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Case Report

Sacral meningeal arteriovenous fistula supplied by branches of the hypogastric artery revealed by conus medullaris infarction

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Objective: Spinal dural arteriovenous fistulas (DAVF), the most common vascular malformations of the spine, are usually supplied by branches of the intercostal or lumbar arteries. Rarely, the DAVF are fed by branches of the hypogastric artery. Only 12 such cases have been reported.

Case report: A 28 year-old man presented with a 2-month history of micturition dysfunction and progressive weakness of the legs. Physical examination showed motor deficit of the lower limbs with brisk knee jerks, absent ankle reflexes and normal plantar reflexes. Cremasteric reflexes were absent. We noted hypoesthesia of the lower limbs with complete anesthesia of the perineum. MRI of the lumbo-sacral spine demonstrated an enlargement of the conus medullaris with high T2 signal intramedullary lesion. It showed also large intradural serpentine vessels. A left iliac angiogram disclosed a nidus of arteriovenous malformation (AVM) supplied by a lateral sacral artery and draining by two enlarged ascending perimedullary veins. No clinical improvement was observed after surgical removal of the AVM.

Conclusion: The screening examination of choice for spinal DAVF remains MRI. When selective spinal arteriography is normal, we have to search for an unusual arterial supply particularly from the hypogastric artery.

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Keywords: spinal dural arteriovenous fistulas; spinal arteriography; hypogastric artery; conus medullaris infarction and medullary vein fistula

Introduction

Spinal dural arteriovenous fistulas (DAVF) are rare, however they are the most common vascular malformations of the spine and represent about 80% of the spinal arterio-venous malformations (AVM).[1–3] Spinal DAVF are usually solitary, small and located within the dura at the level of the intervertebral foramen.[4] Spinal DAVF increases pressure within the coronal venous plexus and may induce spinal cord dysfunction. These patients develop a slowly progressive myelopathy and rarely a radiculopathy and in severe cases can progress to complete paraplegia with sexual and sphincter dysfunction.[4,5] MRI and selective angiography are useful tools for the diagnosis.

The fistula is commonly located in the thoracic and lumbosacral regions and is fed by intercostal and lumbar arteries with venous drainage to dilated radicular and/or perimedullary veins (coronal venous plexus).[4,5] In some cases, the DAVF affects the cervical region and is supplied from branches of the thyrocervical, costocervical, external carotid, or vertebral arteries.[6–8] Rarely, the arterial supply of the fistula is provided by the middle sacral, the laterosacral or hypogastric arteries.[9–11] We report here a case of spinal DAVF supplied by a laterosacral artery.

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Case report

A 28 year-old man presented with a 2-month history of micturition dysfunction. A few days later, he developed acute urinary retention requiring bladder catheterization. One month later, he began to develop progressive weakness of the left lower limb which became bilateral.

Physical examination showed paraplegia with brisk knee jerks, abolished ankle reflexes and normal plantar responses. Cremasteric reflexes were absent. Muscle strength and tone were normal in the upper limbs. Sensory examination yielded hypoesthesia of the lower

limbs with L_1 sensory level, the perineum was completely anesthetic.

Sagittal unenhanced (Figure 1A) and enhanced (Figure 1B) T1 weighted MRI of lumbo-sacral spine demonstrated an enlargement of the conus medullaris with a mild diffuse enhancement. T2 sequence showed an increased intramedullary signal (Figure 1C). Axial T1 image at T_{11} level revealed a slight medullary enhancement. Sagittal T2 weighted image of the lumbar spine showed large intradural serpentine vessels (Figure 2).

Selective angiography of the left T_8 intercostal artery showed a normal Adamkiewicz artery. A left iliac anterior-posterior angiogram demonstrated the nidus of AVM supplied by a lateral sacral artery and draining by two enlarged ascending perimedullary veins (Figure 3A). Selective left sacral lateral angiography showed the DAVF (Figure 3B).

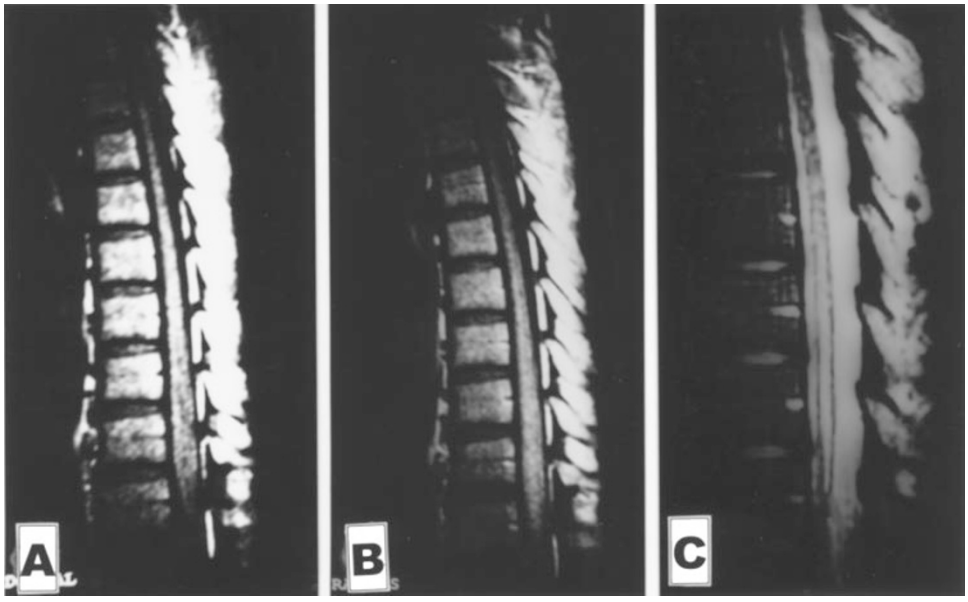

Figure 1 MRI of lumbo-sacral spine. Sagittal T1 weighed sequences showing an enlargement of the conus medullaris (**A**) with diffuse enhancement (**B**). Sagittal T2 weighted sequence demonstrated an increased intramedullary signal (**C**)

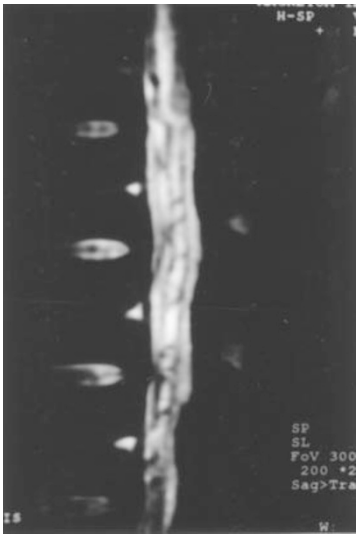

Figure 2 MRI of lumbo-sacral spine. Sagittal T2 weighted sequences showing large intradural serpentine vessels

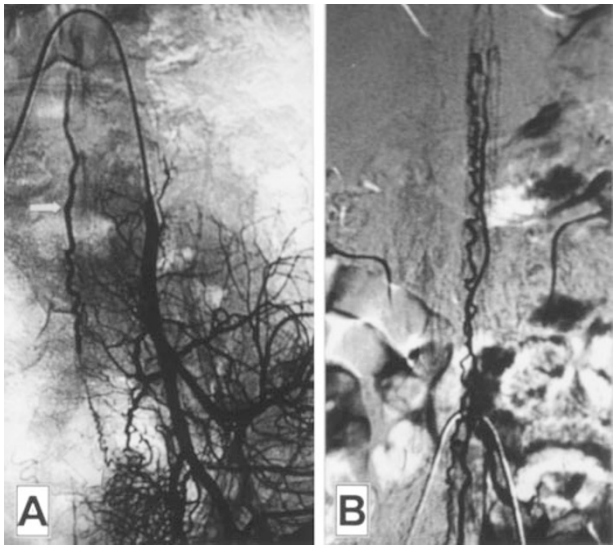

Figure 3 Left iliac angiography demonstrating the nidus of the arterio-venous malformation (thick arrow) supplied by a lateral sacral artery and draining by two enlarged ascending perimedullary veins (thin arrow) (**A**). Anterior-posterior selective left sacral lateral angiography showing the dural arteriovenous fistula (**B**)

No improvement was observed after surgical removal of the AVM, and the patient remained bedridden with bladder dysfunction.

Discussion

The clinical presentation of spinal DAVF supplied by branches of the hypogastric artery does not suggest this unusual arterial supply.[1,12–14] In fact, the fistula can produce symptoms very far from the nidus of the malformation.[4] To the best of our knowledge only 12 cases have been reported in the literature.[4] The clinical picture is non specific and can simulate transverse myelitis, multiple sclerosis, spinal cord tumors and degenerative disc disease. Only selective spinal arteriography can definitively identify site of the fistula. This aggressive examination is rarely performed, so DAVF seems to be underdiagnosed and its incidence is certainly higher than that reported in the literature.

MRI and myelography may show indirect signs suggestive of this malformation. They include spinal cord edema, myelomalacia, tortuosity and enlargement of the perimedullary venous system.[13,15–17] Unfortunately such techniques cannot delineate the shunt itself. Normal MRI and myelography did not exclude the diagnosis of DAVF.

Selective spinal arteriography should then be performed to show the Adamkiewicz artery, to locate the nidus of the malformation, its feeding vessels and the fistula. Slow flow demonstrated in the anterior spinal artery and enlarged perimedullary veins are suggestive of spinal DAVF.[6,18] This finding should prompt the investigation of all intercostal and lumbar radical arteries in addition to both thyrocervical, costocervical, vertebral and iliac arteries until the fistula site is located.[4]

Considering the possible risks of selective spinal angiography (thromboembolic phenomenon) and the accuracy of MRI, the former must best be reserved for atypical cases and for an eventual embolization.[19–21] Selective spinal angiography should also be considered in cases with multiple feeding radicular arteries.[3]

Surgery and embolization are the two modalities of treatment of spinal DAVF. Endovascular embolization must use permanent embolic agents to obtain permanent obliteration of the shunt nidus. Failure of embolization may be due to multiple arterial supply of the malformation or the use of non-permanent particulate agents with recanalization of the spinal DAVF.[14,22] Surgical resection of the site of arteriovenous shunting with interruption of the intradural draining vein is an effective treatment.[12,23]

Neurological improvement after successful obliteration of the shunt by either surgical or endovascular means occurs in more than 80% of cases.[12,24] An increased chance of neurological improvement is associated with mild pre-treatment deficit or shorter duration of symptoms regardless of the treatment modality.[12,25] Unfortunately in our case, subsequent to severe neurological deficit and diagnosis delay, disability remains after complete surgical removal of the fistula. Presence of intramedullary infarct at MRI, observed in our patient, is also indicative of poor prognosis.[26]

We conclude that spinal DAVF produces a non-specific subacute progressive myelopathy. Diagnosis suspected by MRI or myelography, must be rapidly confirmed by spinal angiography. Early aggressive evaluation and treatment should result in a greater chance of neurological recovery for patients with this most common of spinal vascular malformations.

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