Trifocal Monomyelomeric Spinal Cord Arteriovenous Fistulae in a Seven-Year-Old Boy

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Summary

We describe a rare case of multiple arteriovenous fistulae of the spinal cord (SCAVF) in the same myelomer in a five-year-old boy. This case report consists of a trifocal SCAVF at the Th12 myelomeric level without communication between the three different fistulae. Two AVF were located posteriorly, bilaterally, in the spinal cord, fed by left and right posterior radiculopial arteries and one anteriorly in the anterior spinal axis. The venous drainage was independent for each lesion. The patient presents associated lesions characterized by cutaneous stain and inferior limb asymmetry. A metameric distribution is the explanation for the multiplicity of these lesions in a syndromic association related to Cobb syndrome. The patient was treated by transarterial embolization using glue with occlusion of the three different fistulae. The patient achieved a good improvement in neurological status.

Introduction

Multifocal Spinal Cord AVM are exceptional compared to multifocal cerebral AVM. The incidence of SCAVM is about one-tenth of brain AVM¹. The symptoms related to SCAVM in

most cases occur during the second or third decades of life². They are rarely diagnosed in children. Multiple brain arteriovenous malformations are usually associated with Rendu-Osler-Weber disease (ROW) that presents typical features in children represented by high flow single hole arteriovenous fistula and venous ectasia³. A trifocal SCAVF in the same myelomer has never been described before.

Case Report

A five-year-old boy presented sudden pain in both limbs associated with weakness and sphincteral disturbances followed by paraplegia. Lumbar puncture with the CSF analysis revealed SAH. He presents cutaneous "café-au-lait" patches on the face, right lumbar region and posterior portion of the right limb. There is also limb asymmetry with the right side shorter than the left.

Spine MRI revealed a SCAVM at the Th12 level, with hypersignal next to the malformation in the venous aneurysm corresponding to a clot (figures 1, 2). Spinal angiography performed showed a trifocal SCAVM in the Th12 level (figure 3). The first nidus was located in the anterior spinal axis, fed by a radiculomedullary artery from left Th6 anastomosing with a

Figures 1 and 2 Axial and sagittal MR views show abnormal vascular structures in Th 12 level corresponding to the SCAVM. The hyperintensity signal in the sagittal view corresponding to thrombus in metehemoglobin phase inside the venous aneurysm (arrow). There is also a hypersignal in the spinal cord itself, corresponding to cord suffering.

Figure 3 Drawing with the three SCAVF in Th12 myelomeres. One fistula is located anteriorly in the anterior spinal axis, fed by the left Th6 artery (A), draining through right and left L3 veins (V) and the other two fistulae are located posteriorly. Note the independent venous outlet for each fistulae. The posterior fistula on the right side is fed by Th9 and Th12 radiculopial arteries (A) drains through the right L5 veins (V) and the fistula on the left side fed by Th11, Th12 and L2 arteries (A) and drains through the right L1 vein and the L5 vein (V).

radiculomedullary artery from left L1 (figures 4, 5). The second malformation is an arteriovenous fistulae located posteriorly, fed by a radiculopial artery from right Th9 and Th12 (figures 6, 7). The third malformation corresponds to an arteriovenous fistulae located posteriorly, on the left side, fed by a radiculopial artery from left Th11 and Th12 (figures 8, 9). There is no communication between these three malformations and the venous outlet is different in each AVF.

Embolization using histoacryl in a high concentration was done in the left Th11 and right Th12 achieving immediate total occlusion in the two posterior AVF (figures 10, 11). The AVM in the anterior spinal axis was also embolized through superselective catheterism of left L1 artery with occlusion of most of the venous drainage. A control angiogram confirmed the total occlusion of the A-V shunt preserving the anterior spinal axis (figure 12). Clinically, the patient has improved after embolization.

Figures 4 and 5 Selective angiography in the left Th6 and left L1 intercostal arteries shows the SCAVF in the anterior spinal axis (arrow). Note the venous drainage through right (arrowhead), left (double arrowhead) L3 vein and the filum terminale vein (triple arrowhead).

Discussion

Spinal cord arteriovenous fistula (SCAVF) was firstly described by Djindjian et Al. The arteriovenous shunt is located in the subpial space⁴.

In Rodesch et Al⁵ series, SCAVMs occurred in 17% in children under 15 years old⁵. Only 20% of these cases correspond to a direct arteriovenous fistulae. No multiple lesions were found in the paediatric population. Most of the lesions (60%) were revealed by hemorrhage. The apparent aggressiveness of the disease is the same as in a similar adult case, and the early presentation reveals the weakness and failure of the remaining vascular tree^{6,16}.

In children, most of the symptoms have a sudden onset^{4,7}. Single hole A-V fistula usually presents with hemorrhagic accident and not

with acute non hemorrhagic onset^{5,16}. According to Rodesch⁶, venous ectasia can be encountered in 32% of cases and are most frequently found in the thoracolumbar region (68%). These pouches may be intralesional or related to a kinked megadolicoectatic draining vein. These ectasias may be "open" with a distal outlet or "closed" when the exit vein is apparently occluded. These closed pouches may represent the stump of a thrombosed draining vein⁸. The arterial pressure that reigns in these venous ectasias may lead to their rupture with subarachnoid or intramedullary hemorrhage (42%). On the other hand, large pouches may compress the nervous parenchyma and contribute to the progressive onset of neurologic symptoms. Primary treatment should be targetted to the highest flow to decrease the venous drainage congestion and shrink the venous pouches⁶.

Figures 6 and 7 Selective angiography in right Th9 and Th12 arteries with the second fistulae located posteriorly in Th12 myelomer (arrow) on the right side. Note the venous drainage through the right L5 vein (arrowhead).

Single hole high flow arteriovenous fistula have been described in ROW disease and may be associated with giant venous ectasia that is responsible for the occurrence of a cord swelling and venous ischemia secondary to chronic venous hypertension or mechanical compression and pulsatility of the sac⁹. Associated AVM of CNS are present in approximately 8% of patients with ROW. Willinsky et Al¹⁰ reported that 28% of patients with multiple CNS AVM had ROW. Angioarchitectural analysis of these vascular lesions showed that most were direct pial A-V fistulae, without nidus, with a huge ectatic draining vein. There were no clinical or angioarchitectural differences between the A-V fistula associated with ROW and sporadic A-V fistula, the diagnosis being based on the family history³.

Few reports have been published on multiple spinal cord AVM with a multimyelomeric in-

volvement associated or not with vertebral or skin lesions^{2,11}. The present case is the first one with multiple different spinal cord AVF located at the same myelomer. Berenstein et Al reported that 28% with SCAVMs had associated soft tissue vascular malformation or dysplasia and 10% had associated metamer lesions^{7,16}.

Matsumaru et Al described an incidence of 16% of SCAVM associated with other soft tissue vascular malformations in a metameric distribution and these represent the most frequent type of multiplicity. Among 19 patients with multifocal and metameric SCAVM there were bifocal SCAVM in three cases¹¹. Y. Niimi et Al¹² described a case with two distinct single-hole perimedullary AVF near the conus at two different metamers supplied by the left posterior spinal artery associated with Parkes-Weber Syndrome. Identification of the myelomeric level involved in SCAVM allows a segmental

Figures 8 and 9 Selective angiography in the left Th11 and Th12 arteries showing the third SCAVF located posteriorly in Th12 myelomer, on the left side. Note the venous aneurysm (double arrowhead) and the anastomosis point between the left Th11 and Th12 (arrow). See the fistula point in Th12 level (double arrow) and the venous drainage by the right L1 and left L5 vein (arrowhead).

Figures 10 and 11 Immediate post embolization angiogram in the right Th12 and left Th11 arteries after embolization showing total occlusion of both posterior SCAVF (compare with figures 7 and 9 respectively).

Figure 12 Control angiogram in left Th6 six years after embolization with total occlusion of the A-V shunt, preserving the anterior spinal axis.

link between various lesions of mesodermal or neural crest origin¹¹.

In the present case, the involvement of skin and spinal cord in the same myelomer constitutes the features of a "metameric Cobb Syndrome", that could be explained by a dysplastic genetic defect involving the segmented mesodermic origin of the endothelial cells to establish the topographic link between these metameric targets¹³.

Therefore, multiple A-V shunts become an expression of a disease resulting from a group of biological dysfunctions involving the vessel wall remodelling when an earlier causative event occurred creating a high chance of apparent multifocality as a reply to an area of genetic dysfunction^{14,15}.

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