Postero-lateral arterio-venous fistula with particular histological aspects as the main cause of severe spinal thoracal stenosis. Case report

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Postero-lateral arterio-venous fistula with particular histological aspects as the main cause of severe spinal thoracic stenosis. Case report

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ABSTRACT
Spinal dural arteriovenous fistulas (AVFs) are rare spine vascular malformations. We report a case of AVF in the epidural space of T10 and degenerative medial osteophytes leading to clinical symptoms of severe spinal stenosis operated with good evolution.

Object: Our report's objective is to present a particular, extremely rare case of spinal stenosis at T10-T11 level, mostly on the right side, result of the cumulative effect of a vertebral postero-lateral AVF, medial degenerative osteophytes and a synovial cyst at the same level. The AVF is the main cause of the spine stenosis.
Case Report: A 69 year-old man presented for 5 years' pain in the thoracic spine, intermittent paresthesias on the right leg. From one week he complains of painful paresthesias on both legs (predominant on the right side), gait with progressive difficulty.
On examination we observed incomplete paraplegia (Frankel C+/E), sensory examination revealed hypoesthesia with sensitive level T10, knee and ankles reflexes increased bilaterally with predominance on the right side, Babinsky sign present bilateral.
On MRI imaging: Thoracal spine stenosis T10-T11, by degenerative osteophytes T10-T11 predominant on the right side, synovial cyst in the lateral recess who severely compresses the dural sac and the spinal cord.
The patient was operated (Laminectomy T10 and T11 on the right side, microsurgical complete resection of a heterogenous extradural
lesion (bony and vascular, intensively bleeding), severely compressive over the spinal cord.

The postoperative evolution was favorable: the incomplete paraplegia has improved (ASIA C+ on the right side, D on the left side), hipoesthesia diminished.

Figure 1. Coronal T2.: Dorsal spine stenosis T10-T11, by degenerative osteophytes T10-T11 predominant on the right side (Blue arrow) who severely compress the spinal cord.

Figure 2. Sagittal T2: Severe spine stenosis produced by degenerative osteophytes T10-T11 and synovial cyst (green arrow).

Figure 3. Axial contrast T1: Dorsal spine stenosis T10-T11, by medial degenerative osteophytes T10-T11 predominant on the right side (red arrow).

THE HISTOLOGICAL EXAM

Material and method: The probe was sent fresh with a cold ischemic time of 20 minutes, then it was paraffined embedded.

The conventional slides (Haematoxylin – eosine stained) revealed a conjunctive tissue with coiled vessels, some dilated, some collapsed, attached on a spinal node.

Immunohystochemical method was done on Ventana platforme GX, with antibodies CD34 (CLONA QBEnd10); WT1 (CLONA 6FH2)

COMMENTS

Wilms' tumor-1 (WT-1) was originally described as a tumor suppressor gene based on its mutational inactivation in a subset of Wilms' tumor. It plays an essential role in haematopoiesis and angiogenesis by regulating vascular endothelial growth factor, angioproteins, nestin, and proliferation of vascular smooth muscle cells. Human skin vasculature shows cytoplasmic WT1 protein expression, detected by an antibody recognizing the C-terminal of the protein (6F-H2). Reports have demonstrated that WT-1 protein is expressed in a variety of vascular anomalies. Defects in WT1 signalling might underlie the inability of endothelial cells in vascular malformations to undergo physiological apoptosis and remodelling.
CD34 - Commonly used marker of hematopoietic progenitor cells and endothelial cells; Also called hematopoietic progenitor cell antigen CD34; CD34+ stromal cells are called dendritic interstitial cells;

CD34: Membranous stain
WT1: nuclear stain

Endothelium of normal vessels acts as a positive internal control

In our case, the positive reaction was noted in a greater number of vessels to CD34 than to WT1, so the interpretation, correlated with clinical and imagistic data, was of a vascular spinal fistula with loss of WT1 expression (as in AVMs). We couldn’t find other reports about WT1 expression in spinal AVFs.

Figure 4. Cd34, ob4x

Figure 5. Cd34, ob 10x; positive in endothelial cells and in some interstitial dendritic cells

Figure 6. Wt1 positive in a normal vessel, negative in other vessels

Figure 7. Postoperative CT scan

**FOLLOW UP AT 6 MONTHS**

**Discussion:** Extradural (or epidural) AVFs are relatively uncommon malformations characterized by anomalous communication between an extradural branch, usually of a radicular artery, and the epidural venous plexus.

Spinal arteriovenous malformations (AVMs) (constituting 3-4% of all spinal cord space occupying lesions).

After Lim the symptoms of epidural AVFs are related to the pattern of venous drainage. If they are...
exclusively epidural, they may present with local pain or radicular pain or with progressive myelopathy.

Spetzler et al\textsuperscript{11} proposed a modified classification of the spine/spinal cord arteriovenous lesions to arteriovenous malformations (AVMs) and arteriovenous fistulas (AVFs). AVFs are classified according to their location as intradural and extradural. According to Geibprasert et al\textsuperscript{6}, dural AVFs are classified on anatomical bases in ventral group (most of them) and dorsal epidural group.

The dorsal group of dural AV fistulas is related to veins that normally drain the spinal process and lamina at the spinal level. Patients with dural AV fistulas within this space typically present with spontaneous epidural hematomas. These symptomatic lesions are extremely rare. Spinal dural AV fistulas are fed by the radicular arteries and/or the surrounding vertebral branches.

After classification of Spetzler et al\textsuperscript{5} there are extradural AV fistulas ventral or dorsal, with single or multiple feeders. There is no gender predilection (male 52.9\%) of AVFs.

Common to unruptured AVFs are symptoms of myelopathy and pain, such as lower or upper extremity weakness, abnormal sensory, disturbance of gait, back pain, and bladder and/or bowel incontinence. Most cases of ruptured AVFs are manifested as spontaneous epidural hematoma (64.7\%). Typically, initial symptoms of spinal epidural hematoma are rapid development of excruciating back pain, with or without neurological deficit, followed by rapidly progressive severity of myelopathy, such as abnormal sensory, disturbance of gait, bladder and/or bowel incontinence, lower or upper extremity weakness/paralysis due to the location of hematoma.

After Steinmetz\textsuperscript{10} there are two options in the treatment of spinal dural AV fistulas; surgical occlusion of the intradural reflux vein, and endovascular therapy employing embolic material into the fistula. Surgery is a relatively simple and safe intervention, resulting in long-term shunt occlusion in 98\% of cases. Any AVFs with progressive myelopathy have to be treated as soon as diagnoses are made. After Lim\textsuperscript{4}, surgical disconnection of the intradural radicular vein is the curative method, so is endovascular occlusion of the fistula and proximal part of its venous drainage with liquid embolic material. Prompt diagnosis and emergency surgical treatment are crucial.

The best management for AVFs is still surgical operation\textsuperscript{11}.

Our case illustrates a severe compression of the thoracic spinal cord produced by a combination of degenerative lesion (medial degenerative osteophites T10-T11) and posterior extradural AVF. We have realized a correct treatment of these lesions with good results.

**Conclusions**

Surgical treatment was the mandatory method of healing of the AVF and decompression of the dorsal spinal cord.

The delay in diagnosis left residual incomplete paraplegia.

**References**
