Foix-Alajouanine Syndrome

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Subacute necrotizing myelopathy (SNM) or Foix-Alajouanine syndrome is a rare disease characterized by progressive neurological dysfunction caused by a spinal dural arteriovenous fistula (AVF), without evidence of spinal hematoma.

Historically eponymic, the syndrome now has many other names in the literature, including angiodysgenetic necrotizing myelopathy, subacute necrotizing myelopathy, and venous congestive myelopathy.

Charles Foix and Théophile Alajouanine first described the Foix-Alajouanine Syndrome in 2 young men (aged 29 y and 27 y), in 1926 [1].

A translation and review of the original 42-page French report revealed 2 young men who had presented with progressive and unrelenting myelopathy ultimately leading to their deaths. Pathological analysis demonstrated endomesovasculitis of unknown origin, including vessel wall thickening without evidence of luminal narrowing, obliteration of cord vessels, or thrombosis. Foix and Alajouanine also excluded the presence of intramedullary arteriovenous malformations. At the time, dural arteriovenous fistulas (dAVFs) had not been described, and therefore this type of lesion was not specifically sought. In retrospect, it seems possible that both patients had progressive myelopathy due to Type I dAVFs. In the decades since that original report, numerous authors have included spinal cord venous thrombosis as a central feature of Foix-Alajouanine syndrome. The inclusion of thrombosis in the clinical picture of this syndrome is not only incorrect but may leave one with the impression of therapeutic futility, thus possibly preventing successful surgical or endovascular therapy [2].

Epidemiology

Foix-Alajouanine syndrome is a rare entity. No specific statistics are available with regard to its frequency in the United States, but the condition is likely underdiagnosed.

Predominantly affecting the lower thoracic and/or lumbosacral levels; cervical cord involvement is

rare. Findings include necrosis of the affected cord regions. Grey matter (as compared with white matter) structures are more severely involved.

Incidence & Prevalence

Precise epidemiological data for Foix-Alajouanine Syndrome are lacking due to its rarity. The condition is most commonly associated with spinal dural arteriovenous fistulas (SDAVFs), which have an incidence of 5–10 cases per million per year. SDAVFs account for 70% of spinal vascular malformations, making them the most common type. The prevalence of Foix-Alajouanine Syndrome is likely underestimated due to misdiagnosis or late recognition.

Age & Gender

Typically affects middle-aged to elderly individuals (50–70 years). There is a male predominance, with a male-to-female ratio of approximately 5:1.

Risk Factors

Spinal dural arteriovenous fistulas (SDAVFs): Most cases arise from these abnormal vascular connections. Venous hypertension: Leads to progressive myelopathy due to spinal cord ischemia and necrosis. Delayed diagnosis: Many cases go undiagnosed for months or years due to insidious onset.

Geographic Distribution

No specific geographic or ethnic predilection has been documented. Cases have been reported worldwide, with no clear environmental or genetic factors identified.

Pathology

Masses of enlarged, tortuous, thick-walled subarachnoid veins are observed overlying the surface of the cord (primarily on the posterior aspect). Smaller blood vessels with thickened fibrotic walls also are present within the affected spinal cord segments.

Clinical features

Patients are usually over 50 years of age and can present with acute lower extremity dysesthesias or intermittent sciatica. Progression to paraplegia may be slow.

Foix-Alajouanine Syndrome is a progressive myelopathy that primarily affects the lower thoracic and

lumbar spinal cord, leading to a combination of motor, sensory, and autonomic dysfunction. The condition is associated with spinal dural arteriovenous fistulas (SDAVFs), resulting in chronic spinal venous hypertension, ischemia, and necrosis.

1. Progressive Myelopathy (Insidious Onset) Symptoms develop gradually over months to years, often leading to misdiagnosis (e.g., degenerative spine disease, multiple sclerosis). Typically asymmetric at onset but can become bilateral with disease progression. 2. Motor Symptoms (Weakness & Gait Disturbance) Progressive lower limb weakness: Begins as mild weakness or heaviness in the legs. Evolves into spastic paraparesis or paraplegia in advanced cases. Gait disturbances: Often the first symptom, described as “heavy legs” or fatigue while walking. Worsens over time, progressing to difficulty standing and severe disability. 3. Sensory Dysfunction Numbness, tingling, and paresthesia in the lower limbs. “Stocking-like” sensory loss (distal predominant). Dysesthesia and burning pain in the legs and lower back. Loss of vibration and proprioception, leading to sensory ataxia. 4. Autonomic Dysfunction Bladder dysfunction (early feature): Urinary urgency, frequency, or urinary retention. Bowel dysfunction (later stages): Constipation or fecal incontinence. Erectile dysfunction: Common in affected males. 5. Lhermitte’s Sign Electric shock-like sensations down the spine and into the legs with neck flexion (seen in some cases). 6. Upper Limb Sparing Unlike cervical myelopathies, Foix-Alajouanine Syndrome rarely affects the arms. 7. Symptoms Exacerbation with Exercise & Valsalva Worsening of symptoms with exertion due to increased venous pressure in the spinal cord. Temporary improvement with recumbency (lying down may relieve symptoms).

Diagnosis

The diagnosis of FAS continues to be difficult due to its infrequency and varied clinical manifestations, emphasizing the role of MRI and digital subtraction angiography in diagnosis [3)].

Spinal angiography is needed for definitive diagnosis, based on the clues provided by the symptoms.

Computed tomography (CT) scanning and magnetic resonance imaging (MRI) studies may be normal during the early stages of Foix-Alajouanine syndrome.

With disease progression, T1-weighted MRI scans reveal swelling of the cord and decreased signal intensity peripherally within the affected spinal cord segments. On T2-weighted images, the spinal cord lesions are hyperintense in central locations.

Contrast administration often produces serpentine areas of enhancement and reveals the presence of enlarged, tortuous vessels in the subarachnoid space with associated “flow void” phenomena.

Most spinal dural AV fistulas occur in the thoracolumbar spinal cord; less than 6% are cervical or sacral in location.

Angiography

Initially, MR angiograms can more correctly predict the site and extent of the fistula prior to the use of the more invasive technique of catheter angiography. MR angiograms generally demonstrate flow in serpentine perimedullary vessels.

As previously stated, catheter spinal angiography remains the criterion standard for the diagnosis of

Foix-Alajouanine syndrome and spinal dural AV fistulas. It may demonstrate specific arterial feeders and draining dorsal veins [4].

Radiological diagnosis is usually suspected when there is intramedullary nonspecific enhancement and perimedullary flow voids. Ring-enhancement is rarely reported in the scope of AVF, which poses a diagnostic challenge and raises the suspicion of a spinal cord tumor. In such situations, biopsy can be required and delay proper diagnosis. Salomão et al. report the case of a patient with SNM, who underwent biopsy on the assumption of it being a spinal cord tumor [5].

Treatment

Foix-Alajouanine Syndrome treatment.

Prognosis

This clinical entity has been considered to be the result of progressive vascular thrombosis resulting in a necrotic myelopathy; it has therefore been thought to be largely irreversible and hence untreatable.

The outcome of these patients indicates that acute and subacute progression of myelopathy in cases of spinal dural AV fistulas may be caused by venous congestion and not necessarily by thrombosis. Therefore, a clinical diagnosis of Foix-Alajouanine syndrome is of little practical use, as spinal cord dysfunction from venous congestion is a potentially reversible process whereas thrombotic infarction is not. This diagnosis may result in suboptimal management. The recognition of nonhemorrhagic acute or subacute myelopathy as a complication of a spinal dural AV fistula is important since what appears to be irreversible cord injury is often treatable by standard surgical techniques [6].

Case series

Criscuolo et al., report five patients with dural AV fistulas who presented in this manner, and who improved substantially after embolic and surgical therapy. The outcome of these patients indicates that acute and subacute progression of myelopathy in cases of spinal dural AV fistulas may be caused by venous congestion and not necessarily by thrombosis. Therefore, a clinical diagnosis of Foix-Alajouanine syndrome is of little practical use, as spinal cord dysfunction from venous congestion is a potentially reversible process whereas thrombotic infarction is not. This diagnosis may result in suboptimal management. The recognition of nonhemorrhagic acute or subacute myelopathy as a complication of a spinal dural AV fistula is important since what appears to be irreversible cord injury is often treatable by standard surgical techniques [7].

Systematic review and pooled analysis of case reports

Using the PubMed database, MEDLINE, and EMBASE, Atallah et al. collected data on FAS patients and conducted a pooled analysis. The term 'FAS' was used to search for related articles. Our search was restricted to previous clinical case reports or series that were published in English. Non-English articles were excluded. We included the articles in the period from 1974 to 2024. Articles were eligible if the radiographic and clinical findings were indicative of FAS. A thorough research analysis was performed, examining case reports that specifically addressed this issue. This study examines the clinical symptoms, difficulties in diagnosis, methods of treatment, and outcomes related to FAS.

FAS predominantly impacts the elderly population. A total of 26 patients were diagnosed with FAS. The median age of affected individuals was 53 (SD ±15.96). The ratio of males to females is roughly 5:1. The clinical manifestations encompass gradual muscle weakness and sensory impairments. The diagnosis is dependent on radiological evaluations, specifically MRI and digital subtraction angiography. Possible treatments include endovascular therapy, surgical closure of arteriovenous fistula, or a combination of the two. Significant improvements in neurological impairments can be achieved by early intervention.

The diagnosis of FAS continues to be difficult due to its infrequency and varied clinical manifestations. Prompt and precise diagnosis is essential for proper intervention, typically utilizing endovascular or surgical methods. Additional research is required to determine prognostic markers and enhance long-term care techniques for this rare neurological condition [8)].

The study effectively highlights the importance of early recognition and intervention, emphasizing the role of MRI and digital subtraction angiography in diagnosis, as well as the efficacy of endovascular and surgical treatments.

However, several limitations must be acknowledged. The study relies solely on previously published case reports, which may introduce publication bias and limit the generalizability of its findings. Additionally, the exclusion of non-English studies could lead to an incomplete dataset, potentially overlooking critical insights from diverse populations. The lack of standardized diagnostic criteria and treatment protocols in the reviewed cases further complicates the interpretation of pooled results.

Future research should focus on prospective studies, larger multi-center registries, and long-term follow-up data to enhance our understanding of FAS. Identifying predictive markers for early diagnosis and developing standardized treatment algorithms could significantly improve patient outcomes. Despite its limitations, this study serves as an important reference for clinicians encountering FAS, reinforcing the need for heightened awareness and prompt intervention in suspected cases.

Case reports

2025

The case of a 63-year-old male who developed a rapidly progressing thoracic medullary syndrome over a 6-month period, compromising motor function, sphincter control, and sensory function in the lower extremities. The patient was diagnosed with venous congestive myelopathy secondary to a dural arteriovenous fistula and underwent endovascular embolization using hyper-selective catheterization. Over an 8-month period, the patient experienced successful recovery of both motor and sensory functions. This case supports the use of minimally invasive techniques for the treatment

of dural arteriovenous fistulae with spinal involvement [9].

2024

A 68-year-old man with a history of hypertension presented with a sudden headache, proximal paresis of the left upper extremity, impaired pain and temperature sensation in the right upper extremity, dysphagia, and dysarthria. Computed tomography scans showed intraparenchymal hemorrhage in the left medulla oblongata and a linear, continuous high-density area extending from the medulla oblongata to the cervical spinal cord. Magnetic resonance images showed cervical spondylosis at the C5-6 and C6-7 levels, with high signal intensity changes from the medulla oblongata to the lower cervical cord on T2-weighted images. Cerebral angiography showed no abnormal vessels. Conservative treatment gradually improved symptoms and the high signal intensity areas.

Lessons: This case highlights intracranial hemorrhage occurring from extracranial causes and the possibility of VHM due to cervical spondylosis. When hemorrhagic lesions of the craniovertebral junction or spinal parenchymal lesions are encountered, the underlying pathology should be investigated thoroughly and systematicall [10]

The case of spinal viral vasculitis detected by means of spinal MR-angiography. The undoubted viral etiology of vasculitis allows us to attribute this observation to Foix-Alajouanine syndrome [11]

A 71-year-old male patient presented with sudden loss of strength and hypoesthesia in the lower limbs. A spinal cord magnetic resonance imaging (MRI) scan revealed a vascular tangle in the spinal canal region with hypersignal on T1 in the spinal cord at the T8–T9 level, corresponding with the clinical presentation of the patient. Arteriography was performed to study the patient's anatomy and plan further treatment. Dural arteriovenous fistula (DAVF) was confirmed [12].

2016

A young patient with an unusual clinical onset of Foix-Alajounine Syndrome coincidentally occurring after his Outpatient Clinic appointment, illustrates how prompt surgical treatment can result in rapid recovery of neurological function despite preoperative paraplegia.

Venous hypertension with subsequent rapid resolution after surgical treatment is the pathophysiological mechanism underlying a dural arteriovenous fistula, in contrast to historical views suggesting these lesions result from irreversible venous thrombosis resulting in necrotic myelopathy [13].

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