Spinal cord arteriovenous malformation in a neonate

Case report

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A 2-day-old neonate with a spinal cord arteriovenous malformation developed severe paraparesis. The abrupt neurological deterioration was not associated with hemorrhage or aneurysmal dilatation. Ischemic damage of the spinal cord is suggested as the cause of the clinical manifestation.

Key Words: neonate · paraparesis · spinal cord lesion · arteriovenous malformation

Spinal cord arteriovenous malformations (AVM's) are seldom identified in children. In young children the disorder is particularly uncommon: less than 3% of cases become symptomatic before 10 years of age.14 Only two cases of spinal cord AVM that caused spinal cord dysfunction during the 1st year of life have been reported.16 We are presenting the case of a neonate in whom a spinal cord AVM caused severe paraparesis on the 2nd day of life.

Case Report

This baby boy was the product of a 38-week gestation and was delivered by Caesarean section because of cephalopelvic disproportion. The family history was noncontributory. He suffered from mild transitory respiratory depression. Apgar scores were 3 at 1 minute, 6 at 5 minutes, and 9 at 6 minutes. His neurological condition appeared normal until 24 hours after birth, when he abruptly developed irregular heart rate and blanching over the trunk below the umbilicus, particularly in the lower extremities. Shortly after this event he was noted to have profound leg weakness and loss of sensation to pinprick below T-11 bilaterally. Spinal cord infarct was suspected for which he was started on heparin. He was then referred to the University of Virginia Hospital for further evaluation.

Examination. The baby weighed 2.56 kg and measured 46 cm in length. The anterior fontanel was soft and the head circumference was 33 cm. His back was normal without a cutaneous angioma. He had flaccid paralysis of the lower extremities except for preservation of 3/5 strength of the hip flexors on both sides. Pinprick showed sensory loss below T-11 bilaterally. The anal sphincter was patulous. Reflexes were hyperactive in the upper extremities and absent in the lower extremities.

Two days after birth a metrizamide myelogram was obtained. Lumbar puncture produced a slow return of cerebrospinal fluid, and a satisfactory specimen could not be obtained. Initial injection of contrast material allowed some leakage into the epidural spaces. Repositioning of the needle provided a satisfactory subarachnoid injection. The conus medullaris was markedly enlarged terminating at L2-3, and the filum terminale also appeared thickened (Fig. 1 left and center). Although the anterior spinal artery can be prominent in newborns, it appeared abnormally enlarged and tortuous. Radicular arteries were seen entering the subarachnoid space at T-6 and T-12. Computerized tomography (CT) of the spine confirmed enlargement of the spinal cord, and there was no evidence of hemorrhage within the spinal cord.

The differential diagnosis prior to exploration included a spinal cord AVM, an intramedullary tumor at the conus medullaris, tethered cord with fatty tissue attached to the filum terminale, and embolic infarction of the spinal cord. Although the filum terminale appeared thick on myelography, the spinal cord terminated at L2-3, making the tethered cord syndrome
Spinal cord AVM in a neonate

FIG. 1. **Left:** Preoperative myelogram, frontal view, revealing the prominent vasculature, expanded conus medullaris, and thickened filum terminale. The thickened appearance of the filum terminale was due to a feeding artery accompanying the normal filum terminale. **Center:** Preoperative myelogram, lateral view, showing the expanded conus medullaris and thickened filum terminale. **Right:** Operative photograph demonstrating the arteriovenous malformation lying over the dorsal surface of the conus medullaris and within the cauda equina.

unlikely. Embolic spinal cord infarction was also unlikely, since there was no history of a previous invasive procedure, such as umbilical catheterization, before the infant's development of acute paraparesis. A spinal cord AVM or an intramedullary conus medullaris lesion was therefore suspected prior to surgery. Spinal angiography was not performed because of the risk of increased morbidity in a newborn infant.

**Operation.** At 9 days of age, the patient underwent a laminectomy from L-3 to T-5. Upon exposure of the spinal cord, an AVM was encountered extending over the entire area exposed (Fig. 1 right). The malformation was seen lying over the dorsal surface of the spinal cord and between the nerve roots of the cauda equina. The largest feeding artery appeared to be a dilated artery of Adamkiewicz, entering the spinal canal at T-12 from the right. Five other large radicular arteries supplied the malformation: two in the lumbar region and three in the thoracic region. Accompanying the thickened filum terminale was a dilated ascending artery running cephalad onto the conus. The largest draining vein exited the spinal canal at L-2 on the left; additional veins in the lumbar region drained the malformation. No aneurysmal dilatation was seen over the spinal cord. The conus appeared expanded but lacked any evidence of recent hemorrhage. Under the operating microscope, the arterial feeders were divided just as they entered the subarachnoid space. The entire malformation decreased in volume, and the AVM and draining veins were cauterized without difficulty. The small size of the spinal cord in the neonate precluded an attempt to excise the malformation.

After surgery, the patient's neurological condition remained unchanged with preserved strength of the hip flexors. However, he had not recovered any spinal cord function over the ensuing 3 months.

**Discussion**

This case is unique in that a spinal cord AVM caused catastrophic neurological deficits in a neonate. A review of the literature revealed no similar case; only two cases of spinal cord AVM presenting during the 1st year of life have been reported. Hoffman, et al., described the case of an infant who developed progressive spastic weakness of the lower extremities at 12 months of age. Myelography revealed a complete block at T-8. The spinal cord AVM was explored on two occasions and was found to involve the spinal cord at C6–T10. It was associated with aneurysmal dilatations seen in separate areas of the cervical and thoracic cord. The feeding arteries were ligated intradurally. It is of note that angiography performed after an interval of 7 years demonstrated significant enlargement of the spinal cord AVM. Binder, et al., observed an infant in whom motor development of the lower extremities had come to a standstill at the age of 9 months due to a spinal cord AVM. Myelography revealed a complete block at T-8, and angiography outlined the malformation and an associated aneurysmal dilatation filling the entire spinal canal over T10–12. Fifteen months after excision of the AVM and the aneurysm the child was walking with a leg brace.

Clinical manifestations of a spinal cord AVM appear to vary in different age groups. In adults a slowly progressive course is predominant. Aminoff and Logue noted such a clinical course in 80% of a series of 60 patients, an incidence that conforms to the observation made by Tobin and Layton. Bleeding from a spinal cord AVM occurs in 10% of adults during the course of their illness. In contrast, children with spinal cord AVM's more often present with acute clinical problems. Riché, et al., reported the sudden onset of symptoms in 94% of 34 children afflicted with the disorder. Motor weakness was an initial clinical presentation in more than half of the patients, and bleeding from the malformation occurred in 55%. Other authors have also noted the higher incidence of hemorrhage in children. A number of pathogenetic mechanisms, including hemorrhage, compression, steal phenomenon, increased venous pressure, and thrombosis, have been blamed for the development of neurological deterioration in these cases. In our patient, the abrupt paralysis of the lower extremities was associated with
expansion of the caudal spinal cord. The patient had received heparin shortly after the onset of neurological deterioration, and yet CT and operative examinations revealed no evidence of hemorrhage or aneurysmal dilatation to account for the clinical presentation. We therefore believe that the neurological deterioration in our patient resulted from ischemic damage to the spinal cord.

Chatterjee suggested the hypothesis of steal phenomenon based on the assumption that the blood supply to the AVM and cord are not separate.4,5 Contrary to his belief, however, the angiographic appearance of a common vascular supply to the cord and AVM is rare, except in the cervical region.7 More importantly, symptoms improve after occlusion of feeding arteries.9 At present, the significance of steal remains unknown. Aminoff and his colleagues1 outlined the pathological basis for increased venous pressure within the spinal cord adjacent to the AVM. The spinal cord AVM drains into the plexiform coronal veins lying over the dorsal and lateral surfaces of the cord and eventually into the medullary veins accompanying the nerve roots. The arteriovenous shunt presumably causes an increase in pressure in the coronal veins and their tributaries, the intramedullary veins. The increased venous pressure alters normal capillary membrane transport leading to progressive myelomalacia. This theory may explain the typical clinical presentation of this disorder in adults: fluctuating neurological symptoms, frequent neurogenic intermittent claudication, and symptomatic improvement following occlusion of the feeding arteries. Lastly, spontaneous thrombosis in a spinal cord AVM, known as "Foix-Alajouanine disease," occurs infrequently but can lead to the acute or subacute onset of spinal cord dysfunction.11,15 Rand11 described a patient who became paraplegic within 48 hours in association with thrombosis of the extramedullary vessels. Whether the patient harbored concurrent thrombosis of intramedullary vessels is unknown. In our patient, the exact pathogenesis of the expansion of the conus and the apoplectic onset of neurological deficits is unclear. However, lack of demonstrable hemorrhage, aneurysmal dilatation, or neurological improvement following division of the feeding arteries makes thrombosis a most likely cause of the abrupt clinical manifestation.

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References


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