

## Spinal Cord Arteriovenous Malformation: Presenting As Relapsing Paraplegia

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### ABSTRACT

Spinal arteriovenous malformations are rare cause of spinal cord syndromes. It is important in identifying them early as it is a potentially treatable cause. The clinical and radiological features may mimic demyelination. But steroids worsen the symptoms as opposed to demyelinating illnesses. We describe the case of a middle aged man presenting with the relapsing paraplegia.

**Key Words:** AV Malformation, Spinal Cord, Relapsing Paraplegia, FIESTA Sequence, CT Angiogram.

### INTRODUCTION

A 44 year old male belonging to a tribal family, who is a manual labourer by profession, presented with complaints of insidious onset weakness of left lower limb with thinning of the limb and ascending sensory symptoms. 5 months later he noticed weakness and numbness of the right lower limb as well. For the last 3 months he has incomplete bladder evacuation and is constipated.

He was treated from elsewhere as demyelination, but symptoms worsened with steroids. 10 years back, he had an abrupt onset of weakness of both the lower limbs that improved in one month time. His treatment records showed that it was also treated as demyelination. Physical examination showed wasting of the left lower limb with a power of grade 3 MRCC, hypotonia and hyporeflexia. His right lower limb was spastic with hyperreflexia and a power of MRCC grade 4. These features pointed to relapsing paraparesis in a middle aged man.

However the clinical picture did not correlate with a demyelinating illness as there was extensive lower motor neuron sign in one lower limb and upper motor neuron sign in the other limb. This along with the history of worsening with steroids raised the suspicion of dural arterio venous fistula. MRI with FIESTA sequence was taken that showed spinal cord as well as dural arteriovenous malformations. The feeding vessels were delineated with a CT angiogram.

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### DISCUSSION

Vascular malformations of the dura and the spinal cord are treatable causes of progressive myelopathy. They are rare and underdiagnosed entities, which, if not treated properly, can lead to considerable morbidity with progressive cord symptoms.<sup>1</sup>

There are 3 types of spinal vascular malformations- Extra dural, Dural and Intra dural.<sup>2-6</sup> The intradural malformations can be separated into perimedullary fistulas and real intramedullary arteriovenous malformations (AVM). Intradural AVMs are supplied by spinal cord supplying arteries, whereas dural malformations are supplied by meningeal arteries as branches of the radicular artery.<sup>3,6</sup> 80 percent of all spinal AVMs are dural fistulas. In contrast to the probably inborn perimedullary fistulas, dural AV fistulas are most likely acquired.<sup>5,6</sup> The fistula itself is located in the dural layer near the penetration point of the nerve root. Venous drainage runs along the spinal cord veins on the surface of the cord, because the local radicular venous drainage is missing. The high venous pressure is presumed to be the cause of clinical symptoms.<sup>2,3,5,7</sup> Patients usually present with slowly progressive myelopathy and/or radiculopathy, with attacks of sudden or fluctuating deterioration upto the transverse lesion.<sup>4,6,8,9</sup> Most dural AV fistulas are located below the mid thoracic level, usually consisting of a connection between a radicular feeding artery and the dural veins.<sup>10</sup>

The typical presentation is that of a middle aged man presenting with progressive myelopathy that worsens slowly or intermittently. It may have periods of remissions resembling multiple sclerosis. Acute deterioration is due to hemorrhage into the spinal cord. Other symptoms suggestive of dural AV malformation are intermittent claudication and symptoms that change with posture.<sup>10</sup>

Classical syndrome of intra medullary AV malformation is a progressive thoracic myelopathy with paraparesis developing over weeks to months. The pathological feature is the presence of thick hyalinized vessel within the cord (Foix- Alajouanine syndrome).<sup>10</sup>

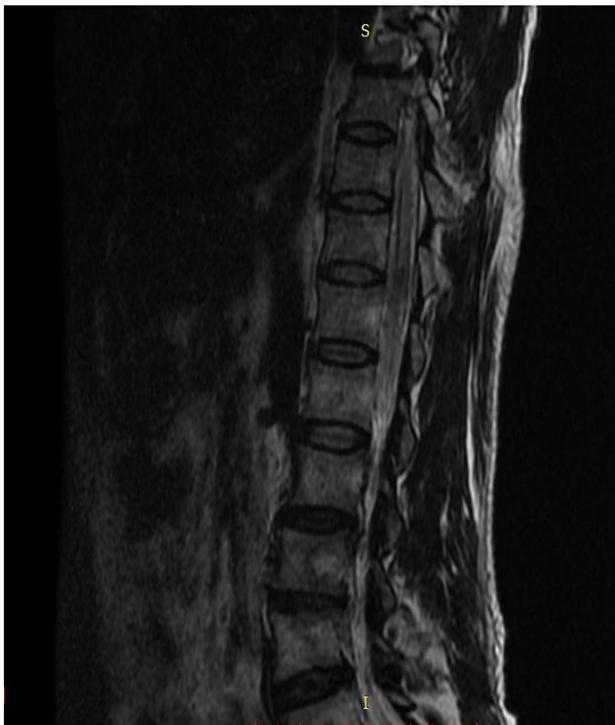
**SPINAL VASCULAR MALFORMATIONS**

1. Dural AVF
2. Perimedullary/cord AVF
3. Cavernous angioma
4. AVM

**DIFFERENCE BETWEEN SPINAL AVM AND AVF**

Spinal AVM	Spinal AVF
20%	80%
Young age	Old age
Congenital	Acquired
Nidus	No Nidus

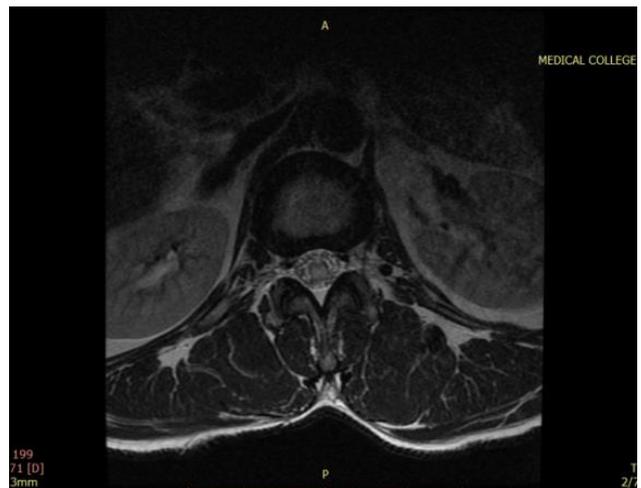
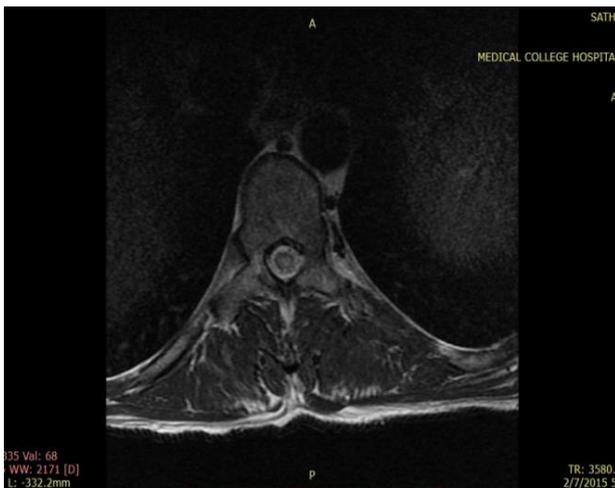
Spinal bruit is infrequent and should be sought after making the patient exert. High resolution MRI may detect some AVMs. Definitive diagnosis requires a spinal angiography. The treatment consists of intra dural interruption of the draining vein with coagulation or excision of the dural fistula. Alternatively, endovascular treatment with liquid embolic material may be attempted.<sup>11-15</sup>



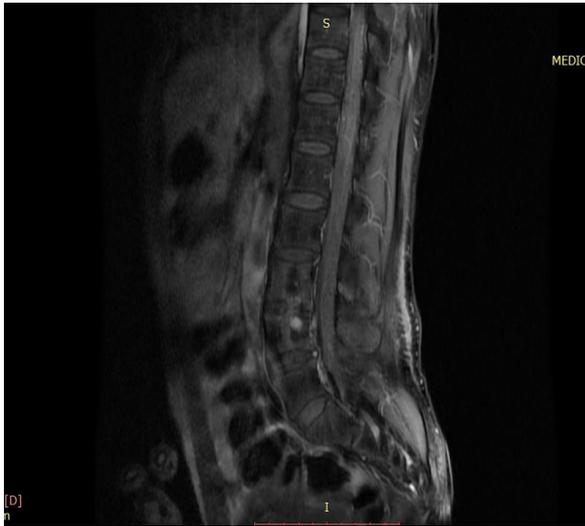
**Fig 1: Sagittal T2 weighted image showing- T2 hyperintense signal involving lower dorsal cord upto conus. Foci of T2 hypointensity also noted.**



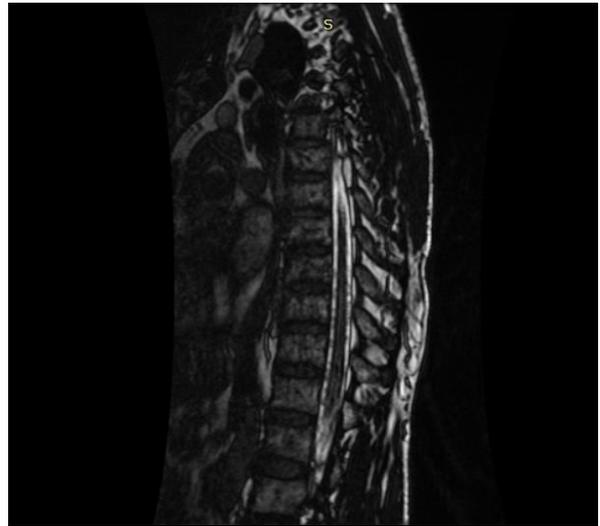
**Fig 2: Sagittal MERGE sequence- showing foci of blooming within the cord – suggestive of haemorrhage**



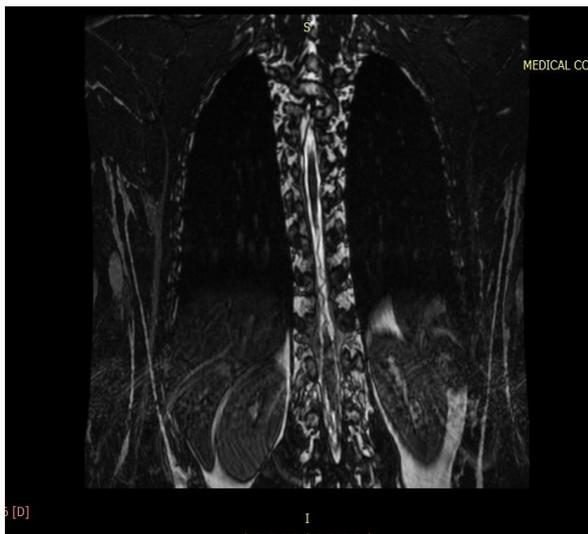
**Fig 3 and 4: Axial sections showing T2 hyperintense signal involving cord**



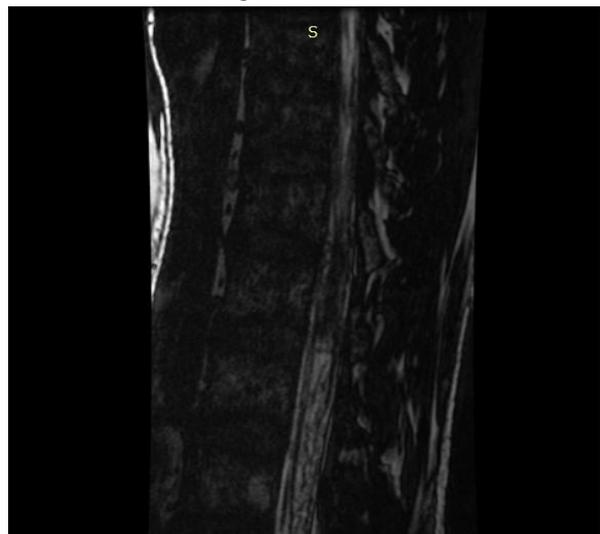
**Fig 5: SAGITTAL T2 POST CONTRAST: heterogeneous post contrast enhancement involving dorsolumbar cord.**



**Fig 6: FIESTA SEQUENCE - multiple serpiginous flow voids involving dura of dorsolumbar cord.**



**Fig 7: CORONAL FIESTA SEQUENCES- serpiginous flow voids involving dural space**



**Fig 8: FIESTA sequence – showing multiple flow voids involving dural space which are serpiginous**



**Fig 9: Sagittal section of contrast enhanced CT showing multiple tiny linear contrast enhanced vessels involving the dorsolumbar spinal cord. There is also a tangle of vessels noted within the cord at T11-T12 level. Arterial feeder appears to arise from the lumbar aorta and the draining vein being the hemiazygos vein.**



**Fig 10: Axial contrast CT section showing abnormal intramedullary contrast enhancement within the thoracic spinal cord**



**Fig 11: Coronal section of the contrast enhanced CT showing tiny serpiginous contrast enhancing vessels within the dorsolumbar spinal cord with a tangle of vessels appearing to form a nidus at T11-T12 level.**

## CONCLUSION

Spinal AV malformation should be considered in the differential diagnosis of a relapsing neurological symptom mimicking demyelinating illness. It may have a radiological resemblance to demyelination, but clinical neurological correlation and careful search for radiological evidence should be sought.

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