowledge of the MRI findings in this type of DAVF is important because many patients with myelopathy are first examined by MRI.

We report the MR findings in a patient in whom progressive cervical myelopathy developed as a consequence of intracranial DAVF. Of 23 cases of cervical myelopathy associated with intracranial DAVF and given MR information found in the literature,⁵⁻¹³ none had the diffuse intense enhancement of the cervical cord found in our patient. Moreover, postcontrast MR examination performed after endovascular treatment showed a dramatic decrease in the enhancement intensity.

Case report

A year before admission, a 36 year old man without relevant history noticed a progressive right lower and upper limb numbness. Initially this numbness happened after physical exercise. It had worsened in the past month and became responsible for falls. For 4 months he also complained of an abnormal tingling sensation involving his right cheek and lip, which recently spread up to his forehead.

On the day of admission, clinical examination was within normal limits. Magnetic resonance imaging disclosed a mildly enlarged medulla and cervical cord with abnormal hyperintensity on the long repetition time images (T2 weighted images) (figure A). Post-contrast sagittal T1 weighted images showed diffuse intense contrast enhancement of the medulla and cervical cord, extending to the C-6 level (figure B). Flow voids were present ventrally and dorsally to the cervical cord (figure A, B), suggesting enlarged vessels.

The day after MRI, the patient's condition deteriorated rapidly and he experienced a tetraparesis. A conventional angiography was performed in emergency, showing the intracranial DAVF located at the tentorium cerebelli with venous drainage into the spinal veins (figure C). The major feeding artery was the left occipital artery. In the hope of decreasing venous pressure, an occlusion of this artery was performed. A marked reduction in the blood supply of the fistula was achieved and the tetraparesis improved rapidly but partially.

A 2 week follow up MRI showed decreased extent of the high intensity signal on the long repetition time images and dramatically decreased intensity of cord enhancement on postcontrast T1 weighted images.

Discussion

Intracranial DAVFs are made up of a meshwork of arteriovenous shunts located within the intracranial dura. Their angioarchitecture is that of an "arteriovenous fistula" with multiple arteries converging into a single venous structure.¹⁴ The venous drainage of intracranial DAVFs seems a dominant point for explaining most signs and symptoms. That is why some authors have classified DAVFs according to their venous drainage.^{2, 14, 15} Among the different patterns of venous drainage, the Vth type corresponds to a drainage into spinal perimedullary veins.² This type may rarely be associated with ascending myelopathy.⁴⁻¹³

Recognition of the MR findings of intracranial DAVFs is important because most patients with symptoms of myelopathy are usually first examined by MRI. An improper diagnosis might result in delayed or incorrect treatment.

Of the 23 reported cases of intracranial DAVF with spinal venous drainage and given MR features,⁵⁻¹³ findings were:

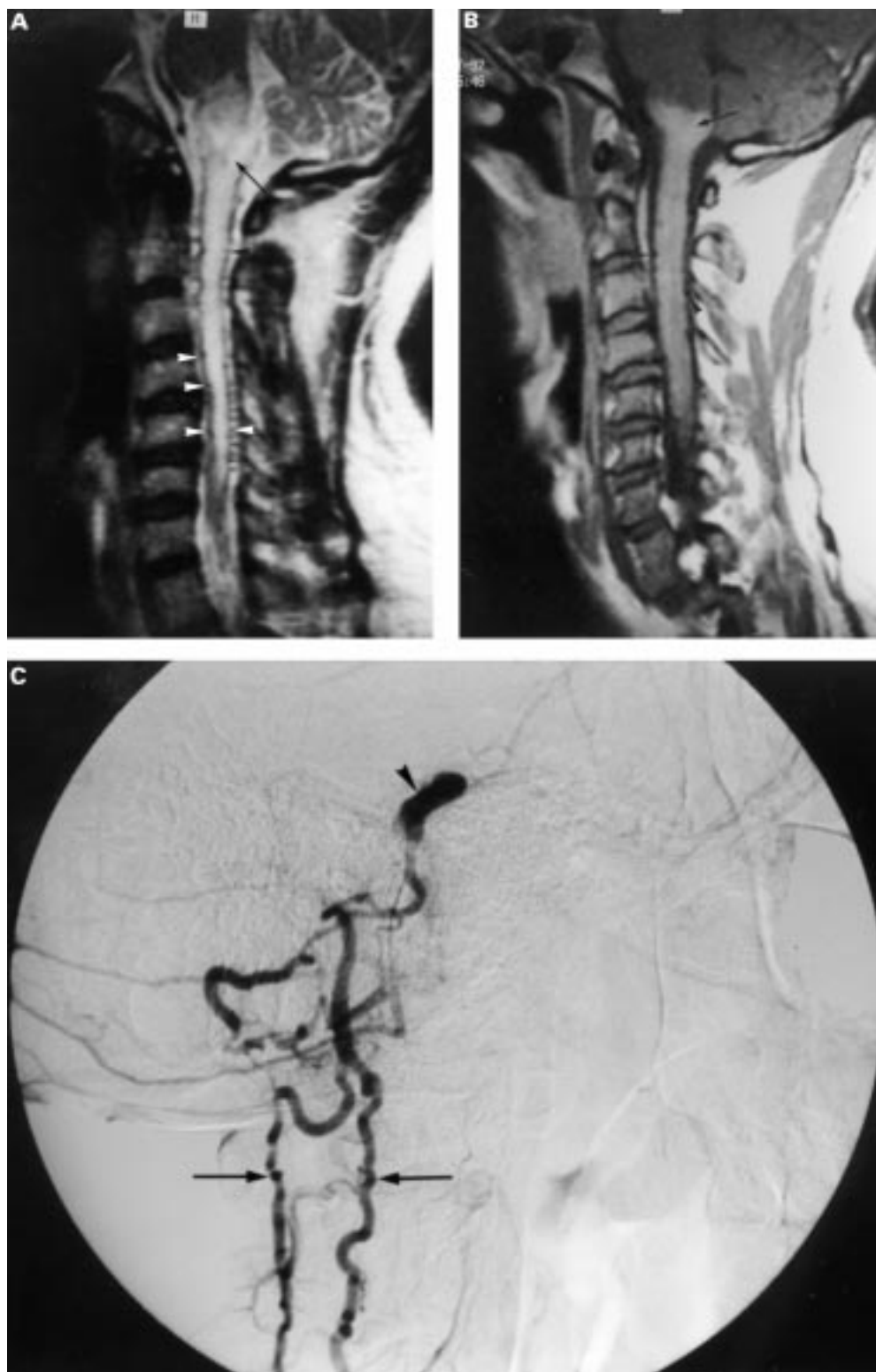
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(A) Sagittal T2 weighted image (3500/91/3, repetition time/effective echo time/excitations) shows abnormal hyperintensity in the medulla and cervical cord (arrow). Abnormally enlarged veins are present ventrally and dorsally to the brainstem and cervical cord (arrowheads). (B) Postcontrast sagittal T1 weighted image (500/15/3) shows intense parenchymal contrast enhancement of the medulla and spinal cord extended to the C-6 level (arrow) with flow voids of perimedullary veins (arrowheads). (C) Lateral view of a selective left occipital arteriogram shows anterior and posterior perimedullary veins (arrows) and the intracranial DAVF (arrowhead).

- A swollen cervical cord in nine cases,^{5-8 11-13}
- A central hyperintense signal of the cervical spinal cord on proton density and T2 weighted images in eight cases,^{5 10 12} of the thoracic spinal cord in one case,⁷ and of the conus medullaris in one case,⁹
- Perimedullary flow voids on T1 weighted or T2 weighted images, corresponding to the enlarged perimedullary veins, in 12 cases.^{5 7-13}
- Postcontrast T1 weighted images were detailed in five cases.^{8 9 11-13} Versari *et al*⁸ saw an intensely enhancing vascular lesion in the left petroclival region that they interpreted as venous drainage lying on the ventral surface of the brainstem. Ernst *et al*¹¹ found a prominent enhancement of the cervical cord in one of his patients at the C-4 level on MRI at 4 year follow up after surgical treatment of the fistula. Chen *et al*¹³ found an enhancement of the serpiginous flow voids along the surface of the cord. None disclosed a diffuse and intense enhancement of the spinal cord such as that found in our patient. Therefore, to our knowledge, diffuse parenchymal enhancement associated with intracranial DAVF has never been reported. However, mild to marked parenchymal contrast enhancement of the spinal cord on postcontrast T1 weighted images has been described in spinal DAVFs.^{16 17}

The pathogenesis of myelopathy in patients with intracranial DAVF draining into the perimedullary veins remains an intriguing issue. The pathophysiological mechanism which has gained the most acceptance^{2 4 5 7 8 10} is the theory of venous hypertension first proposed by Aminoff *et al* in 1974,¹⁸ expanded further by Merland *et al*¹⁹ and supported by rare pathological studies.²⁰ Venous hypertension might be produced by any cranial or spinal DAVF that gains access to the venous system of the spinal cord. The result of anastomoses between the arterIALIZED draining veins of the fistula and the coronal plexus of veins that normally drains the spinal cord parenchyma is a relative hypertension. Venous hypertension may be transmitted to the intrinsic veins of the spinal cord, resulting in reduction of the arteriovenous pressure gradient within the cord. Stagnation of blood flow and chronic hypoxaemia resulting from decreased perfusion leads to progressive necrosis of the medulla. Pathological study showed mural thickening with hyalinisation of both pial and intramedullary vessels with damage to the underlying neuropil.²⁰ Angiographic studies in patients with spinal DAVFs showed stagnation of contrast medium in the artery of Adamkiewicz with delayed normal venous return.²⁰ It also showed a lack of normal draining radicular veins when the patient was symptomatic. In a series of 12 patients with type V cranial DAVFs,¹⁰ six patients had a myelopathy. These patients had slow flow spinal perimedullary venous drainage that extended to the thoracic or lumbar region. A common angiographic characteristic of the six patients without myelopathy was that the spinal perimedullary veins draining the DAVF could be followed

down only to the cervical cord. At the cervical level, the perimedullary veins drained into the epidural veins via a medullary radicular vein. This type of drainage was then thought to prevent the venous hypertension in the spinal cord.

We suggest that the strong parenchymal enhancement of the spinal cord in our patient constitutes a sign of venous congestion and may represent stagnation of contrast medium within the enlarged intramedullary veins. It could also be the result of a blood-cord barrier disruption due to cord ischaemia. Three facts support the hypothesis of accumulation of contrast medium. Firstly, the congestion was probably very intense as the patient experienced an acute tetraparesis on the day after the MRI. It has been proposed that the Foix-Alajouanine syndrome of acute neurological deterioration in patients with DAVFs might be due to exacerbation of venous hypertension.²¹ Secondly, angiographic findings were in agreement with the previous series¹⁰ with perimedullary veins that could be followed by angiography down to the T-4 level. Finally, the rapid improvement of the symptoms found after endovascular occlusion of the main feeding artery, with a dramatic decrease of cord enhancement on postcontrast MR images, argues in favour of the haemodynamic theory rather than a cord-blood barrier disruption due to cord ischemia.

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HISTORICAL NOTE

A note on the use of botulinum toxin

Alan Scott, at the Smith-Kettlewell Eye Research Institute, San Francisco, pioneered the therapeutic use of botulinum toxin in focal hypercontraction of skeletal muscles.¹ The toxin also inhibits the release of acetylcholine at motor nerve terminals and at cholinergic parasympathetic and sympathetic terminals producing autonomic symptoms. This autonomic effect has been used successfully in the treatment of hyperhidrosis² and in smooth muscle hypercontraction of achalasia.³ Christian Andreas Justinus Kerner (1786–1862) was a German physician and poet. He published the earliest account of foodborne botulism in 1817⁴ and later published two monographs.^{5,6} He followed the clinical course of his patients. More importantly, he extracted the toxin and showed its effect on various animals. Kerner correctly concluded that it paralysed both skeletal and parasympathetic function, proposing its use as a therapeutic agent in neurological disorders characterised by involuntary movements such as chorea.

He thought that the toxin present in sausages was a “fatty acid” responsible for the signs of botulism. In his second monograph⁶ he described 155 cases of botulism and in detail described their autonomic symptoms: “The tear fluid disappears, the gullet becomes a dead and motionless tube; in all mucous cavities of the human machine the secretions of the normal mucus stands still, from the largest, the stomach, to the tear duct and the excretory ducts of the lingual glands. No saliva is secreted. No drop of wetness is felt in the mouth...”

Chapter 8, entitled “About the fatty acid as a possible therapeutic drug”, suggests the use of the toxin not only in muscular hypercontractions (“in such doses that its action could be restricted to the sphere of the sympathetic

nervous system only”) but also in hyperhidrosis and hypersalivation.⁷ Its current use in detrusor dysfunction, dysphonia and speech disorders, as well as dystonias and dyssynergia of skeletal muscles, justifies Kerner’s conclusions. He had conceded that what he had said about the “fatty acid” as a therapeutic agent “belongs to the realm of hypothesis and may be confirmed or disproved by observations in the future”.

Kerner wrote a letter to the King pleading for funds to allow him to continue his research. Unfortunately, it seems to have been refused. He quickly abandoned his research and turned toward medical practice and romantic poetry. He was closely associated with Robert and Clara Schumann. He wrote the text of *Der tote Muller*, set by Henri Vieuxtemps as *Die Sterne Tale stehn*.

Ermengem was to discover the organism *Clostridium botulinus* in cases of food poisoning in ham in 1897.⁸

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