

SPINAL CORD ARTERIOVENOUS MALFORMATION

A pediatric presentation

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Spinal vascular malformations in child are rare and still under-diagnosed entities that typically lead to progressive spinal cord symptoms and myelopathy if not properly treated¹. This malformation can be differentiated into congenital (arteriovenous malformations and cavernomas) or acquired (dural arteriovenous fistulae) forms¹. Spinal cord arteriovenous malformations (SCAVM) comprise a complex and heterogeneous group of lesions with a number of subtypes that have distinct clinical presentations, anatomic features and natural histories^{2,3}. While there are important differences between subtypes, the majority of patients with SCAVM present first clinical manifestations only in adolescence or adulthood².

We describe the characteristics on child with SCAVM and show the possible treatment of this disease on childhood because only few cases have been published.

CASE

A 4-year-old girl presented sudden onset of lumbar and lower limbs pain, weakness in right lower limb, difficulty walking, constipation and urinary retention. Her parents denied previous similar episodes. She was born at term and was the first child of non-consanguineous parents and the only affected case in the family. The psychomotor development was normal.

Physical examination did not reveal any abnormalities. Neurological examination showed hypotonia in lower limbs (right more than left); muscle strength grade 3 (MRC scale) in the right lower limb and grade 5 in the bilateral upper and left lower limbs; absent right patellar reflex; bilateral Babinski sign; decreased pain, pinprick and light touch sensibility in the right lower limb; and abnormal joint position sense in the right lower limb.

The symptoms and signs were thought to be consistent with spinal cord abnormality, and the investigation showed a cerebrospinal fluid with 5 leukocytes/mm³, 140 erythrocytes/mm³,

glucose 55 mg/dl and protein 25.7 mg/dl; needle electromyography and sensory nerve conduction study were normal, but motor nerve conduction study demonstrated absent F-wave in right peroneal nerve; magnetic resonance imaging (MRI) revealed high signal on T1-weighted images and hypointense signal on T2-weighted images in area of the T11–T12 spinal cord with concomitant swelling of the T11–L1 cord (Fig 1); and selective spinal angiography demonstrated a small arteriovenous fistula with posterior aneurysmatic formation on T11–T12 spine cord area which was fed by the anterior spinal artery arising from the right 12th and left 9th intercostal arteries (Fig 2A and 2B).

After this initial investigation the patient was submitted to management of the SCAVM. Surgery was performed with clipping of the supplies arteries resulting complete obliteration of the fistula on postoperative spinal angiography (Fig 2C). Follow-up shows that her symptoms and neurological status improved after surgery.

All studies were done following informed consent of parents.

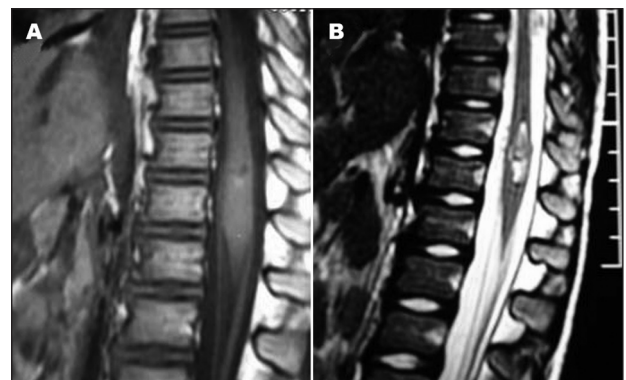


Fig 1. [A] Sagittal T1-weighted MRI shows the main arteriovenous shunt located in the thoracic spine (T11–T12). [B] Sagittal T2-weighted MRI reveals swelling due to spinal cord venous congestion between T11–L1 levels.

MALFORMAÇÃO ARTERIOVENOSA MEDULAR: APRESENTAÇÃO PEDIÁTRICA

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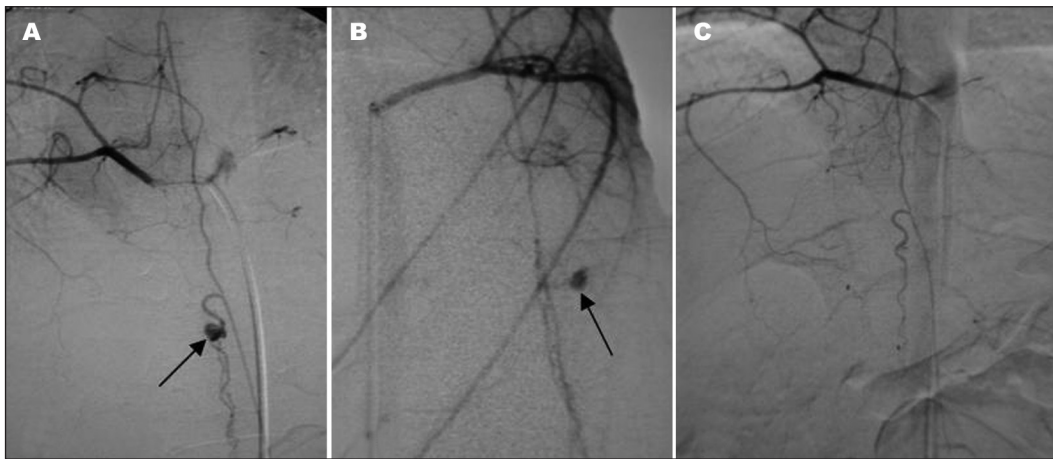


Fig 2. Selective spinal angiography demonstrated a small arteriovenous fistula with posterior aneurysmatic formation (arrows) on T11–T12 spine cord area which was fed by the anterior spinal artery arising from the right 12th and left 9th intercostal arteries [A and B]; with complete obliteration of the fistula on postoperative spinal angiography [C].

DISCUSSION

SCAVM is a rare congenital vascular malformation of the spinal cord characterized by an arteriovenous shunt, with or without an intervening vascular network, between the feeding artery and the draining vein³⁻⁵. They are fed by one or many arteries that normally supply the spinal cord⁵. The SCAVM incidence ranging between 20 to 30% of all spinal vascular shunts, and it were rarely observed during the childhood period^{1,6}. SCAVM presentation in the first 2 years of life is exceedingly rare².

Clinical manifestation of the SCAVM depending of the degree of the malformations, but the pathogenesis in spinal cord includes venous congestion and hemorrhage causing myelopathy¹. Clinical manifestations can include pain, acute or progressive myelopathy, and radiculopathy⁷. A minority of patients with SCAVM are symptomatic as neonates or infants². In these cases, symptomatology is unspecific if the patient did not present an acute hemorrhage¹. Neurological symptoms are attributable to the spinal cord lesion and patients can present weakness or paraplegia associated with sensory deficit or sphincter dysfunction, similar our case². In some cases, SCAVM can be seen genetically associated with hereditary hemorrhagic telangiectasia^{2,4,6}.

Natural history of neurovascular disease in the pediatric population differs from that of adults in many significant ways, and SCAVM are no exception. Hemorrhagic presentation is more frequent in pediatric population than in adults, except in extremely young children⁶. Importantly, however, neonatal and infant age groups represent a yet more specific group into pediatric population^{2,6}. For example, in contrast to report of Rodesch et al which found that 70% of pediatric patients with SCAVM presented with hemorrhage (vs 45% of adults), in neonatal and infant cases reported by Cullen et al, hemorrhagic presentation was

23%^{2,6,8}. SCAMV causing venous congestion possibly could be the main pathophysiology feature in neonatal and infant cases². In addition, while nidus SCAVM type predominates in the pediatric population (67%), the majority of patients have fistulas in the neonatal and infant series^{2,8}.

MRI diagnosis is characterized by serpiginous or tubular structures, corresponding to large draining veins⁴. The MRI can still show the effect of the vascular malformation in the spinal cord: hemorrhage, congestion, compression and deviation by vessels abnormalities⁴. However, it is difficult to precise the shunt involved (fed, drainage, type and localization) by MRI^{4,9}. Angiography remains the gold standard for analysis of the anatomical, morphological and architectural features necessary for therapeutic decisions in both pediatric and adult populations⁴. When a SCAVM of any type is clinically suspected on the basis of unexplained myelopathy or hemorrhage, angiography should be performed even if prior noninvasive imaging studies were unrevealing⁹.

They have given rise to a large literature, dealing with different analyses that have oriented patients towards various treatment modalities that range from surgery, embolization or a combination of both^{3,4,10}. Experiences suggest that endovascular therapy should be the primary treatment of these lesions in the neonatal and infant population^{2,4,6}. However, fistulous SCAVM with a small caliber of the feeding artery can prohibits catheter placement close to the fistula; therefore, a surgery treatment should be favored, as in our case¹. Fistulous SCAVM with dilated feed vessel can be submitted to a surgical management or a super selective catheterization to close the fistula by embolized procedures^{1-3,10}. In infants, despite early presentation and severe symptoms, these lesions are frequently amenable to endovascular treatment, often with anatomic cure achieved and with improvement or stabilization of symptoms after partial targeted treatment⁶. A

partial embolization, to obliterate the nidus in glomerular SCAVM, also can improve the prognosis of the patient^{1,3}. In juvenile SCAVM, only those parts of the SCAVM responsible by symptomatology should be treated^{1,3}.

Despite the increasing use of endovascular techniques there is a significant role for operative management of these conditions at infancy^{3,10}. Therefore, treatment should be individualized and integrated by endovascular and surgical procedures, depending on lesional angioarchitecture and clinical status of the patient⁷.

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