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CASE REPORT

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Congestive Myelopathy due to Intradural Spinal AVM Supplied by Artery of Adamkiewicz: Case Report with Brief Literature Review and Analysis of the Foix-Alajouanine Syndrome Definition

Authors' Contribution:
A Study Design
B Data Collection
C Statistical Analysis
D Data Interpretation
E Manuscript Preparation
F Literature Search
G Funds Collection

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Summary

Background: Spinal arteriovenous malformations (AVMs) can lead to development of congestive myelopathy (Foix-Alajouanine syndrome). Spinal AVMs are rare and so is this syndrome. Diagnosis is often missed due to its rarity and confusing definitions of the Foix-Alajouanine syndrome.

Case Report: We report a case of a 47-year-old male patient suffering from this rare syndrome with an AVM arising from the artery of Adamkiewicz, which is another rarity. Our patient was treated by embolization of the lesion with 20% glue, after which he showed mild improvement of symptoms. We also present a brief review of literature on spinal AVMs and elucidate the evolution of the term Foix-Alajouanine syndrome.

Conclusions: Use of the term "Foix-Alajouanine syndrome" should be restricted to patients with progressive subacute to chronic neurological symptoms due to congestive myelopathy caused by intradural spinal AVMs. CT angiography should supplement DSA as preliminary Imaging modality. Patients may be treated with surgery or endovascular procedures.

MeSH Keywords: Angiography, Digital Subtraction • Central Nervous System Vascular Malformations • Magnetic Resonance Imaging • Multidetector Computed Tomography • Spinal Cord Ischemia

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Background

Spinal AVMs are uncommon. According to Spetzler (2002), an arteriovenous fistula (AVF) is a spinal AVM without a nidus [1]. An intradural AVF may cause congestive myelopathy leading to subacute or chronic neurological symptoms, the so called Foix-Alajouanine syndrome. On rare occasions this AVF may arise from the artery of Adamkiewicz. Congestive myelopathy is usually reversible if patients are treated early; hence an early diagnosis of this syndrome using clinical and imaging characteristics is of importance.

Case Report

A 47-year-old male presented with gradually progressing bilateral lower limb weakness and urinary incontinence for the past five months. No muscle wasting was noted on examination. Muscles of the calves and the thighs showed bilaterally increased tone with exaggerated ankle and knee

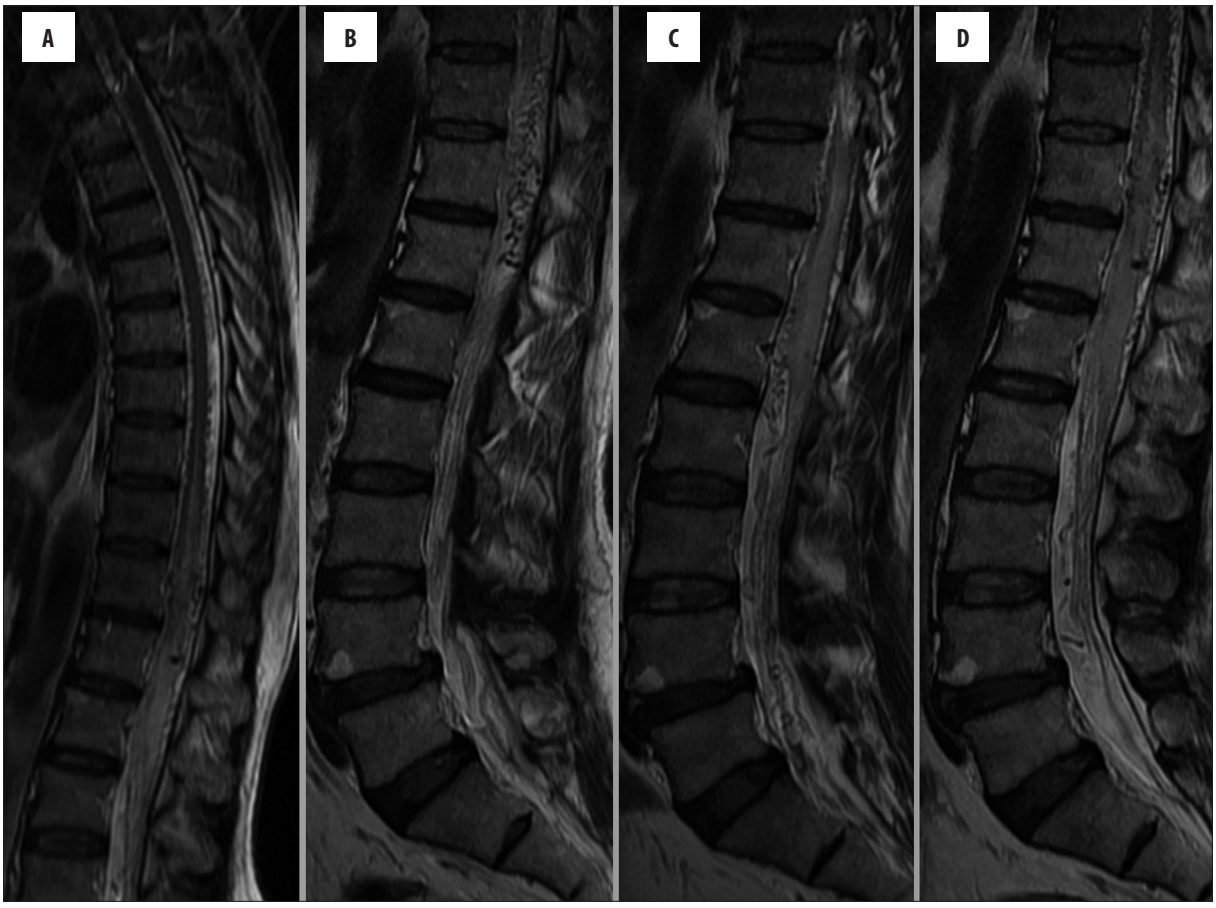

Figure 1. Sagittal T2-weighted (TR=3000 ms, TE=88 ms) images (**A–D**) of the spine demonstrate bulky lower dorsolumbar cord and conus medullaris with increased signal intensity and multiple intradural extramedullary areas of signal void. Grade 1 anterolisthesis of L4 over L5 vertebral body is noted.

jerk reflexes. Muscle power was assessed as grade 3-4 out of 5 according to Medical Research Council (MRC) grading. Patient had paresthesia in both lower limbs in non-dermatomal distribution. Laboratory investigations including a hemogram, renal, liver and thyroid function tests were unremarkable. Serum vitamin B12 levels were normal.

Magnetic resonance imaging (MRI) of the dorsolumbar spine performed with a 1.5T scanner (Signa Excite 1.5T, GE) revealed bulky lower lumbar cord and conus medullaris with high T2 signal. Multiple intradural extramedullary areas signal void were seen from Th6 to S1 level (Figure 1), more abundantly on the left side of Th10–Th12, causing rightward displacement of the lower part of the spinal cord. Spinal cord exhibited intense, relatively homogenous enhancement after intravenous gadolinium injection (Figure 2). Subsequent spinal computed tomography angiography (CTA) was performed with a 16-slice scanner (Philips Brilliance 16, Philips Medical Systems) with non-ionic iodinated contrast agent (Iohexol, 350 mg/mL) at dose of 2 mL/kg injected into the antecubital vein through pressure injector at a rate of 4 mL/sec with bolus tracking, with ROI placed over the abdominal aorta (scanning parameters: increment 1.0 mm, reconstruction interval 0.75 mm, slice thickness 2.0 mm, pitch 1.188, rotation time 0.75s, kVp 120, mA 200) CTA revealed an intradural extramedullary arteriovenous malformation (AVM) from Th10 to Th12 level (Figure 3) supplied by a branch of a great radicular artery (artery of Adamkiewicz). In our case it originated from the abdominal aorta on the left side and ran subcostally, entering the spinal canal through the Th12–L1 intervertebral foramen (Figures 4–6). The AVM drained into the right internal iliac vein through an elongated, tortuous venous channel. Adiagnosis of intradural perimedullary AVM with congestive myelopathy (Foix-Alajouanine syndrome) was made based on the findings.

The patient underwent digital subtraction angiography (DSA) – guided embolization of the AVM with 20-percent cyanoacrylate glue. Immediate postoperative period was uneventful. Postoperatively, over a period of 8 months of regular physiotherapy, the patient showed gradual mild improvement of symptoms, with muscle power of 3 to 4 acc. to MRC grading in both lower limbs. Postoperative MR imaging performed at eight months showed mildly thinned lower cord and conus medullaris with irregular surface and persistent intramedullary signal, which was likely due to gliotic changes in the cord.

Discussion

Definition of Foix-Alajouanine syndrome has been confusing over time. In 1926 Foix and Alajouanine first described two fatal cases of patients with neurological deficits and

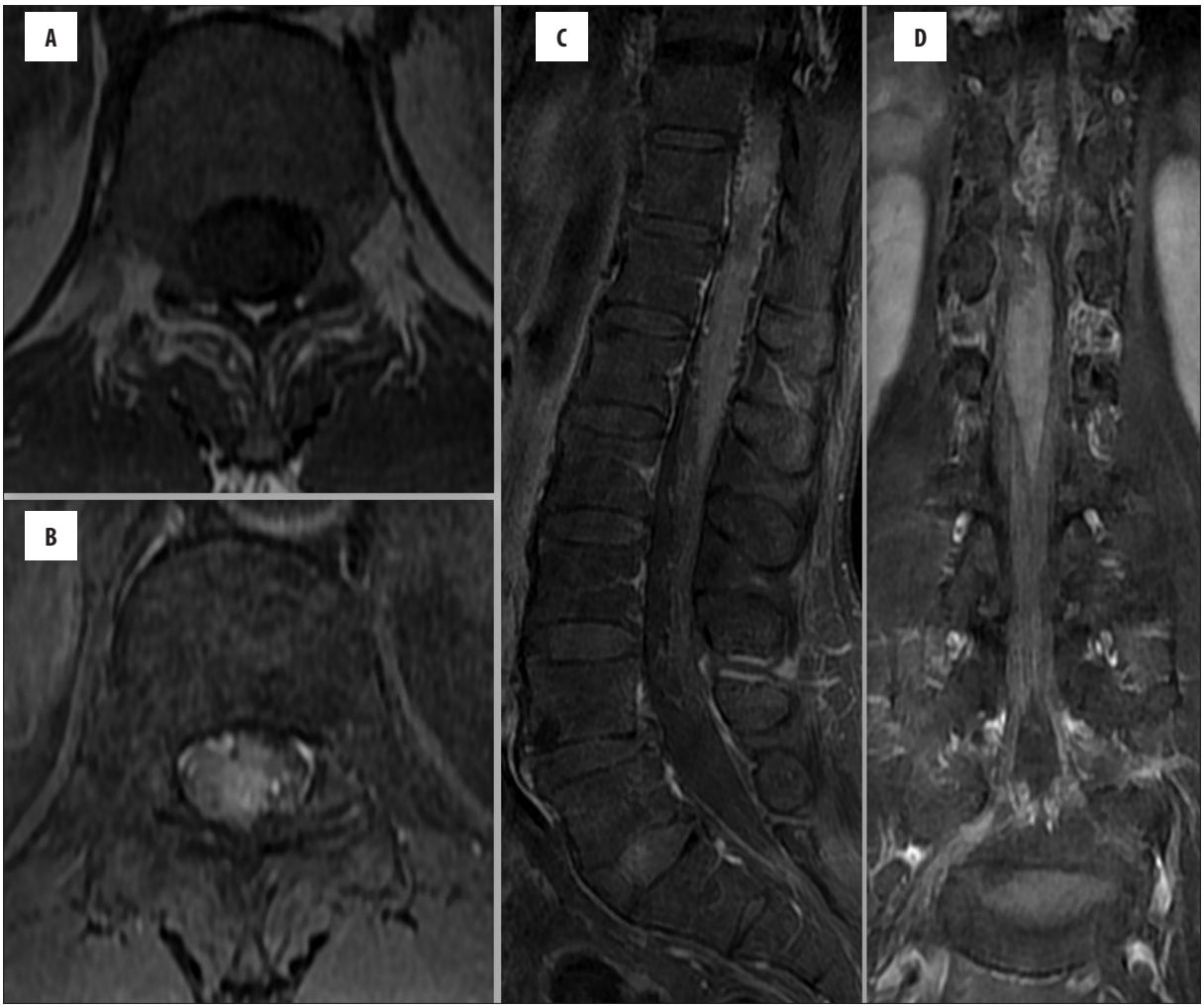

Figure 2. Axial T1-weighted (TR=640 ms, TE=9.4 ms) noncontrast image at lower dorsal level (**A**) and following intravenous gadolinium administration, fat saturated, T1-weighted (TR=640 ms, TE=9.4 ms) axial (**B**), sagittal (**C**) and coronal (**D**) images demonstrate significant post-contrast enhancement of the lower cord and conus medullaris.

tortous vessels on the spinal surface identified post mortem [2]. Later, in 1931 Lhermitte described similar findings and called it necrotizing myelopathy [3]. Over the years, the use of the term Foix-Alajouanine syndrome has been a subject of debates. Initially, this term was associated with spinal artery thrombosis leading to myelopathy, which should be associated with poor prognosis. However, some patients diagnosed with Foix-Alajouanine syndrome improved, making thrombosis of the spinal artery an unlikely cause. Afterward, in 1989 Criscuolo et al. also pointed against its association with thrombosis and explained that the symptoms of this syndrome could be explained by congestive myelopathy, which is a reversible process. Thus, nowadays spinal AVMs with congestive myelopathy without hemorrhage should be referred to as Foix-Alajouanine syndrome. Further narrowing down the terminology, it should be applied specifically to patients with clinically subacute to chronic progressive neurological symptoms due to intradural AVFs leading to congestive myelopathy without hemorrhage [4].

Spinal AVMs are rare entities, but when present they can be disabling. Their classification has changed over time. In the 90s they were classified as: Type I – dural AVFs, Type II – intramedullary glomus AVMs, Type III – juvenile or mixed AVMs, and Type IV – intradural perimedullary AVFs [5]. Later, in 2002 Spetzler and others proposed a new classification for spinal AV malformations (Table 1) [1]. The importance of classification is such that different treatment approach is indicated for each category. An intradural AVF (Type I AVM acc. to Spetzler classification) can cause Foix-Alajouanine syndrome, which may be of dorsal or ventral type. Dorsal type is characterized by an anastomosis between the artery and vein near the dural sleeve and ventral type involves anastomosis between the anterior spinal artery and a vein. In our case the feeding artery was arising from the Artery of Adamkiewicz, which was a branch of the left subcostal artery in this particular case. Such intradural AVFs are rare and not clearly classified. These intradural AVFs may cause congestive myelopathy. In 1974 Aminoff et al. demonstrated that intramedullary venous pressure is increased in case of AVMs, which raises the arteriovenous gradient leading to decreased perfusion of the cord. Congestive myelopathy has predilection for the lower part of the cord due to the effect of gravity and secondly, due to paucity of collaterals in the lower part of the spinal cord [6].

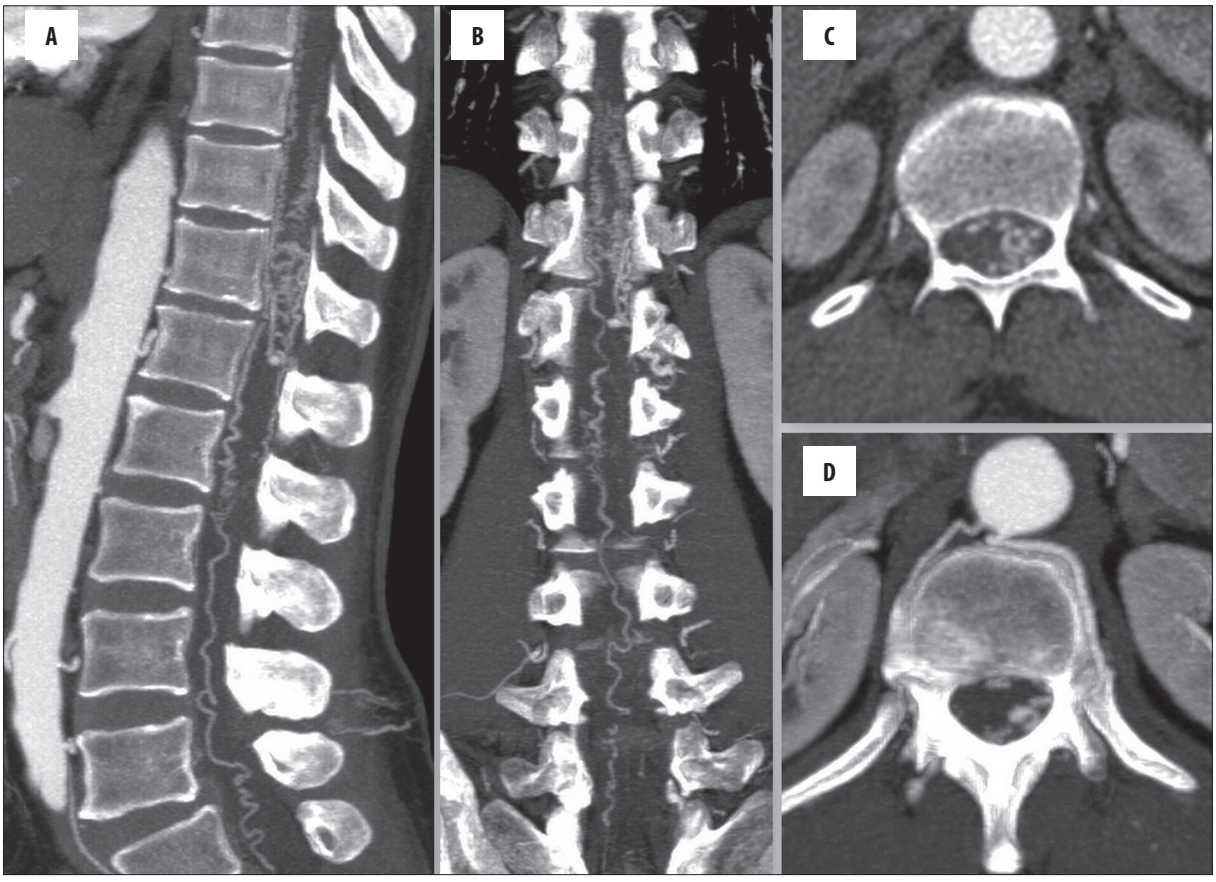

Figure 3. Sagittal (**A**), coronal (**B**) and axial (**C, D**) spinal computed tomography angiography images, maximum-intensity projection showing intradural AVM with cord displacement.

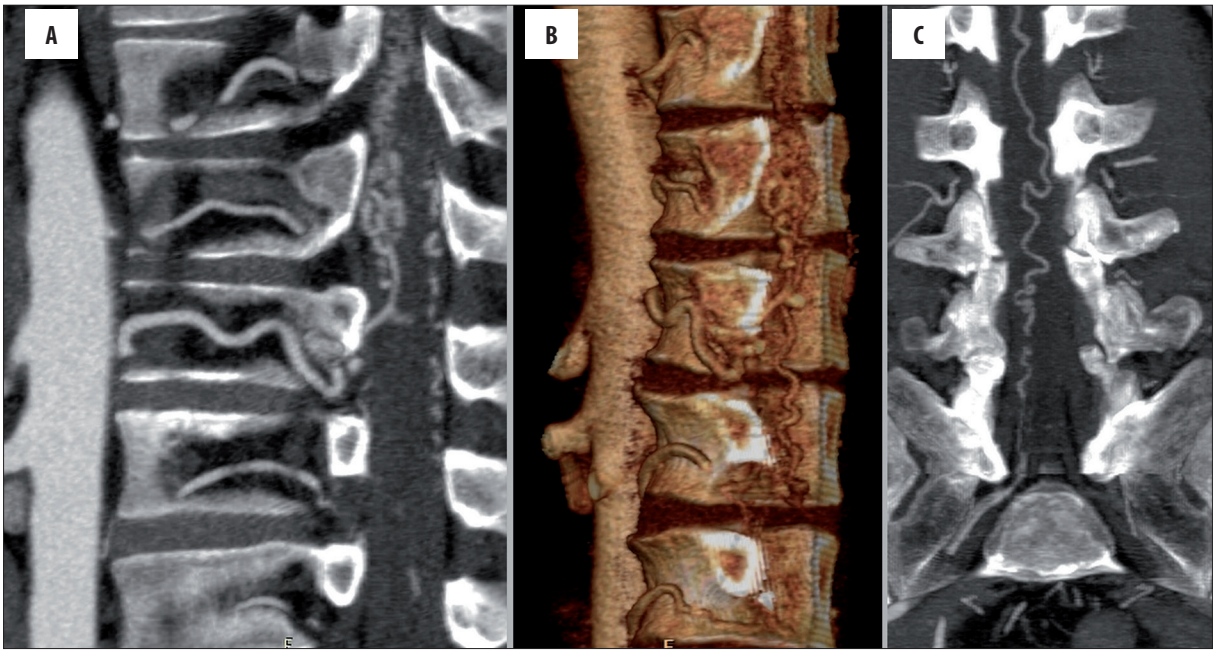

Figure 4. Sagittal curved planar reformat of (**A**) and volume rendered image of CTA (**B**) demonstrate the artery of Adamkiewicz feeding the AVM. Coronal curved reformat (**C**) showing drainage of the AVM through right 1st sacral foramen via an elongated tortuous vein.

Spinal AVMS may present with sudden subarachnoid hemorrhage, i.e. coup de poignard of Michon or gradually progressing myelopathy, i.e. Foix-Alajouanine syndrome. Symptoms such as lower limb paresis, difficulty walking, sphincter dysfunction and even sensory loss may be present. Usual duration of symptoms may vary from months

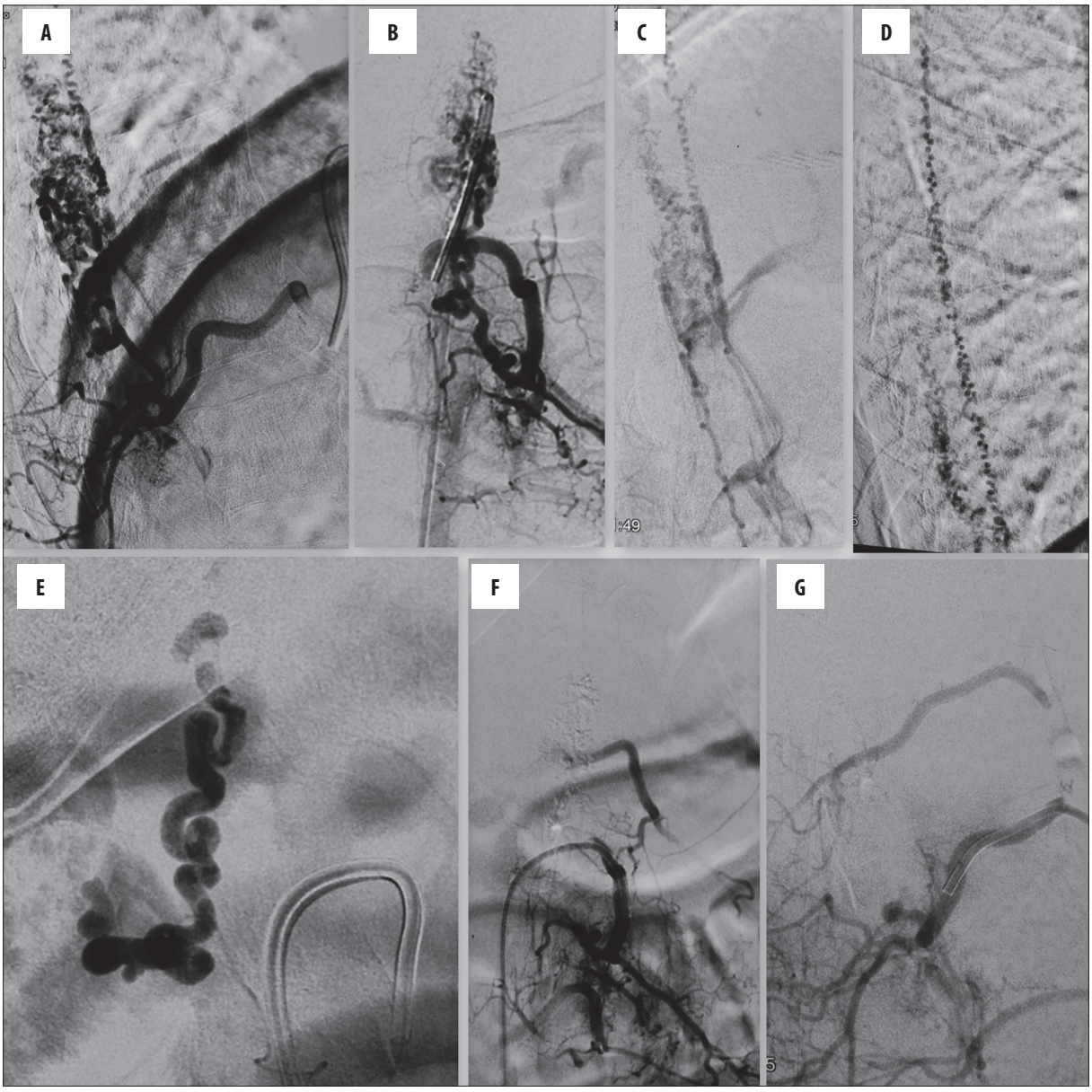

Figure 5. Digital subtraction angiography was performed by catheterizing the feeding artery. Lateral (**A**) and right anterior oblique (**B**) view in arterial phase show multiple dilated vessels within the spinal canal. Lateral views in the venous phase (**C, D**) show tortuous venous channels in the spinal canal. The lesion was embolized with 20% glue mixed with lipiodol (**E**). Post-procedure antero-posterior (**F**) and lateral (**G**) images show no opacification of the AVM.

to years. The clinico-radiological diagnosis of this syndrome is difficult due to its rarity as well as its somewhat unclear definition. Clinically, it can be confused with intramedullary tumor or polyneuropathy. The diagnosis of spinal AVMs is primarily based on imaging and useful modalities include digital subtraction angiography (DSA), spinal computed tomography angiography (CTA), and spinal magnetic resonance angiography (MRA). DSA is the modality of choice, as it visualizes smaller and finer vessels in their greatest detail. However, it is associated with high radiation dose and inability to demonstrate mural and extravascular details. CTA can provide adequate details but is not as conclusive as DSA. According to Gio Si-jia et al. (2009), CT angiography should be used for screening before spinal DSA to guide the procedure. One should look for the feeding vessel and location of the nidus, if present, which helps in classifying the AVM type. MR imaging is useful for assessing the status of the spinal cord, which can demonstrate hemorrhage or congestive changes associated with AVM in addition to multiple areas of flow void within the lesion [7].

Congestive myelopathy should be suspected in a patient with chronic neurological complaints and T2 enhancement in the lower part of the spinal cord and a follow-up examination with spinal CTA or DSA should be planned to look for spinal AVM, which can be treated surgically or with endovascular techniques, the former being the preferred approach. Post-treatment imaging is necessary to check for surgical complications and to monitor recovery

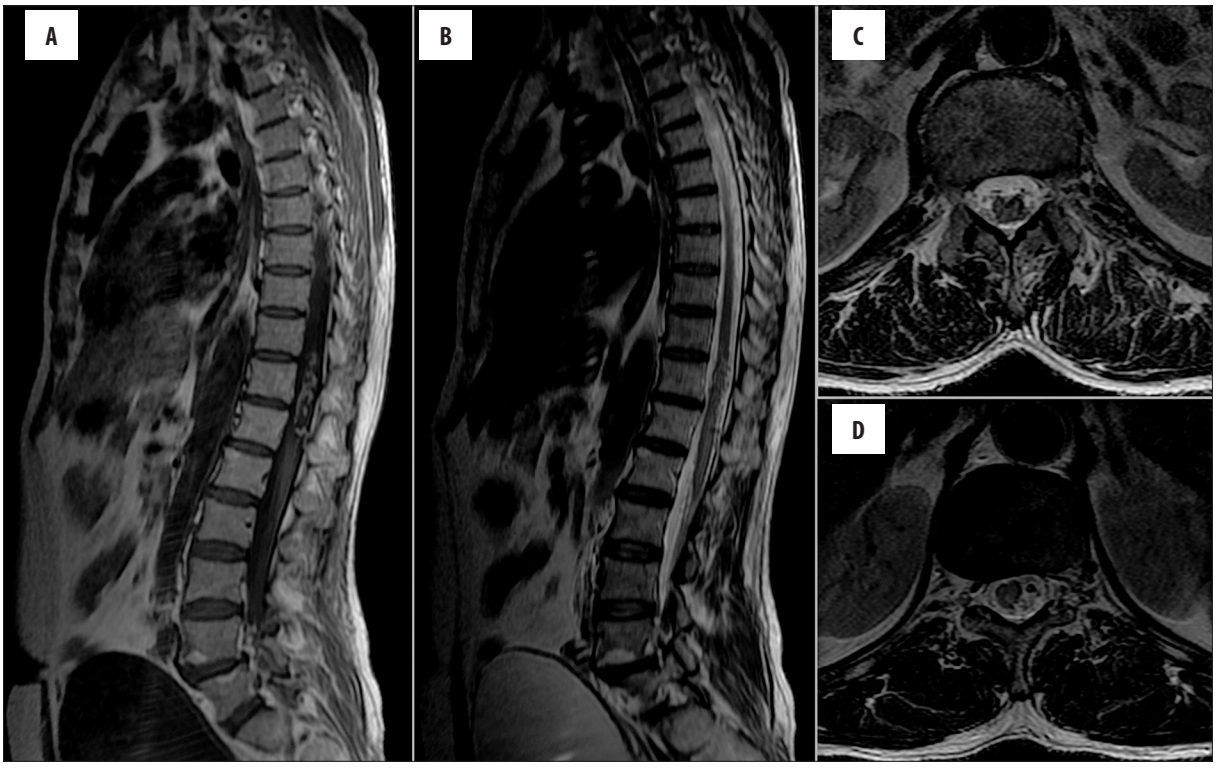

Figure 6. MR performed eight months after the procedure. Sagittal T1-weighted (**A**) and T2-weighted (**B**), as well as axial T2-weighted (**C, D**) images show thrombosed AVM (arrow in **A**) with irregular outline of the lower cord and conus, reduced bulk and altered signal suggesting gliotic changes.

Table 1. Spetzler classification of spinal AVFs (2002) [8].

Extradural
Intradural ventral Shunt between the anterior spinal artery and a draining vein Also known as Type IV lesions, intradural perimedullary AVFs (usually high-flow) Can be small, medium or large
Intradural dorsal Shunt between the artery and vein at the level of a dural root sleeve also known as spinal dorsal arteriovenous fistula or SDAVF (usually low-flow) Can have single or multiple feeders

process. There are suggestions of postsurgical improvement in patients with spinal AVMs arising from the anterior spinal artery [8]. In our case, where the AVM arose from the artery of Adamkiewicz, mild improvement of symptoms was seen after DSA-guided glue embolization of the AVM.

Conclusions

The term Foix-Alajouanine syndrome should be limited to patients with progressive subacute to chronic neurological symptoms due to congestive myelopathy caused by intradural spinal AVMs. CT angiography should complement DSA as a preliminary imaging modality. Patients may be treated surgically or with endovascular procedures.

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