Spinal Arteriovenous Malformation Associated with a Radicular Arteriovenous Fistula Suggested a Metameric Disease

A Case Report

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Summary

A spinal intramedullary arteriovenous malformation (AVM) associated with a radicular arteriovenous fistula (AVF) is reported. The patient had mild myelopathy and low back pain. Spinal angiography revealed the AVM fed by the anterior spinal artery via left T10, T11 and right L1 radiculomedullary arteries and the radiculopial arteries of left L1, L2 and right T11, L3 levels and the radicular AVF at the left L4 level. There were three radiculomedullary arteries within four levels in our case. This spinal AVM associated with a radicular AVF is considered a genetic nonhereditary lesion with metameric link.

Introduction

Multiple intradural spinal cord malformations are rare. A case of intramedullary spinal arteriovenous malformation (AVM) associated with radicular arteriovenous fistula (AVF) is reported.

Case Report

A 16-year-old girl presented with a 13 month history of stepwise progressing lumbago, motor weakness and numbness of the left lower limb. Examinations on admission disclosed mild mo-

tor weakness of the left lower limb and hypalgesia up to L2 level. Magnetic resonance (MR) imaging on T2 weighted image revealed a flow void on the conus medullaris. Spinal angiography revealed an intramedullary AVM fed by the anterior spinal artery, which was supplied by three radiculomedullary arteries on injection of the left T10 and T11 intercostal arteries and right L1 lumbar artery (figures 1,2), and the radiculopial arteries of the left L1, L2 and right L3 lumbar arteries, and right T11 intercostal artery (figures 1,2).

The intranidal aneurysmal dilatation was also seen. Additionally, spinal angiography on injection of the left L4 lumbar artery revealed the radicular AVF (figure 3). Endovascular embolization was attempted via the T-11 radiculomedullary artery using the flow-guided microcatheter. But it was impossible due to the severe tortuosity of the anterior spinal artery.

The approach via the right L1 radiculomedullary artery was also impossible due to its small caliber. After partial embolization, surgery was performed at least to obliterate an aneurysmal dilatation along one of the main feeders to prevent its rupture and, if possible, resect the AVM to delay or cease neurological deterioration. At surgery, the AVM was partially resected through the posterior median sulcus with posterior laminotomies.

Figure 1 Angiograms (anteroposterior view, A) left T10 intercostal injection, B) left T11 intercostal artery injection, C) left L1 lumbar artery injection, D) left L2 lumbar artery injection) demonstrating an arteriovenous malformation (nidus type) at the L4 myelomere in a 16-year old girl. The lesion is fed by radiculopial arteries arising from L1 and L2 and radiculomedullary arteries arising from T10 and T11, which contribute to the anterior spinal artery.

The feeder including its aneurysmal dilatation was successfully trapped.

Discussion

In the first few weeks of development from the rostral to the caudal end of the embryo, 31

somites are formed. Each of these receives one pair of arteries from the dorsal aorta, which are the segmental arteries. From the sixth week to the fourth month of intrauterine life, progressive longitudinal changes occur and result in a mature pattern. The two ventral neural arterial channels of the cord fuse on the midline to be-

Figure 2 Angiograms (anteroposterior view, A) right T11 intercostal injection, B) right L1 lumbar artery injection, C) right L3 lumbar artery injection) demonstrating an arteriovenous malformation (nidus type) at the L4 myelomere. The lesion is fed by radiculopial arteries arising from T11 and L3 and a radiculomedullary artery arising from L1, which contributes to the anterior spinal artery.

come a single ventral longitudinal arterial axis. Simultaneously, desegmentations occur; this corresponds to regression of most of the supply respective to the ventral axis and pial network. Of 62 metameric arteries (31 right and 31 left), 4-8 ventral radicular arteries and 10-20 dorsal radicular arteries remain, the others regress. Such as this, every segmental artery has a potential to be a radiculomedullary artery or a radiculopial artery¹. This is because three radiculomedullary arteries were located within only four levels in our case, which is rare.

Multiple spinal cord AVMs (SCAVMs) with metameric lesion are rare, with an incidence of 7 to 12% of the intradural SCAVMs in the literature^{2,3}. The term "metameric" is reserved for vascular malformations that can involve the spinal cord, bony structures, paraspinal musculature, subcutaneous tissues, and skin in the same segmental dermatome.

Rodesch⁴ proposed a reappraisal of the classification of spinal cord arteriovenous shunts (SCAVSs) that takes into account the supposed origin of each shunt and its relationship to a possible underlying disease. His classification primarily distinguished AVMs and AVFs and secondarily categorized these SCAVSs into three groups. The first group encompassed single shunts attributable to a genetic hereditary disorder in which the vascular cells are affected early by the disease, in the germinal stage.

The main syndrome in which genetic links can be recognized is a hereditary haemorrhagic telangiectasia, or Rendu-Osler-Weber disease. The second group involves SCAVSs that are always multiple, which share potential metameric links. These SCAVSs are not related to a hereditary disorder; they are genetic but non-hereditary. Two types of genetic non-hereditary lesions can be recognized. The first type is multiple shunts with a metameric link.

The association of SCAVSs and radicular AV shunts is metameric if the myelomere involved with the SCAVS corresponds to that of the involved nerve. The second type of genetic non-hereditary lesions is multiple shunts for which the metameric disposition cannot always be clearly demonstrated but can be postulated. The third group contains single lesions (either AVMs or microfistulae).

The lesions are intradural and affect one of the nervous structures found in this anatomic space, i.e., single focal cord, root, or filum

Figure 3 Angiogram (anteroposterior view, left L4 lumbar artery injection) demonstrating a radicular arteriovenous fistula.

shunts of either the AVM or AVF type. The association of SCAVM and radicular AVF is rare and potentially metameric if the myelomere involved with the SCAVM corresponds to that of the nerve involved. Our case is a genetic non-hereditary lesion with a metameric link under his classification.

The angiographic finding of radicular AVF is similar to one of the dural AVF. The dural AVF is the shunt between the dural branch and the radicular vein in the region of the interverte-

bral foramen. On the other hand, the radicular AVF is the shunt between the radicular artery and radicular vein.

The radicular lesions were localized on the spinal nerve intradurally and were supplied by radiculopial or radiculomedullary arteries and drained into a spinal cord vein. The shunting point of radicular AVF is located slightly far from one of the dural AVF. In the dural AVF, the supply from the contralateral radicular artery at the same level can contribute to the dural AVF.

Conclusions

A rare case of SCAVM associated with radicular AVF is described. There were three radiculomedullary arteries within four segments in our case, which is also rare.

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