A 53-year-old man presented with urinary retention, bilateral lower extremity weakness, and numbness after a L2–L5 lumbar laminectomy 6 weeks prior. MRI of the lumbar region revealed spinal cord edema and dilated peri-medullary vessels on T2 images. Digital subtraction angiography revealed early venous filling arising from the segmental T12 artery supplying the spinal cord, confirming the diagnosis.

Spinal dural arteriovenous fistulas (AVFs) are the most common vascular malformation of the spine. It is presumed that spinal AVFs are an acquired disease although their exact etiology is unclear. The diagnosis is suspected when patients present with symptoms of spinal cord venous congestion [1]. Spinal cord edema may be at a distant location from the arteriovenous shunt.

Connective tissue diseases such as fibromuscular dysplasia, neurofibromatosis type 1, Marfan syndrome, and Ehlers–Danlos syndrome are often seen with AVFs [2]. Arteriovenous fistula may be associated with RASA1 gene mutations as part of a capillary malformation-arteriovenous malformation (CM-AVM) syndrome or Parkes Weber Syndrome [3]. CM-AVM and Parkes Weber syndromes are characterized by multiple small capillary malformations and arteriovenous fistula. Capillary malformation along the involved dermatome in association with AVF should raise suspicion for Cobb’s syndrome.

Currently, no serum or genetic testing is routinely indicated in patients with arteriovenous fistulas. In patients with an associated syndrome, genetic testing for RASA1 gene mutations may be offered, although other unidentified genes are also likely involved in all of the above mentioned syndromes. Treatment of spinal arteriovenous fistulas is by endovascular embolization or surgical occlusion of the intra-dural vein [4].

Figure 1. Spinal angiogram and thoracic MRI site revealing evidence of an arteriovenous fistula. (a) Superselective injection of the left T12 artery (red arrow) in arterial phase showing early venous filling from the site of the fistula (green arrow) and retrograde flow through an arterialized tortuous intradural extra-medullary spinal vein (blue arrow). (B) T2 sagittal view revealing spinal cord edema (red arrow) and dilated peri-medullary veins (blue arrows).

We searched PubMed to identify articles using the search terms (and synonyms) “arteriovenous fistula” in combination with “spinal cord,” “dural,” “imaging,” “genetic testing,” “serum,” and “markers”. HS and MB selected papers identified through these searches, supplemented with additional papers from personal records. High impact reviews and where appropriate original papers were included from 1995 to 2017.
References