

Spinal Dural Arteriovenous Malformation : A Cause of Myelopathy

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Summary

Spinal vascular malformation is an uncommon but potentially treatable cause of myelopathy. We describe two cases of angiographically proven spinal vascular malformation in Malaysia. The first case is a 47-year-old man who had a progressive myelopathy and the second a 60-year-old man with intermittent attacks of transient paraparesis leading to paraplegia. As the clinical presentation of spinal vascular malformation is variable, it should be considered as a cause of patients with myelopathy.

Key Words: Spinal Dural Arteriovenous Fistula, Myelopathy

Introduction

Spinal vascular malformation causing myelopathy is often underestimated. According to McCormick's classification of spinal vascular malformation, spinal dural arteriovenous fistulas (SDAF) account for 75% of these abnormalities¹. We report two cases of SDAF documented on spinal angiogram.

Case Report

Case 1

A 47-year-old Chinese man presented with progressive spastic paraparesis associated with sphincteric loss of his bladder over three years. A year after the onset of illness, he was confined to his home. The impairment was associated with frequent episodes of painful flexor spasms.

Neurological examination showed spastic paraparesis of grade 4 power associated with hyperreflexia and clonus in both lower limbs. There was loss of sensation to all modalities from T8 dermatomes downwards. Examination of the upper limbs, cranial nerves and other systems was normal. Baseline blood

investigations, plain X-rays of the thoraco-lumbar spine were normal. Cerebrospinal fluid microscopy and IgG index were normal.

The supine myelogram showed a tortuous dilated serpiginous filling defect at the thoraco-lumbar junction suggestive of a spinal vascular malformation. Spinal angiogram showed a single feeding vessel to the SDAF from the right T9 intercostal artery (Fig. 1).

Embolisation of the SDAF was done in Singapore. Although the patient continued to have spastic paraparesis at 9 months of follow-up, there was no further progression of the myelopathy following embolisation.

Case 2

A 60-year-old Sikh man with longstanding hypertension and diabetes mellitus presented with several episodes of transient paraparesis lasting from one to twelve hours duration over a two-month period. This was associated with low back pain and urinary retention.

Examination during one such episode showed

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paraparesis with sensory loss at T9 downwards. There was hyperreflexia in the lower limbs and bilateral extensor plantar responses.

Routine blood investigations, cerebrospinal fluid studies and gadolinium enhanced MRI of the thoraco-lumbar spine were normal. Supine myelogram showed serpiginous filling defects at the thoraco-lumbar region. Spinal angiogram showed a dural fistula at T8 intercostal artery consistent with a SDAF (Fig. 2).

This patient declined any further treatment and eventually developed paraplegia two months later.

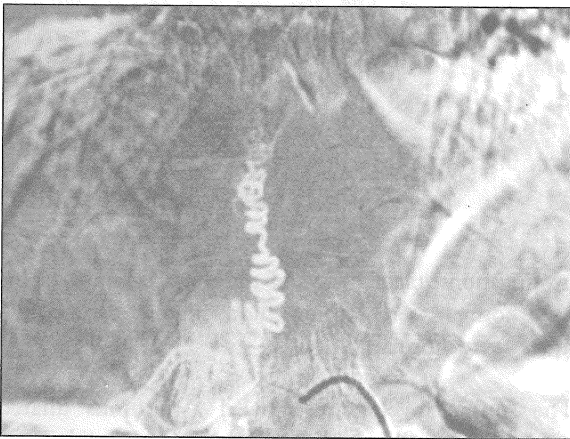


Fig. 1: Spinal angiogram showing selective catheterisation of T9 intercostal artery supplying the SDAF

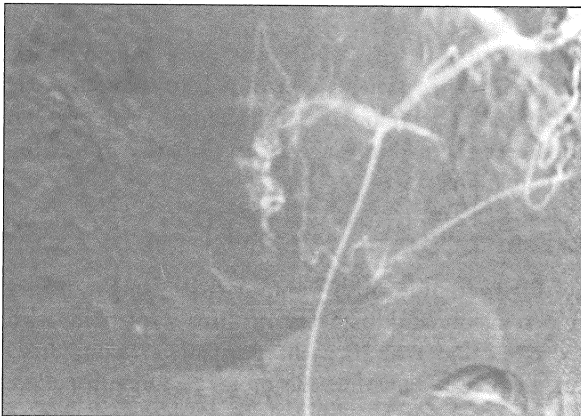


Fig. 2: Spinal angiogram showing a dural fistula at T8 intercostal artery supplying a SDAF

Discussion

Spinal dural arteriovenous fistulas (SDAF) is a relatively new pathological entity first described by Kendall and Logue in 1977². It has been recognised as the most common type of spinal vascular malformation in adults, occurring most commonly in the thoraco-lumbar region. It is more common in men than women, with a male to female ratio of 8:1. The males affected are usually middle-aged or elderly¹.

We describe two cases of angiographically proven spinal dural arteriovenous fistula at the thoraco-lumbar region. Both patients presented with symptoms of myelopathy with differing temporal sequence. The former case had a progressive spastic paraparesis and the latter recurrent episodes of transient paraparesis.

The variability of the presentation of SDAF is well recognised: this may be acute, subacute, episodic or progressive myelopathy¹. The two cases described were consistent with the known demographic and clinical manifestation of SDAF.

The pathological features of SDAF are due to an acquired dural fistula communicating between a radicular artery and vein located on the dural nerve sleeve. The varied clinical presentation is believed to be due to a combination of chronic venous hypertension and associated dural sinus thrombosis¹. However, a more recent paper on a proven case of SDAF by angiography and pathology supported increased venous pressure as a mechanism of neurologic dysfunction³.

Supine myelograms performed on both patients showed serpiginous filling defects in the thoraco-lumbar regions. However, in both patients the MRI with gadolinium enhancement failed to demonstrate these lesions. This is most likely related to the magnetic strength of the MRI machine which was 0.5 Tesla in both patients. SDAF was reported to be visible in only 25-63% of cases².

Both supine myelogram and MRI with gadolinium enhancement are usually performed to seek corroborative evidence prior to spinal angiography. Spinal angiography is a tedious procedure and has

significant morbidity with selective cannulation of the intercostal arteries, lumbar arteries and the vertebral arteries.

Interventional neuroradiology has an important role in the treatment of SDAF. The first case was treated with endovascular embolisation which halted the progression

of the paraparesis. Unfortunately, the second patient declined treatment.

In conclusion, spinal dural arteriovenous fistula is a potentially treatable cause of myelopathy. Spinal angiography remains the gold standard for confirming or refuting the presence of SDAF.

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Psychiatric Presentation of Huntington's Disease in a Malaysian Family

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Summary

A 32-year-old Chinese lady presented to the Psychiatric Clinic with a history of change in personality for 2 years and abnormal movements for a year. After thorough investigations and observation a diagnosis of Huntington's Disease was made. Her elder brother was traced and found to have Huntington's Disease as well. He had a long standing history of antisocial behaviour and substance abuse long before the onset of the choreiform movements. Her younger brother also has choreiform movements for the last 2 years and had recent change in personality. Their mother also had abnormal movements and was recorded to be depressed and attempted suicide. The maternal grandfather had a mental illness and was warded at a mental institution till his death in 1942. Psychiatric presentation of Huntington's Disease in this Malaysian family is prominent and preceded the characteristic movements in the present generation.

Key Words: Huntington's Disease, Psychiatric symptomatology