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Foix Alajouanine Syndrome Mimicking Longitudinally Extensive Transverse Myelitis

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Foix-Alajouanine Syndrome Mimicking Longitudinally Extensive Transverse Myelitis

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Introduction

In 1926, Drs. Foix and Alajouanine described subacute congestive myelopathy leading progressive paraplegia and eventual death of two men. Upon autopsy, they discovering necrosis of the spinal cord and multiple tortuous, thickened blood vessels on the surface of the spinal cord. They called this necrotizing myelopathy[1]. Several years later, this necrotizing myopathy was associated with the presence of a small but abnormal connection between a radicular vein and artery, or arteriovenous fistula (AVF).

AVF can create increased venous pressure and subsequently a decrease in the arteriovenous pressure gradient, leading to a decrease in spinal cord perfusion, and consequently spinal cord edema. The lower part of the spinal cord is most often affected due to gravity of the lower spinal cord and lack of valves in the intraspinal venous system[2].

Foix-Alajouanine Syndrome continues to remain a rare condition. A neurologist may only see one case approximately every 4-8 years [2]. It is also often misdiagnosed, underdiagnosed, or diagnosed late, which may lead to the patient receiving ineffective treatments or surgeries, and poor outcomes[2,3]. Here, we present one such case with unique radiographic findings and a relatively early diagnosis and treatment.

Case Description

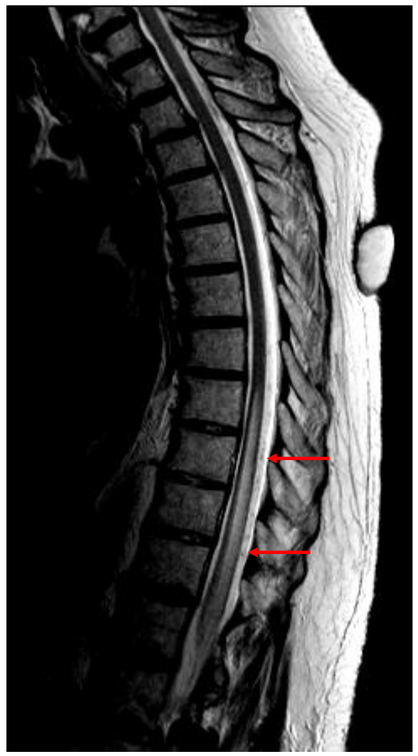
A 77-year-old man presented to clinic with 8 months of progressive lower extremity weakness, paresthesia, gait instability, and recurrent falls to the point that he had incurred a fracture to his orbital floor. He had no lumbar or leg pain. He had no urinary sphincter dysfunction.

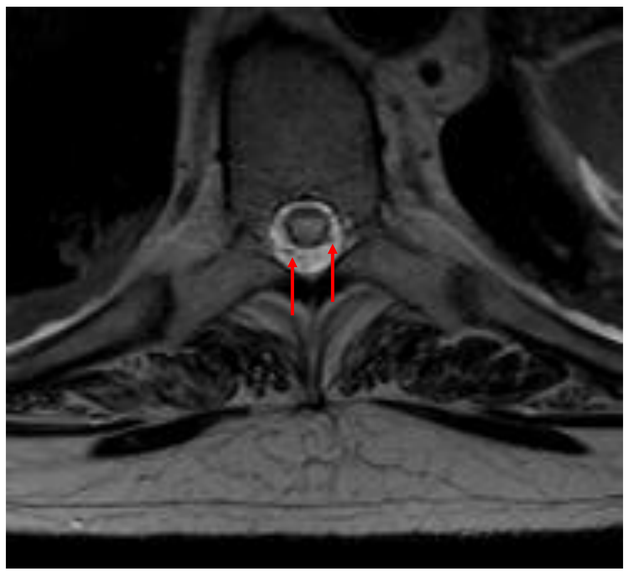
Prior to presentation to clinic, his symptoms were attributed to lumbar spinal stenosis and he had undergone an L3-L4 lumbar decompression with no improvement. He had also undergone an EMG which was reported to be normal.

Workup included a normal MRI of the brain and cerebrospinal fluid analysis including Aquaporin-4 antibody, IgG index, and myelin oligodendrocyte glycoprotein (MOG) antibody. MRI of the lumbar spine without contrast showed a longitudinally extensive transverse myelitis (LETM) from T8 through the tip of the conus medullaris and flow voids at the dorsal aspect of consistent with a spinal dural arteriovenous fistula (SDAF). This was confirmed by the spinal angiogram, with findings confirming a Type I SDAF diverting flow from the Artery of Adamkiewicz.

The patient underwent microsurgical obliteration of the SDAF and regained 50% of his lower extremity strength within 48 hours.

Radiographic Images





FIGURES 1, 2: MRI of the lumbar spine without contrast (sagittal view on the left, axial view above) showing a longitudinally extensive transverse myelitis (LETM) from T8 through the tip of the conus medullaris and flow voids (red arrows) at the dorsal aspect of T9-L1.

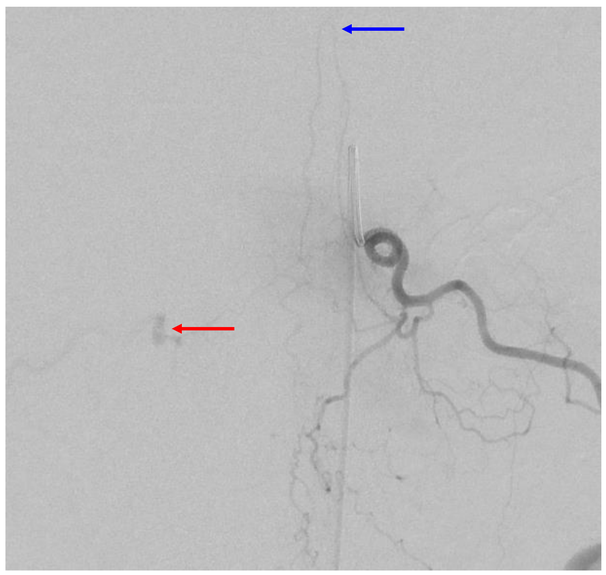


FIGURE 3: Catheter angiography of the lumbar spine showing a Type I spinal dural arteriovenous fistula (red arrow) diverting flow from the Artery of Adamkiewicz (blue arrow)

Discussion

Patients with Foix-Alajouanine Syndrome often present with subacute spastic paraparesis which can progress over 1-5 years to flaccid paraparesis. They may also exhibit gait imbalance, sensory symptoms, radicular pain, and/or urinary retention. Interestingly, this patient did not exhibit any urinary sphincter dysfunction which may be attributed being in the early phase of the disease[2] .

Due to a nonspecific presentation of symptoms, this condition can often be misdiagnosed for neuromuscular conditions. Patients can also often undergo operations for lumbar disc prolapses not unlike this case. Therefore it is important to differentiate between compressive and non-compressive myelopathy prior to offering a patient surgical intervention[2,4].

However given this case, AVDF should also be considered in the diagnosis for LETM, which is also a rare condition. The most common causes for LETM include NMO Spectrum Disorder, Multiple Sclerosis, paraneoplastic, neoplastic, or metabolic disorders. The treatment for many autoimmune causes of LETM includes intravenous steroids. Unfortunately pulses of intravenous methylprednisolone can cause immediate and severe worsening of motor and sensory symptoms when administered to patients with SDAF[3]. Thankfully, the presence of flow voids in the posterior epidural space of the thoracolumbar spinal cord prompted us to consider a vascular etiology for this patient's LETM. Flow voids are tortuous and dilated veins that can be found in 35–91% of patients, with increasing sensitivity of detection with the advancement of MR technique. Catheter angiography continues to remain the gold standard in SDAF[2] .

Treatment is either endovascular embolization with liquid polymer (46% technical success) or surgical ligation of the fistula (98% technical success), as underwent by this patient[5]. Duration of symptoms is a significant contributor to outcomes. As this patient was treated within one year of symptoms, this is likely a major factor in his return to baseline. As a result, SDAVF is a rare but reversible cause of Foix-Alajouanine Syndrome and should be considered in the differential diagnosis of LETM.

References

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