Spontaneous Thrombosis of a Spinal Conus Perimedullary Arteriovenous Fistula
—Case Report—

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Abstract

A 49-year-old male presented with a rare case of acute lower extremity paresis caused by spontaneous thrombosis of a spinal conus perimedullary arteriovenous fistula (AVF) after a subacute myelopathic course. Magnetic resonance imaging obtained after deterioration showed that the flow voids around the conus medullaris had changed from hypointense to hyperintense lesions. Surgery with thoracic laminoplasty was performed to determine the nature of the lesion because angiography was negative. During surgery, thrombosed abnormal vessels were observed, consistent with thrombosis of a spinal conus perimedullary AVF. Histological specimens of the thrombosed vessels exhibited vascular wall injury such as intimal alteration, wall dissection, and mural thrombus. Hemorrhage and infection were excluded. Vascular wall injury of draining veins and varices were probably one of the causes of thrombosis in the present case. Spinal arteriovenous malformation generally causes progressive venous congestive myelopathy, but the congestive myelopathy may rarely rapidly deteriorate with spontaneous thrombosis, known as Foix and Alajouanine syndrome.

Key words: Foix-Alajouanine syndrome, spinal arteriovenous malformation, spontaneous thrombosis, histological finding, vascular wall injury

Introduction

Spinal arteriovenous malformation (AVM) is known to manifest as myelopathy, and subarachnoid and intramedullary hemorrhages.6,8,11,12,14) Progressive myelopathy is caused by venous congestion due to arterial blood flow reflux into the spinal coronal veins through an abnormal arteriovenous shunt.1,4) Subarachnoid and intramedullary hemorrhages are caused by rupture of high-flow perimedullary and intramedullary abnormal arteriovenous shunts. Thrombosis of spinal AVM is much rarer, with only a few case reports, and is called Foix and Alajouanine syndrome.5) Several causes for thrombosis have been suggested, such as subarachnoid or intramedullary hemorrhage,2,13) infection,9) or use of contrast medium,7) but the mechanism of thrombosis remains unclear. Here we present a case of a spinal conus perimedullary arteriovenous fistula (AVF) associated with spontaneous thrombosis and discuss the causes for thrombosis based on the histological findings.

Case Report

A 49-year-old male presented with a 3-week history of slowly progressive paresis and hypesthesia of the left lower extremity. In retrospect, lumbar magnetic resonance (MR) imaging obtained on his first visit to another institution showed swelling of the conus medullaris and its surrounding flow voids, which was supposed to be spinal AVM. However, these findings were misdiagnosed at that time. One month later, right lower extremity paresis suddenly occurred within days.

On admission, he exhibited motor weakness, pain, and hypesthesia of the distal lower extremities, particularly on the right, and decreased knee and ankle jerk reflexes bilaterally. He did not have bladder or rectal disturbance. He was evaluated for the cause of acute deterioration of paraparesis. Lumbar puncture at myelography showed no subarachnoid hemorrhage or infectious disease. Myelography and postmyelography computed tomography demonstrated multiple intradural extramedullary lesions around the conus medullaris. T1-weighted MR imaging obtained after deterioration showed that the flow voids around the conus medullaris had changed from hypointense to hyperintense lesions (Fig. 1). These lesions were thought to be thrombosis of the coronal veins of the spinal AVM. No intramedullary hemorrhage was identified. Spinal angiography was performed carefully with selective
Fig. 1  T1-weighted magnetic resonance images showing that the flow voids (arrow) around the conus medullaris had changed from hypointense (A) to hyperintense lesions (B) after deterioration.

Fig. 2 A: Intraoperative photograph showing total thrombosis of the spinal varicose veins and varices. B: Illustration showing a posterior spinal artery (arrowheads).

Fig. 3 Intraoperative photograph showing an arteriovenous shunt (asterisk), varicose draining vein, and varices.

Fig. 4 A, B: Photomicrographs of the dilated veins showing intimal thickening (asterisk) and disruption of elastic fibers (arrow). Elastica van Gieson stain, original magnification ×20. C: Photomicrograph of varices showing a remarkably dilated vein composed of a luminal thrombus. Hematoxylin and eosin stain, original magnification ×10. D, E: Photomicrographs showing the intima had become extremely thin in large areas and thick in others. Azan stain, original magnification ×40. F: Photomicrograph showing wall dissection. Azan stain, original magnification ×40.

injections of all intercostal and lumbar arteries by neurointerventional specialists, but no abnormal vessels were identified. In retrospect, this clinical course was due to total thrombosis of the draining veins of the spinal AVM. No diagnosis could be established because spinal angiography was negative, although thrombosis of the spinal AVM was suspected.

Surgery with laminoplasty was performed from T10 to T12 to determine the nature of the lesion. Arachnoid thickening and abnormal dilated spinal veins were observed and many of these veins were thrombosed (Fig. 2). Close examination of these veins finally found two direct perimedullary arteriovenous shunts from the posterior spinal artery to the varices and dilated varicose veins (Fig. 3). These two arteriovenous shunts were coagulated and part of the thrombosed vein was removed. The postoperative course was stable and follow-up MR imaging obtained after the operation showed the flow voids had diminished. His neurological symptoms slightly improved. Histological examination revealed remarkably dilated veins com-
posed of luminal thrombi. Characteristic findings were vascular dilatation, intimal alteration, and mural thrombus (Fig. 4). The intima was extremely thin in large areas and thick in others. Wall dissection was also observed. In addition, the mural thrombi had been formed at different times. The final diagnosis of this lesion was spontaneous thrombosis of a spinal conus perimedullary AVF based on the operative findings and specific location.

**Discussion**

Conus AVMs/AVFs generally have multiple direct arteriovenous shunts that derive from the anterior/posterior spinal arteries and have glomus-type niduses and/or varices that are usually extramedullary and pial based, and are always located in the conus medullaris and cauda equina. Symptomatically, these conus AVMs/AVFs can manifest with venous hypertension, compression, or hemorrhage, much like the present case. \(^{13}\) This is a clear case of acute deterioration of lower extremity paresis caused by spontaneous thrombosis of a spinal conus perimedullary AVF. Thrombosis of a draining vein from this lesion is likely precipitate a rapid increase in venous pressure in the remaining draining veins, leading to acute deterioration after a subacute myelopathic course. Only 12 cases with thrombosis of spinal AVM, including our case, have been reported (Table 1). \(^{1-3,5,7,9,10,15}\) These 12 cases included 4 cases with dural AVF, 3 with intramedullary AVM, 1 with perimedullary AVF, and 4 with conus AVM/AVF.

Vascular wall injury of draining veins and varices such as intimal alteration, wall dissection, and mural thrombus could be one of the causes of thrombosis of spinal AVM. Important similar findings support this hypothesis. Vascular wall findings of thrombosed vessels were reported in 7 of the 12 cases. In all 7 cases, vascular wall injury was observed such as vascular dilatation, intimal alteration, and mural thrombi in the walls of varices and varicose veins (Table 1). For example, in two cases of dural AVF, dilated and hypertrophic, “onion-bulb” aspects of varicose veins were observed around the necrotic spinal cord. \(^{5}\) In three cases of intramedullary/conus AVM with large varices, thick mural thrombus of varices were confirmed surgically. \(^{3}\) In an autopsy case of a perimedullary AVF, thick mural thrombus was observed in a varicose draining vein. \(^{9}\) In spinal AVM, retrograde high arterial blood flow into spinal coronal veins may cause vascular wall damage. Vascular wall injury such as intimal alteration or wall dissection could cause mural thrombus as a repair process, which might result in thrombosis. Of course, other causes of thrombosis have been reported such as subarachnoid and intramedullary hemorrhages, \(^{2,15}\) infection, \(^{5}\) or use of contrast medium. \(^{7}\) However, in the present case, subarachnoid and intramedullary hemorrhages and infection were excluded by lumbar puncture and MR imaging. Use of contrast medium was also excluded because the thrombosis had occurred spontaneously before angiography and MR imaging with contrast medium.

**Disclaimer**

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

**References**

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