Extradural Arterio-Venous Malformation Arising From Dorsal Nerve Mimicking A Tumor : A Case Report

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Abstract
A young male developed sudden weakness of both lower limbs with D8 sensory level. MR imaging revealed extradural lesion at D5-6. Laminectomy revealed brown colored highly vascular circumscribed lesion arising from D5 extradural nerve root. Lesion was excised completely and histopathology confirmed it to be arteriovenous malformation.

INTRODUCTION
Spinal extradural nerve root arterio-venous malformations are rare and those presenting as a mass are still rarer. There are no pathognomonic clinical features and diagnosis depends on high level of clinical suspicion. As there was no evidence of enlarged or abnormal vessels on neuroimaging and preoperatively on the involved nerve root or adjoining dura, possibility tumor was kept in mind but surgery revealed it to be a vascular mass and histopathology confirmed it to be arteriovenous malformation. Because of it mimicking a tumour, the present case is being reported.

CASE REPORT
A 25-year-old man was admitted on July 11, 2005 with sudden onset of painless weakness of both lower limbs of 2 days duration and bladder and bowel involvement of one day duration. There was no history suggestive of tuberculosis or diabetes. General physical examination was normal. On neurological examination tone was increased in the lower limbs and power was grade 1 in all muscle groups. Knee and ankle jerks were exaggerated superficial abdominal reflexes were absent. Anal tone was normal. All modalities of sensation were diminished by 90% up to D8 level.

On investigation hemoglobin and urinalysis were normal. X-ray thoracic spine were normal MRI of spine revealed an extradural lesion at D5-6 level on the left side. Lesion was iso-intense on T1 and hyper-intense on T2 with hypo-intense area in the lesion (Fig 1a, b, d).

Fig 1. Sagittal T2 weighted image showing hyperintense lesion with central hypointensities at D5-6 (a), isointense on T1 (b) irregular patchy contrast enhancement (c). Axial T2 image showing hyperintense lesion with central hypointensity on the left side (d) and irregular enhancement with gadolinium.

Lesion showed irregular contrast enhancement (Fig 1c, e). There was no evidence of dilated coronal venous plexus on the dorsal aspect of the spinal cord.

D 5-6 laminectomy revealed a brown colored highly vascular well circumscribed lesion in the left extradural location arising from D5 root. Root sheath was ballooned to form the capsule of the lesion. Lesion was excised completely along with part of the D5 root which was cut flush with dura. Patient improved postoperatively and is asymptomatic for the last 3 years.

Histopathologic examination was consistent with arteriovenous malformation. On H&E staining a collection of numerous dilated variable sized blood vessels were seen and Van-Gieson elastic stain highlighted the elastic lamina (Fig 2a, b).

Microphotograph showing collection of numerous dilated,
variable sized blood vessels (arrow) in close proximity (a) (H&E stain x100) and internal elastic lamina (arrow) in thick walled blood vessel (b) (Van-Gieson stain).

Figure 2

DISCUSSION

The spinal extradural arteriovenous malformations usually present with slowly progressive myelopathy and/or radiculopathy and rarely with spinal extradural hemorrhage resulting in acute cord compression rarely with recurrent intermittent paraplegia [[4-6]]. Most commonly the nidus is located on the dura in relation to the dorsal nerve root, drained by the intradural coronal venous system and most likely cause neurological deficits due to raised venous pressure within the spinal cord. Majority of spinal root arteriovenous malformations are intradural and may usually be associated with intramedullary angioma. Usually the feeding artery/arteries are visible along the root or dura and the enlarged veins may compress the root, however at times the feeding vessel may not be identifiable on imaging study. Symptoms in these patients are produced by venous congestion due to high venous pressure from arteriovenous shunt. At times the lesion may present as mass lesion with signal void and the lesion enhances with contrast. Nizuma et all reported a case of extradural arteriovenous malformation presenting as mass in relation to L4 root but did not mention whether it was arising from root or was separate from root.

There are reports of extradural radiculomeningeal AVMs in the cervical, lumbar and sacral regions however, their location in the dorsal region and sudden acute presentation probably due narrow spinal canal in dorsal region has not been reported. Operative and histopathological findings in our case suggested extradural arteriovenous malformation without connection to dural/intradural vessels, which is extremely rare.

Differential diagnosis of this lesion includes neurofibroma, meningioma, intramedullary tumor. Tuberculosis in the form of granuloma or abscess should be considered in the differential diagnosis in developing countries like India. In our patient though course was acute but there was no evidence of bleed, acuteness could be attributed to increased blood flow resulting in increased size of the mass with subsequent compression of the cord.

Various diagnostic techniques like MRI, CT myelography and dynamic CT have been used but selective spinal angiography confirms the diagnosis. Duration from onset of symptoms until diagnosis, the age at the time of treatment, the condition of deep tendon reflexes in the lower limbs as well as the severity of gait and urinary disturbance before the treatment were significantly correlated with functional outcome. However our patient made excellent recovery and is asymptomatic for the last three years.

CONCLUSION

We conclude that extradural arteriovenous may present atypically like tumor in the dorsal region.

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