Perimedullary Spinal Arteriovenous Malformation in an Elderly Female: A Case Report

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Abstract

Perimedullary spinal AVM is a rare type of spinal arteriovenous malformations. We present a case of 70 yrs female who presented with motor weakness in her bilateral limbs. Initial MRI was misinterpreted as ependymal myxoma; however, histopathology revealed spinal AVM. MRA or DSA should be conducted if AVM is suspected.

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Abstract

Perimedullary spinal AVM is a rare type of spinal arteriovenous malformations. We present a case of 70 yrs female who presented with motor weakness in her bilateral limbs. Initial MRI was misinterpreted as

ependymal myxoma; however, histopathology revealed spinal AVM. MRA or DSA should be conducted if AVM is suspected.

Key Clinical Message

This case report highlights how rare spinal vascular diseases (spinal AVM) may be misinterpreted in conventional MRI. So we should be vigilant about spinal vascular diseases and conduct Digital subtraction angiography (DSA) when appropriate.

Keywords : Spinal Arteriovenous malformation, Spinal AVM, Perimedullary Spinal AV Fistula

Introduction :

Spinal arteriovenous malformations, along with cavernous malformations and spinal cord infarction, fall into a group of spinal vascular diseases. They are abnormal connections between arteries and veins of the spine which bypass the capillary network. Spinal arteriovenous fistula can be classified into Dural AVF, intramedullary glomus AVM, intramedullary juvenile AVM, Perimedullary AVF, and Extradural AVF according to a classification proposed by Takai et al.(1). We present a case of perimedullary AV Fistula, which was misinterpreted as myxopapillary ependymoma in MRI. However, it was correctly identified by histopathology and removed surgically.

Case Presentation:

A 70-year-old right-hand dominant Asian female presented with progressive bilateral lower extremity weakness, tingling sensation, and numbness over ten months of duration. At the time of the presentation, she was unable to walk on her own and was wheelchair-bound. She further complained of urinary incontinence and constipation. There was no associated fever, dizziness, blurring of vision, auditory abnormalities, speech abnormalities, weakness in the upper limbs, and history of trauma. She has a history of hemorrhoids and mild aortic regurgitation. She is currently not under any medication and doesn't have an account of surgical intervention in the past. She doesn't have a significant history of known allergies or similar illnesses in her family members.

On physical examination, her vitals were within the normal limits. A neurological examination revealed decreased muscle strength on her bilateral lower limbs with a power of 3/5 bilaterally. Deep tendon reflexes were hypoactive in her lower limbs and normal in the upper limbs. There was decreased touch and pain sensation in the bilateral lower limbs. Her PCR for SARS CoV 2 was negative, and her blood reports were normal.

An MRI of the thoracolumbar spine (plain and contrast-enhanced) was done. The MRI revealed an ill-defined heterogeneously enhancing lesion in conus medullaris with dural enhancement likely to be myxopapillary ependymoma.

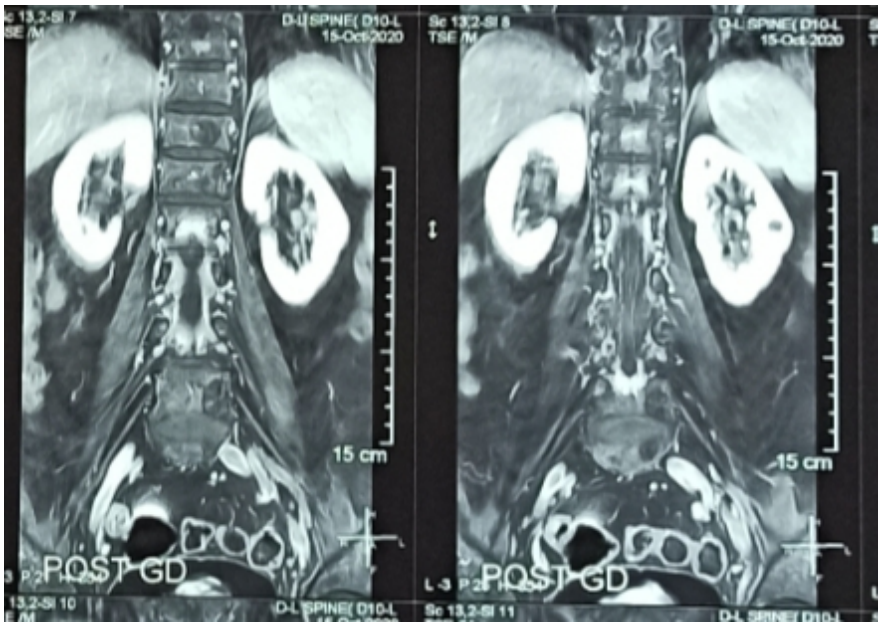

Figure 1 MRI with contrast T2 weighted showing intramedullary mass. (Coronal view)

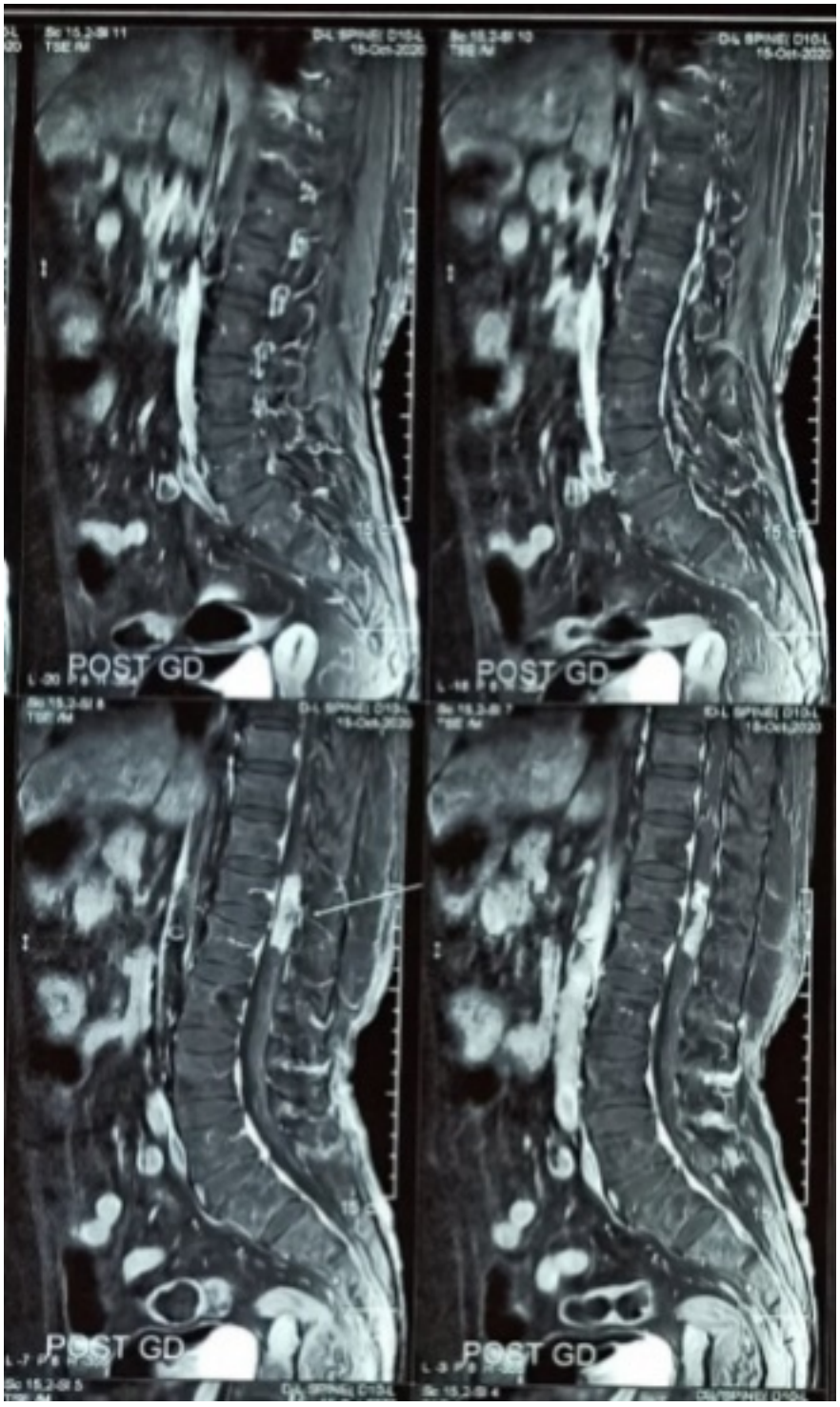

Figure 2 Sagittal view of T2 weighted Contrast MRI showing intramedullary mass at thoracolumbar level

Under general anesthesia, a laminectomy at the D11-L2 level and exploration was performed by an experienced neurosurgeon. The findings were a large tortuous vascular channel on the dorsal surface of the spinal

cord and a large tuft of vascular channels (nidus) on the right side of the dorsal root on the spinal cord at D12-L1 level. Excision of spinal AVM was done under GA, and AVM nidus was dissected off the pial surface of the spinal cord. First, the feeding artery was coagulated, followed by coagulation of the draining vein. Next, a feeding artery arising from neural foramen was coagulated and cut.

A tissue biopsy from nidus was taken and sent for histopathological examination, which further confirmed the diagnosis of spinal AV malformation. Postoperatively she was kept on vancomycin 500mg IV, fentanyl 50 microgram per hour IV, pregabalin 75 mg PO, amitriptyline, ondansetron 4 mg IV, labetalol, and thiamine. She was discharged after 12 days of surgery.

She was followed up 6 months later, on May 15, by phone call. There was mild improvement in the motor function of the patient after the operation. There were no complications or adverse effects.

Discussion

We presented a case of a Type IV spinal AV fistula. Spletzler et al.(2002) classified Type IV spinal AV fistula into three types according to size and flow of the fistula: the size, flow, venous tension increasing from Type A to Type C (2). In a study of patients with perimedullary fistulas, the mean duration from the appearance of symptoms to diagnosis was 24.6 months for chronic symptoms and 0.5 months for acute symptoms (3). Similarly, in our case, it was diagnosed within 11 months from the initiation of the symptom.

In a systematic pooled analysis of 213 patients with type IV spinal AV fistula, the annual rate of hemorrhage was found to be 2.5 %; however, for hemorrhagic lesions, the annual rate increased up to 5.6% (4). MRI should be used as the first diagnostic tool when Spinal AVM is suspected. Spinal AVMs are seen as flow voids on T2 weighted MRI and are seen as mixed hypo or hyperintense tubules in T1 weighted MRI depending on their flow velocity and direction (5). However, conventional MRI may have some difficulty in identifying spinal vascular malformation. In a retrospective study, sensitivity and specificity of conventional T2 MRI were found to be 82.8% and 89.4%, respectively, for spinal vascular lesions. Similarly, in another retrospective study, only 53% of cases were reported as spinal vascular malformation in conventional MRI (6). There have been case reports of perimedulary fistulas where conventional MRI has been unable to identify the lesion; as in the case report by Marshman et al. (7), MRI failed to identify flow voids. Also, in the case report by Suntharalingam et al. (8), perimedullary fistula was misinterpreted as a neurinoma. As in our case, it was initially misinterpreted as ependymal myxoma on MRI findings.

Embolization is a viable option for type IV b spinal AV fistula because of the diameter of feeding arteries. But studies have shown that embolization does not provide complete obliteration in all cases, because of which, they may require a surgical approach afterward (9,10).

Conclusion

This case demonstrates how MRI can misread rare spinal vascular disorders. So we should be vigilant about spinal vascular diseases and conduct DSA when appropriate.

Patient Perspective:

The patient had anxiety before surgery, and there was only a slight improvement in her symptoms after surgery, and she was satisfied with the treatment and care she received.

Consent: Written informed consent was obtained from the patient's daughter to publish the case report and the accompanying images.

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