Spinal arteriovenous malformations and neurogenic claudication

Report of two cases

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Spinal arteriovenous malformations (AVM's) can present with symptoms of neurogenic claudication indistinguishable from those of lumbar spondylosis. Spinal AVM's occur most frequently in males of middle age or older; lumbar spondylosis is often also present in these patients. The myelographic appearance of the abnormal vessels may resemble that of the dilated veins or redundant nerve roots sometimes seen adjacent to regions of spinal block, obscuring the diagnosis. Two patients are described who presented with clinical histories and myelographic findings that led to laminectomies for presumed spinal stenosis; ultimately, both were found to have an AVM. Treatment of the AVM arrested the neurological decline in one patient, and resulted in dramatic improvement in the other. A hypothesis related to hemodynamic consequences of venous hypertension is presented in an attempt to link the pathophysiology of the two conditions.

KEY WORDS • spinal arteriovenous malformation • spinal stenosis • neurogenic claudication • lumbar spondylosis

The similarity of the symptoms presented by spinal stenosis and peripheral vascular disease of the lower extremities, which can both cause claudication (literally, limping or gait-associated symptoms), is a well-known diagnostic issue. Arteriovenous malformations (AVM's) or arteriovenous fistulae represent an apparently quite distinct hemodynamic etiology of spinal claudication that is well recognized but rare. Given the prevalence of spinal spondylosis in the age group at which spinal AVM's usually become symptomatic, these lesions may occur occasionally in the setting of a narrowing spinal canal.

We have recently managed two patients with dorsal extramedullary (type I) spinal AVM's who had been treated previously for spinal stenosis. Following initial laminectomies, the condition of both patients deteriorated and, after undergoing additional decompressive laminectomies, they were sent to us for further evaluation. In one case surgical treatment of the vascular malformation stabilized but did not improve the patient's neurological status; in the other case marked improvement followed. These patients prompt consideration of the similarities between neurogenic claudication associated with spondylosis and that arising from spinal AVM's.

Case Reports

Case 1

This 59-year-old man presented with a 3-month history of pain in the right calf while walking, subjective weakness and numbness in both legs but worse on the right (also aggravated by walking), and urinary hesitancy. Myelography demonstrated a complete block at the L3-4 level, and computerized tomography (CT) showed a narrow canal at L-4 and L-5. Complete lumbar laminectomies at L-3, L-4, and L-5 were performed. Immediately after surgery his condition was neurologically unchanged, but on the 1st postoperative day he noticed increased numbness in the right foot. Examination revealed significant new weakness in the inverter and everter muscles of the right foot and mild new weakness in the right gastrocnemius muscle. Deterioration continued, and on repeat examination 45 minutes later there was no detectable voluntary contraction of the anterior tibialis or peroneal muscles. There was
also evidence of new weakness in the toe flexor muscles on the right and in the extensor hallucis longus muscle on the left. An emergency myelogram showed a persistent block at the L3-4 level, and he underwent emergency surgical reexploration. No compressive lesion was found. Widening the laminectomy and opening the dura over the cauda equina revealed no cause for the acute deterioration, although the possibility of arachnoiditis was raised. After the second operation, there was no improvement in right leg strength and the left leg was clearly weaker. He was transferred to our neurosurgery service for evaluation.

**Examination.** Admission examination showed profound right leg weakness (1/5 strength in the quadriceps, hamstrings, and toe flexor muscles; 0/5 in the ankle plantar and dorsiflexor muscles) and severe weakness in all groups on the left leg (2/5 to 3/5 throughout). Proprioception was absent in the toes, and there was a loss of touch and pinprick sensation, worse over the right leg than the left. Perineal sensation was intact. There was a sensory level at T-12 on the right leg and at T-11 on the left. The knee jerks were diminished, and ankle jerks and plantar responses were absent.

A repeat myelogram showed persistent narrowing of the thecal sac, particularly at L4–5, in spite of a very adequate bone decompression (Fig. 1 left). There were distended tortuous structures in the region of the conus, interpreted as most likely representing dilated veins from distal spinal stenosis, but also consistent with an AVM or with the redundant nerve roots sometimes seen in lumbar spondylosis (Fig. 1 right). An emergency spinal arteriogram was performed to study the segmental arteries from T-2 to L-2 bilaterally, as well as the thyrocervical, vertebral, and hypogastric arteries. Retrospective analysis of the films showed that the right T-12 artery was inadvertently omitted. No evidence of an AVM or fistula was seen in this arteriogram. A magnetic resonance (MR) image of the spine using surface coils did not show evidence of an AVM. By the next day the patient was essentially paraplegic and had lost any trace of bladder control.

**Operation.** In the face of a progressive conus syndrome of uncertain etiology, surgery was undertaken in spite of the negative angiogram. Laminectomies were performed from T-12 through L-2. Upon opening the dura over the conus, a tortuous complex of coiled arterialized veins was evident. There was no swelling of the cord suggestive of an intramedullary component. A large feeding vessel was found entering with the nerve root at the T12–L1 level on the right. This vessel was occluded with a temporary aneurysm clip, and initially there was no change in the arterialized coil of veins. However, after 4 or 5 minutes the veins became blue, and there was obvious evidence of stasis and early thrombosis throughout. The feeding vessel was then coagulated and divided, and the dura and wound were closed.

**Postoperative Course.** Immediately after surgery there was only slight improvement in the patient’s neurological status; however, over the next several weeks there was gradual return of sensation, strength, and bladder control. By the time of discharge from inpatient rehabilitation 2 months later, he could walk with a foot brace and control his urine. Six months postoperatively he had returned to full-time work as a surgeon.

**Case 2**

This 74-year-old man had developed severe gait-associated pain in both calves 5 years before admission; this deficit progressed and he ultimately needed the help of a cane for ambulation. Spinal CT and myelography showed a stenotic canal at the L-3 and L-4 levels, which were treated by laminectomy. Three years later, exacerbation of the symptoms led to further evaluation and to lumbar decompression by laminectomies at L-1 and L-2. Postoperatively, the pain worsened and he developed progressive bladder symptoms, requiring use of the Credé maneuver to void. Suspecting worsening scar tissue, another myelogram was performed which showed dilated abnormal veins.

**Examination.** The patient was referred to our institution for spinal angiography. Aortic atherosclerosis made the study difficult, but all intercostal and lumbar arteries from T-8 to L-3 were opacified. Slight filling of an early draining vein resulted from injection of contrast medium into the right L-2 artery, but additional supply to the fistula (perhaps from a hypogastric artery) was suspected because of the low volume of flow. For this reason, angiographic embolization (which had been considered) was not performed, and arrangements were made for surgical treatment of the AVM.

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**Fig. 1. Myelograms of the spinal arteriovenous malformation in Case 1. Thecal narrowing is evident (left) and distended vessels are visible over the conus (right).**

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Operation. At surgery, complete laminectomies were performed at T-11 and T-12, and the epidural scar tissue over the previous L1-4 laminectomies was carefully dissected. Upon opening the dura, a typical dorsal coiled arterialized vein was noted over the conus. The arterialized vessel was followed and the fistula was disclosed between the roots of the cauda equina at about the L-3 level. Occlusion of a feeder (probably an arterialized vein) with a clip resulted in slackening of the malformation, which became completely blue and soft after cauterization of an additional descending feeder found entering the spinal canal with a nerve root.

Postoperative Course. Postoperatively, the patient’s strength was unchanged from the preoperative status. He resumed walking on the 3rd postoperative day. At a follow-up visit 7 weeks after the operation, he still had no significant recovery from his preoperative deficit, and required a walker to ambulate. His condition has remained stable for 2 years.

Discussion

These two patients were operated on by competent neurosurgeons who assumed that their typical complaints were due to neurogenic claudication caused by spinal stenosis; this diagnosis was corroborated by the clinical examination results and confirmed by the radiographic findings. Unfortunately, a second pathological process was present, and treatment of the spinal stenosis (which was certainly present in both cases) was followed by worsening of both patients’ deficits. In Case 2, deterioration was slow and may simply have been a result of the natural typically progressive history of spinal AVM’s.2-4,12,16,17,22 In Case 1, however, deterioration after each of the laminectomies suggests that the operations themselves contributed in some way to the deterioration. These cases prompted us to review the historical and possible pathophysiological links between spinal AVM’s and spinal stenosis. The clinical similarity of spinal claudication in the two situations makes this differential diagnosis factor one of considerable practical importance.

The earliest clinical descriptions of spinal claudication recognized the potential for vascular malformations to produce this syndrome. Indeed, Wyburn-Mason25 in his 1943 review of 67 cases of vascular malformations, indicated that nine of the clinical histories mentioned spinal claudication as later defined by Verbiest.24 In some cases of spinal claudication where no malformation is present, there has also been consideration of possible underlying vascular components of the pathogenesis; this has been prompted in large part by the similarity of symptoms between these patients and patients with claudication from iliac or femoral arterial occlusive disease. Verbiest made the strong argument that bone compression (that is, spinal stenosis) rather than a vascular lesion is the cause of neurogenic claudication. His reasoning was an attack on the hypothesis that the syndrome could be caused by arterial insufficiency; other hemodynamic disturbances were not considered.

In the meantime, new interpretations of the hemodynamic abnormalities accompanying spinal AVM’s have emerged. The currently accepted view contends that the slowly progressive, apparently ischemic symptoms may arise largely as a result of venous hypertension.2,15-17,19-21,22 This interpretation is consistent with the good results obtained with these lesions after interruption of the feeders by surgery or embolization,5,7,9,16,18-20,22,23,26 and with the fact that symptoms of spinal AVM’s may be severe and progressive in spite of very low flow through the lesions in comparison with cerebral AVM’s. Venous hypertension is now thought to develop because the arterialized complex of vessels seen on the dorsal surface of the spinal cord is in fact the coronal venous plexus that normally drains most of the substance of the cord. This plexus becomes dilated and tortuous as a result of arterIALIZATION through one or more “feeders” which drain the actual AVM or fistula, which is usually extradural and associated with the dural sleeve of one of the spinal nerves.15,16,19,20,23

If venous hypertension can significantly contribute to problems caused by AVM’s and even cause neurological symptoms when the source of venous obstruction is outside the spinal canal, it certainly could complicate spinal stenosis as well. This would be expected in cases where venous hypertension results from an extradural fistula or AVM (as in the two cases reported here), and also in cases where venous hypertension results from some other cause. The same bone changes that compress the cauda and roots in cases of spinal stenosis (namely, anular bulging, medial hypertrophy of the facets with narrowing of the lateral recess, kinking of the root by hypertrophic pedicles, hypertrophy and buckling of the ligamentum flavum, and laminar hypertrophy with “shingling”) can potentially impede venous outflow by directly compressing the venous plexus or the veins that drain it and accompany nerve roots through the right foraminae. All of these mechanical effects become exacerbated in the lordic posture assumed during ambulation, which could lead to further restriction of venous outflow and an increase in venous pressure. Although the actual pressures in the intradural veins have not been measured, distension of the venous plexus adjacent to spondylotic blocks has been well documented both myelographically and with CT.13 It seems likely, therefore, that venous hypertension plays a role in lumbar spondylisis, although direct mechanical pressure on nerve roots is probably primary.

A further, related pathophysiological similarity between the two conditions is the presence of pathological mass effect. In uncomplicated spondylisis, this results from bone changes; in spinal AVM’s the distended vessels provide the compressing mass. Although distinct from venous hypertension as a cause of tissue injury, the two mechanisms are closely related in the cases under consideration. Because the vessels of the malformation have high capacitance, they dilate significantly

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with increased pressure. In general, direct compression of neural structures by the lesion is probably of secondary importance, since the coils of type I AVM’s are almost never large enough to cause a block.\(^5\)\(^,\)\(^10\) In patients with coexisting spondylosis, however, the spondylosis may exacerbate the venous hypertension (particularly during ambulation) by further impairing the already marginal venous outflow. The interaction between the potential pathophysiological mechanisms is diagrammed in Fig. 2. Distinguishing the relative contributions of these two potential sources remains difficult.

The venous hypertension hypothesis may also offer an explanation for the deterioration seen in the first case after each laminectomy, which was performed with the patient in the prone position. It is possible that an exacerbation of the venous hypertension secondary to abdominal compression while in the prone position was a significant factor. We have no other plausible explanation of how a laminectomy at L3-5 performed by a competent neurosurgeon could have caused or influenced a neurological syndrome, which, as it evolved after the second laminectomy, was clearly referable to the area of the conus medullaris.

These cases raise practical as well as theoretical issues. The most difficult aspect in each was recognition of the correct diagnosis. The problem is how to distinguish symptoms due to common spondylosis from those associated with the much rarer arteriovenous fistulae, given the similarities in clinical presentation and the age group at risk. There are no clear rules to avoid diagnostic confusion, as the conclusions of the neurosurgeons who first treated the present patients can attest. While the symptoms are similar (principally pain, usually radicular, on ambulation), the objective signs tend to be more impressive in patients with spinal AVM’s. Serious fixed or progressive neurological deficits are uncommon in patients with lumbar spondylosis but are the rule in untreated patients with AVM’s. Unfortu-

nately, reliance on more severe clinical deficits to distinguish the causes does not help with the early diagnosis of spinal AVM’s, which should ideally be detected before irreversible damage occurs. Myelographic views of the conus, although less frequently obtained now with the increasing use of water-soluble contrast agents, will supply the diagnosis in some cases, and should be insisted upon in patients with neurogenic claudication especially in cases with long-tract signs or suggestion of bladder involvement. Even with such studies, however, some cases will be missed. Deterioration of function, either immediately or later after decompressive surgery, should always suggest the possibility of an underlying AVM. Unfortunately, MR imaging has not invariably been helpful, as we learned in Case 1. A recent report has confirmed that some of these lesions, particularly the posterior extramedullary type of AVM, can be missed by MR imaging, although most of the larger lesions (particularly the intramedullary type) can be readily imaged by this technique.\(^8\)

Once the diagnosis has been made, treatment may be surgical or endovascular. In those cases where spinal angiography is nondiagnostic, surgical exploration for a suspected but unconfirmed lesion may be required, as in our Case 1.

References

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