INTRODUCTION

Foix-Alajouanine Syndrome was founded by neurologists Charles Foix and Theophile Alajouanine in 1926 and was described as a rare myelopathy caused by dural arteriovenous malformations (DAVF) of the spinal cord. An arteriovenous fistula (AVF) is an abnormal shunt between an artery and a vein. Spinal AVFs are characterized by tortuous sub arachnoid veins due to the invading increased meningeal arterial pressure. The etiology of this syndrome involves a reflux of arterial blood typically supplied by the meningeal arteries, into their respective dural venous plexus. This causes constriction of venous outflow of the spinal cord, and the increase in pressure leads to venous stasis, thrombosis, and ultimately necrotic myelopathy. The most affected areas are the thoracic and lumbar spine. The present case highlights an important point, that when clinical and radiological suspicion for a spinal vascular lesion is high, an inconclusive CT does not rule out spinal AVF and should not mislead clinicians. Follow-up imaging with MRI showing a T2 hyperintensity with flow voids in the spinal subarachnoid space and direct visualization of the fistula with Digital Subtraction Angiography (DSA) are gold standard for definitive diagnosis [1]. This is conjunction with embolization or minimally invasive microsurgery leads to ligation of the fistulae and potential resolution for the patient.[2]  

THERAPEUTIC INTERVENTIONS

The suspicion of the DAVF along with the evidence of brainstem edema and fourth ventricular hemorrhage, necessitates a right frontal Burr hole and placement of an external ventricular drain. Neurosurgery clears the patient for C7/T1 Laminectomies with ligation of the C8 DAVF. While exploring the intradural compartment, they identify a large which is densely adherent to the C8 motor root. There are also multiple tortuous dilated veins noted on the surface of the spinal cord, confirming venous hypertension due to this fistula. Using meticulous microdissection techniques, they fully separate the multiple venous structures arising from the nerve root sleeve from the motor fibers themselves. This procedure reveals a left-sided DAVF at the level of C7-T1, which is treated utilizing Onyx 18 embolization material. Evidence emerges of an enlarged branch arising from the left thyrrocervical trunk supplying a symptomatic right tentorial DAVF, characterized by Borden grade III and Cognard grade III. This vein is clipped successfully, and subsequent Digital Subtraction Angiography scans confirm the ligation of the fistula with evidence of a few more remaining Type 1 fistulas around the C3/C4 region. Under surgical exploration, a large tortuous vein is found, arising from the right C4 nerve root sleeve, which was embedded with a division of the ventral motor rootlets of C4. This vein is clipped, and a repeat DSA scan confirmed cessation of arterialized flow within the visualized vein.