Case Report- Foix Alajaouanine Syndrome

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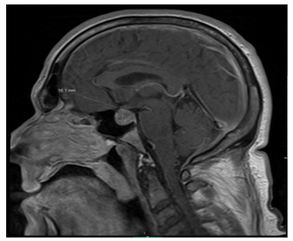
INTRODUCTION

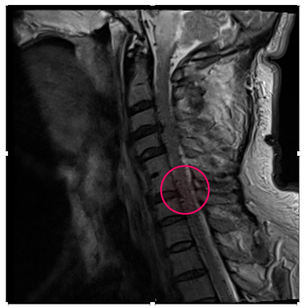
Foix-Alajouanine Syndrome was founded by neurologists Charles Foix and Theophile Alajouanine in 1926 and was described as a rare myelopathy caused by dural arteriovenous malformations (DAVF) of the spinal cord. An arteriovenous fistula (AVF) is an abnormal shunt between an artery and a vein. Spinal AVFs 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 congestion 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 AVM 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

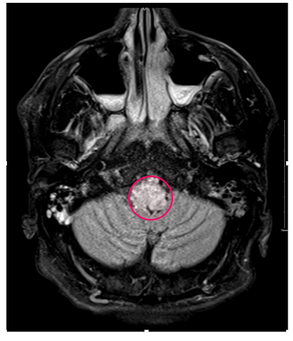
The suspicion of the DAVF along with the evidence of brainstem edema and fourth ventricular hemorrhage, necessitates a right frontal burr hole and placement of an external ventricular drain. Neurosurgery clears the patient for C7/T1 Laminectomies with ligation of the C8 DAVF. While exploring the intradural 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 thyrocervical 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.



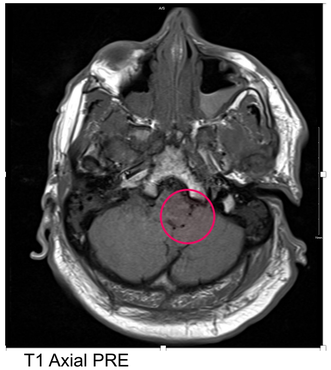
SAG T1 POST GAD BRAIN MRI W/O contrast



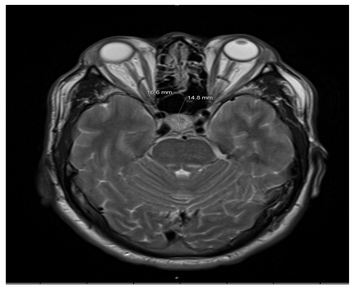
SAG T2 CSP MRI Spine Cervical Contrast (Flow Voids)



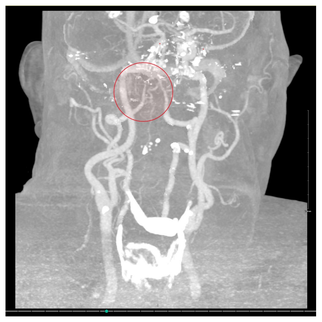
Flair ax MRI BRAIN WO CONTRAST
(stable 4th ventricular hemorrhage)



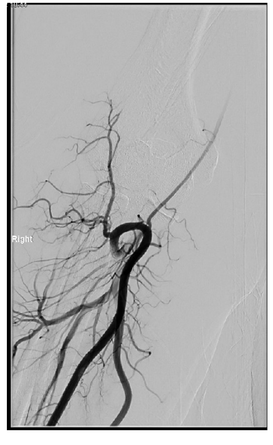
T1 Axial PRE
MRI Brian WO Contrast
Multiple intracranial flow voids



AX T2 Brain MRI Brain W WO Contrast ; Pituatary Macroadenoma abnormal signal involving the pontine medullary junction with subtle enlargement of the medulla



CT ANGIOGRAM HEAD NECK W WO CONTRAST - Numerous abnormal small dilated veins surrounding the medulla and cervical cord corresponding with flow voids seen on recent MRI cervical spine



IR NEURO ANGIO- Right petro-tentorial dural arteriovenous fistula, Right C3-C4 type I spinal dural arteriovenous fistula

CASE PRESENTATION

The patient is a 58-year-old male with a history of hyperlipidemia, hypertension, and chronic headache, who presents to the ED with complaints of intractable nausea, vomiting, vertigo, worsening vision, dysarthria, dysphagia, lip swelling, dysphonia, and posterior headaches. He has an abnormal blood pressure of 202/109 with an inconclusive CT of the head. In addition, the patient develops epiglottic swelling that requires mechanical ventilation. An MRI of the head and spine is taken the next day, which reveals a pituitary macroadenoma, prominent flow voids at the pontomedullary junction, and extensive transverse myelitis throughout the cervical spine. In the following week, his symptoms significantly deteriorated with bilateral leg weakness and a further deterioration in balance. The situation exacerbates with a rapid decline in patients' mental status and the development of acute onset seizures. A CT reveals acute bleeding in the fourth ventricle of approximately 13 mm with stability and without midline shift.

DISCUSSION

Cervical DAVFs are rare, and their presentation can be complex. This case highlights the challenges of diagnosing cervical DAVFs, and myelopathic symptoms that fail to respond to standard treatment should lead to a high index of suspicion. The clinical presentation of DAVFs ranges from symptoms of hemorrhage to progressive myelopathy with a broad, non-specific presentation. There should be a low threshold for a repeat MRI or progressing to DSA when faced with a patient who fails to improve, or further deteriorates, despite commencing treatment for myelopathy. The successful treatment of DAVFs should aim for the complete disconnection of venous drainage. Treatment decisions are based on symptom severity and the grade of DAVF, with improving functional outcomes possible after definitive management.

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