A sacral dural arteriovenous fistula presenting with an intermittent myelopathy aggravated by menstruation

Case report

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The case is reported of a woman with a spinal dural arteriovenous fistula whose intermittent myelopathy became aggravated with menstruation. Her symptoms recurred in spite of successful acrylic embolization of the lateral sacral arteriovenous fistula. Repeat angiography showed venous drainage from the uterus toward the medullary vein. Total abdominal hysterectomy cured her symptoms. The pathophysiological basis for this peculiar clinical manifestation and its management are discussed.

KEY WORDS • spinal anomaly • arteriovenous fistula • venous drainage • hysterectomy • menstruation

Spinal dural arteriovenous (AV) fistulas have been recognized as an acquired AV shunt that presents as a progressive myelopathy secondary to venous hypertension of medullary veins. The anatomicopathological lesion consists of a feeding dural artery, an AV fistulous nidus, and an arterialized intradural draining vein. The symptoms include a slowly progressive myelopathy and/or radiculopathy with varying degrees of spastic or flaccid paraparesis, muscle atrophy, sphincter disturbances, and dorsal-column sensory signs. Neurological deterioration has been associated with high intraspinal venous pressure with a reduced AV pressure gradient leading to hypoxic damage of the spinal cord. This intraspinal venous pressure could also be influenced by various hemodynamic factors. Deterioration can be stabilized or reversed by therapeutic interventions such as embolization and/or surgical obliteration of the dural AV fistula.

We report a case in which the patient developed intermittent myelopathy during her menstrual cycle from a dural AV fistula. The case prompted consideration of the hemodynamic aspect of myelopathy associated with spinal dural AV fistula and its proper management.

Case Report

This 46-year-old woman presented with a few months' history of intermittent episodes of weakness and a cold sensation in both legs at the beginning of her menstrual periods. She usually regained function in about 2 hours. She occasionally experienced a burning sensation on the soles of her feet. Her neurological examination on admission revealed mild weakness (4/5) of the proximal muscles of the left lower extremity. The right patellar and both Achilles reflexes were absent, and superficial sensation below the knees was diminished bilaterally.

A thoracolumbar myelogram revealed scalloped filling defects in the region of the conus medullaris, consistent with dilated veins of spinal AV malformation. Computerized tomography (CT)-myelography demonstrated a slight expansion of the lower thoracic spinal cord surrounded by multiple tiny filling defects. Selective spinal angiography revealed a spinal dural AV fistula from the left lateral sacral artery draining toward the spinal canal via the filum terminale vein (Fig. 1 left). The artery feeding the lumbar enlargement originated from the left L-2 artery. Slow flow in the anterior spinal artery was noted.

Operation. Prior to embolization, the proximal segment of the left internal iliac artery posterior division was protected by placement of a Gianturco coil in the vessel. The fistula was successfully embolized with 0.15 ml of isobutyl 2-cyanoacrylate mixture using the "push" technique. A control angiogram of both internal iliac
arteries verified complete closure of the AV fistula (Fig. 1 right).

Postoperative Course. Immediately after embolization, there was significant improvement in the patient’s neurological status and she had no episode of weakness for one subsequent menstrual cycle. She then began to develop transient weakness in the lower extremities similar to those prior to embolization, although not as severe; these events were related to her menstrual cycles. Repeat spinal angiography performed 2 months after embolization confirmed exclusion of the fistula and normal circulation time through the artery feeding the lumbar enlargement. The muscular branches of both uterine arteries were still tortuous and hypertrophied, and an intense persistent capillary blush was noted in the corpus uterus (Fig. 2A). In the later phase, venous drainage into the same filum terminale vein through the sacral vein was seen (Fig. 2B).

A decision was made to recommend a hysterectomy so that the venous drainage into the sacral venous system would be reduced. One month later, the patient underwent a total abdominal hysterectomy and bilateral salpingo-oophorectomy. A large collection of aberrant vessels including large dilated varicose veins were noted on the left pelvic wall extraperitoneally. At pathological examination, the uterus and ovaries were unremarkable. Dramatic improvement of the patient’s neurological symptoms followed. At her recent 5-year follow-up examination, she was found to be symptom-free.

Discussion

This case raises several important issues for consideration. These include the basic pathophysiological mechanisms of the symptoms of spinal dural AV fistulas and their hemodynamic relationship to the spinal cord venous circulation.

Normally, venous drainage of the spinal cord begins from its intramedullary portion of the medullary veins, pierces the dura, and exits toward the epidural venous plexus. There is usually no retrograde reflux from the epidural vein to the intradural vein. It has been widely accepted that, in most patients harboring a spinal dural AV fistula, there is a dural branch (or branches) of the spinal ramus of the intercostal or lumbar segmental artery that supplies the actual fistula in the dura. Blood flow through the fistula runs toward the medullary veins and drains into the coronal and/or longitudinal venous plexus of the spinal cord, which forms the arterialized complex of vessels seen on the surface of the cord. This venous dilatation and stagnation may also be explained by the presence of a defect in the outflow to the epidural venous system. Therefore, the direct cause of hypoxic myelopathy is a decreased intramedullary AV pressure gradient due to raised pressure in the perimedullary venous system and/or by outflow obstruction. This phenomenon has been confirmed by Hassler, et al., who, in a direct measurement of intravascular pressure in the draining veins, found that it was about 70% of systemic arterial pressure.

Venous hypertension in the absence of normal epidural venous drainage can also be influenced by various hemodynamic factors. Exercise or certain postures which temporarily induce systemic arterial or venous hypertension exacerbate the symptoms in 42% to 70% of patients with a diagnosis of spinal dural AV fistula. Other reported aggravating factors are trauma, surgery, angiography, lumbar puncture, pregnancy, and systemic arterial hypertension. The precipitating factor that was unique in this case was the onset of menstruation. Myelopathy was consistently preceded by menstruation, even after successful fistula embolization.

It is well known that the vertebral veins, with their rich, valveless ramifications and connections, offer an important alternative route of venous drainage from the pelvis and lower extremities. Injection experiments with simulated abdominal straining showed venous flow from the pelvic veins was into the paravertebral venous system. Under certain circumstances, spinal medullary veins might be filled retrogradely from the epidural venous plexus in spite of a valve-like barrier at the dura. The retrograde filling of intradural veins, occasionally observed above an obstruction of epidural veins due to spinal stenosis, has been reported in a series of lumbar and cervical epidural venograms. As in the case of long-standing occlusion of the inferior vena cava, the diversion of significant venous drainage into the epidural venous plexus can produce symptomatic spinal venous hypertension.
Myelopathy aggravated by menstruation

FIG. 2. Follow-up angiograms obtained 2 months after embolization. A: The muscular branches of the left uterine artery (curved arrow) were tortuous and hypertrophied. An intense capillary blush was noted. B: Late venous phase showing persistent staining in the uterine musculature which drains only into the filum terminale vein through a sacral venous collateral vessel (curved arrows). The left hypogastric vein, which is the normal route of uterine venous drainage, is not opacified.

in which a posttraumatic pelvic AV fistula caused progressive paraplegia because of voluminous shunting into the epidural venous system after surgical obliteration of the usual pelvic venous pathways.20

One possible explanation in our patient is that the uterine hyperemia during the menstrual periods increased the venous return to an already compromised pelvic venous system, which in turn drained into the medullary vein through a sacral venous collateral route. This phenomenon is supported not only by a repeat angiogram which disclosed persistent uterine hyperemia with aberrant drainage into the filum terminale vein, but also by the surgically verified pathological condition in the pelvic veins.

The combined treatment of closure of the spinal dural fistula by embolization and surgical removal of the uterus completely cured the patient’s neurological dysfunction. Early recognition of the pathophysiology of symptoms based on the functional vascular anatomy and physiology played a significant role in the proper management of this lesion.

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