Spinal dural arteriovenous malformation presented with intracranial hypertension in a young patient

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SUMMARY

Spinal dural arteriovenous malformations (AVM), the most common type of spinal cord vascular malformation, can be a challenge to diagnosis and prompt treatment. The disorder is rare, and the presenting clinical symptoms and signs are non-specific and insidious at onset. Spinal dural AVMs preferentially affect middle-aged men, and patients most commonly present with gait abnormality or lower-extremity weakness and sensory disturbances. It may rarely present with symptoms of intracranial hypertension including headache and visual disorders.

In this report, we present the radiological findings of a 19-year-old male patient with spinal dural AVM presenting with raised intracranial pressure symptoms including papilloedema and headache, and discuss with recent literature.

BACKGROUND

Spinal dural arteriovenous malformations (AVMs) are uncommon, accounting for 5–10% of all spinal cord lesions. These lesions are thought to be acquired conditions and affect predominantly middle-aged men; at least 80% of patients in most series are male.¹ The average age at diagnosis is 58–63 years, and more than two-thirds of patients are in the sixth or seventh decade of life at the time of diagnosis.² It is very uncommon for a spinal dural AVM to be diagnosed in a patient younger than 30 years.¹

Patients with spinal dural AVM typically present with back pain, root dysfunction or myelopathy. There are also a few studies which reported that intracranial hypertension symptoms including papilloedema and headache were present in some patients.³ MRI is essential, and it should encompass the entire spine, as localising physical examination findings (such as sensory level) do not mostly correlate well with lesion level.⁴

In this report, we present the radiological findings of a 19-year-old male patient with spinal dural AVM presenting with raised intracranial pressure symptoms including papilloedema and headache, and discuss with recent literature.

CASE PRESENTATION

A 19-year-old male patient presented with a 3-month history of headache and a 1-week history of visual disorders. There was no history of cervical or lower back pain and no bladder or bowel disturbances. Physical examination showed normal power of upper and lower limbs with normal muscle tone. Cognitive functions, speech and upper limb reflexes were normal, along with intact sensations and cerebellar functions. He had been in a

good state of health in the past and his family history was negative for any hereditary vascular disorders or AVMs.

INVESTIGATIONS

Brain CT imaging studies were conducted to investigate any intracranial pathology responsible for his headache and visual disorders. After not having detected any pathological findings on CT, a brain and cervical MRI were ordered. MRI studies revealed normal findings in the brain, however, signal void-dilated vascular lesions which comply with a cervical spinal AVM in the dorsal intradural space in the upper-cervical region (figures 1 and 2). Although there was no intramedullary involvement, the spinal cord was mildly displaced and compressed by the AVM.

OUTCOME AND FOLLOW-UP

The diagnosis consisted of a spinal dural AVM with raised intracranial pressure. He was referred to the neurosurgeon for further treatment.

DISCUSSION

Spinal dural AVMs are congenital lesions occurring between the fourth and eighth weeks of embryonic development when vessels differentiate into arteries, veins and capillaries.⁵ These lesions behave as shunts with the arteries communicating directly with veins rather than through the capillary bed.⁶ There are three biologically distinct types of vascular malformations of the spinal cord and its coverings: dural AVMs, intradural vascular malformation and cavernous angiomas. Clinical manifestations are variable in different types. Haemorrhage, compression, venous hypertension and ischaemia of the spinal cord cause the symptoms. Intradural AVMs are more likely to present in an apoplectic manner with pain and acute neurological dysfunction than spinal dural AVMs.⁷

Although rare, spinal dural AVM and other spinal lesions, including tumours, must be considered in differential diagnosis in patients with progressive headache and visual disorders.⁸ The exact frequency of spinal dural AVM has not been determined. They account for between 3% and 11% of all spinal lesions across all ages, although rarer in young patients.⁸ In a review of 304 spinal dural AVMs, only 10% occurred in young patients less than 20 years of age.⁹ The most common site for a spinal dural AVM is the lower thoracic spinal cord, although they can occur throughout the spinal cord, including the cervical region.^{8 10}

Raised intracranial pressure in patients with spinal dural AVMs is usually secondary to

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Figure 1 (A,B) Midsagittal T2-weighted cervical MRI study demonstrating signal void-dilated vascular lesions in the dorsal intradural space in the upper-cervical region.

subarachnoid haemorrhage with consequent communicating hydrocephalus and ventriculomegaly. The high flow rates through the AVM may also provoke sagittal sinus thrombosis and then produce pseudotumour by interfering with cerebrospinal fluid absorption.¹¹ In our patient, none of these factors were present. Other causes of raised intracranial pressure such as cerebral oedema, mass effect or ventricular obstruction were also absent. The occurrence of raised intracranial pressure in patients with a spinal dural AVM is rarely mentioned in the literature. In a review of 545 patients with arteriovenous malformations not one patient is mentioned with papilloedema.¹² Of 110 patients reported by Paterson *et al*,¹³ 17 had papilloedema, 14 secondary to subarachnoid haemorrhage, 1 from non-communicating hydrocephalus and the other cause was not specified.

MRI is essential in diagnosis of spinal dural AVMs, and it should encompass the entire spine. The prominent perimedullary signal void vessels on T2-weighted images are strongly suggestive of a spinal dural AVM. However, these findings do not help locate the site of the shunt. Three-dimensional phase-contrast MR angiography may be used to demonstrate the location of a spinal dural AVM.¹⁴ Location of dural AVMs with

dynamic MR or MR angiography will greatly facilitate spinal angiography. The emphasis during angiography then can be directed to defining the angioarchitecture of the malformation and to endovascular treatment.¹⁵ With further improvements in either dynamic gadolinium-enhanced MR or MR angiography, follow-up angiography is not necessary.

The optimal management of spinal AVM in both young patients and adults is unclear. Embolisation is generally recognised as the first stage in treatment, the aim is to stabilise the size of the arteriovenous shunt and thus prevent further deterioration.¹⁶ Although different types of spinal AVM need to be managed differently, the goal in treating spinal dural vascular malformation is to permanently obliterate the nidus of the malformation without damaging the cord tissue or compromising spinal cord blood supply.

In conclusion, prompt diagnosis and early management are essential in spinal dural AVMs to prevent long-term sequelae. Therefore, a high index of suspicion is needed to prevent delays in treatment. New and/or progressive neurological signs, including headache and visual disorders, can be one of various presentations in spinal dural AVMs. MRI is essential, and it should encompass the entire spine.

Figure 2 (A,B) Axial T2-weighted cervical MRI study demonstrating signal void-dilated vascular lesions in the dorsal intradural space in the upper-cervical region.

Learning points

- ▶ Spinal dural arteriovenous malformations (AVMs) are congenital lesions occurring between the fourth and eighth weeks of embryonic development when vessels differentiate into arteries, veins and capillaries.
- ▶ Although rare, spinal dural AVM and other spinal lesions including tumours must be considered in differential diagnosis in patients with progressive headache and visual disorders.
- ▶ MRI is essential in diagnosis of spinal dural AVMs, and it should encompass the entire spine.

Competing interests None.

Patient consent Obtained.

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