(S83) FOIX-ALAJOUANINE SYNDROME MIMICKING DEMYELINATING DISEASE

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Background: Foix-Alajouanine syndrome is an acquired spinal dural arteriovenous fistula (SDAVF) presenting as a progressive myelopathy that can be mistaken for many disorders, thus delaying diagnosis and treatment. Objectives: To describe Foix-Alajouanine syndrome in the differential diagnosis of demyelinating disease. Methods: Case presentation and literature review. A 65-year-old man with family history of multiple sclerosis (MS) presented with a 4-year history of progressive lower-extremity paresthesias worsening over 4 months; a 3-year history of urinary retention and erectile dysfunction; and 1 year of fatigue, progressive leg weakness, and gait imbalance. Work-up for myelopathy included magnetic resonance imaging (MRI) and spinal angiography. Results: MRI showed two nonspecific white-matter brain lesions; mild thoracic cord expansion, abnormal T2 signal from T7 to T12, and contrast enhancement. Transverse myelitis was suspected by an outside provider, but a spinal angiogram demonstrated a right-sided SDAVF arising from T6/T7 segmental arteries, which was successfully treated with onyx embolization. Conclusions: SDAVF is the most common vascular malformation of the spinal cord, yet diagnosis is frequently delayed months to years because of a nonspecific presentation mimicking more common disorders such as inflammatory demyelinating disease, spinal cord tumor, or degenerative disc disease. The course is slowly progressive, but remission in a stepwise fashion may occur. This patient had a 4-year delay in diagnosis, with symptoms attributed to presumed peripheral neuropathy and transverse myelitis. Unlike patients with demyelinating disease, SDAVF patients are typically male (80%) and in the sixth to seventh decade (mean age, 60 years). Diagnosis is difficult, as SDAVF symptoms overlap with those of other spinal cord disorders: leg weakness (48%), leg paresthesias (35%), back pain (22%), and bladder dysfunction (7%). MRI findings are useful to distinguish SDAVF from demyelinating disease: homogenous, centrally increased T2 signal, enhancement over 6 to 7 lower thoracic or upper lumbar vertebral levels, and dilation of congested coronal plexus veins should raise suspicion for SDAVF. This case highlights the need to consider SDAVF early in the differential diagnosis of progressive myelopathy, as failure results in delayed diagnosis and treatment and accrual of neurologic disability.

Supported by: National Multiple Sclerosis Society

Disclosure: Nothing to disclose