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Case report

Vertebrojugular fistula mimicking an intradural schwannoma

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1. Introduction

Arteriovenous fistulas (AVFs) are relatively uncommon lesions of the spine. Despite their low overall incidence, AVFs are the most common vascular anomaly of the spinal cord [1]. AVFs between the vertebral artery (VA) and the jugular vein (JV) are a rare subgroup of spinal AVFs. Most are post-traumatic in origin, following direct injury or iatrogenic manipulations. Spontaneous fistulas are often associated with neurofibromatosis or fibromuscular dysplasia [2].

In this article, we present a unique case of a spontaneous vertebrojugular fistula that manifested as a diffusely enhancing lesion on the cervical spine, which mimicked an intradural extramedullary mass lesion such as a schwannoma.

2. Case report

A 43-year-old woman was admitted to our hospital with a 6-month history of cervical pain radiating to the left arm. The physical and neurological examinations were unremarkable, and her medical history was negative for trauma, surgery or endovascular procedures. No cutaneous stigmata for neurofibromatosis were noticed.

Magnetic resonance imaging (MRI) of the cervical spine revealed an intradural, extramedullary, diffuse contrast-enhancing tumor-like lesion extending from the C2 to the C6 vertebrae (Fig. 1A and B). The preoperative diagnosis at the time of admission was that of a schwannoma.

2.1. Surgical approach

A standard posterior midline approach was used to expose the posterior aspects of C1–C6. A C2–C5 total laminectomy was performed. A tortuous, well-demarcated, purple-colored lesion extending through vertebrae C2–C5 was observed. The spinal cord was compressed to the right lateral side of the mass lesion. An attempt to incise the biopsy caused a massive hemorrhage. Due to the suspicion of a vascular lesion, the procedure was terminated as soon as hemostasis was achieved. The postoperative period was uneventful.

On the second postoperative day, digital subtraction angiography (DSA) was performed and showed a communication between the extracranial VA and the internal jugular vein (IJV) (Fig. 2a).

2.2. Endovascular approach

Angiographic images of the bilateral VA were obtained using a right transfemoral artery approach. Angiograms revealed an arteriovenous fistula between the left VA V2 segment and the left IJV (Fig. 2a). The left VA V3 segment and the distal intracranial segment could not be shown in angiograms because of the high flow rate of the fistula (Fig. 2b). The right VA, the basilar artery and both anterior and posterior inferior cerebellar arteries were normal. Selective right vertebral angiography images indicated that the distal portions of the left VA and the left posterior inferior cerebellar artery were patent. These arteries filled with retrograde flow. After the diagnostic angiograms, a 6 F 90 cm guiding catheter (Chaperon,

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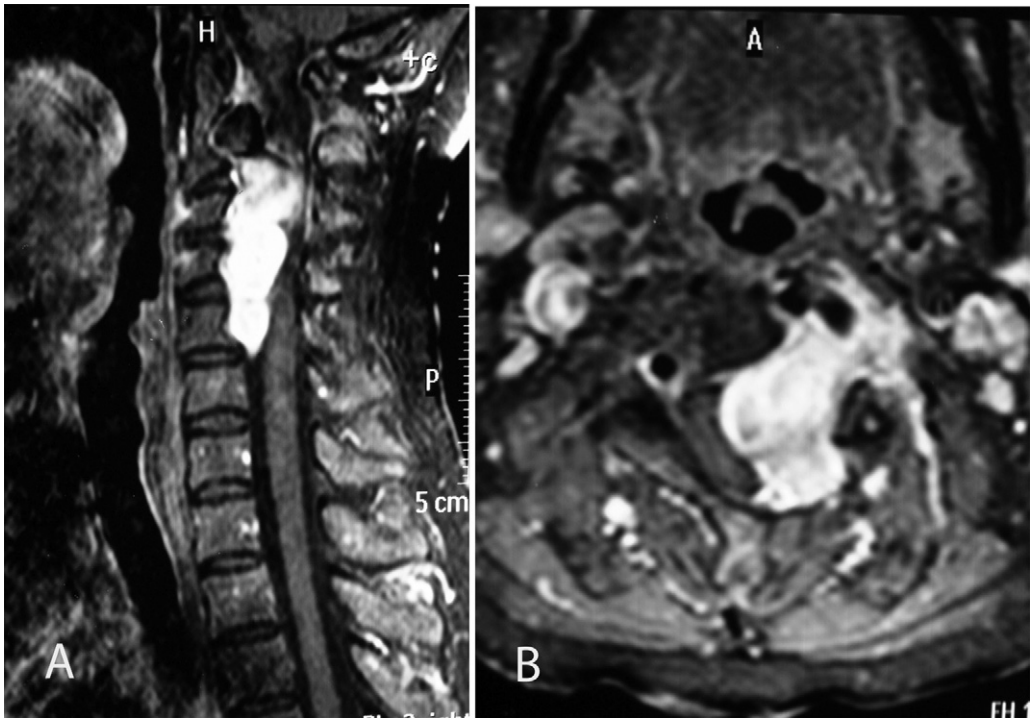

Fig. 1. Magnetic resonance imaging of the cervical spine revealed an intradural, extramedullary, diffusely enhancing mass extending from vertebrae C2 to C6. (A) Preoperative sagittal T1-weighted magnetic resonance imaging demonstrating an extensive intradural upper cervical mass. (B) An intradural mass suggestive of a schwannoma in the upper cervical cord demonstrating compression of the cord laterally. The mass extends into the left neural foramen in the axial T1-weighted image.

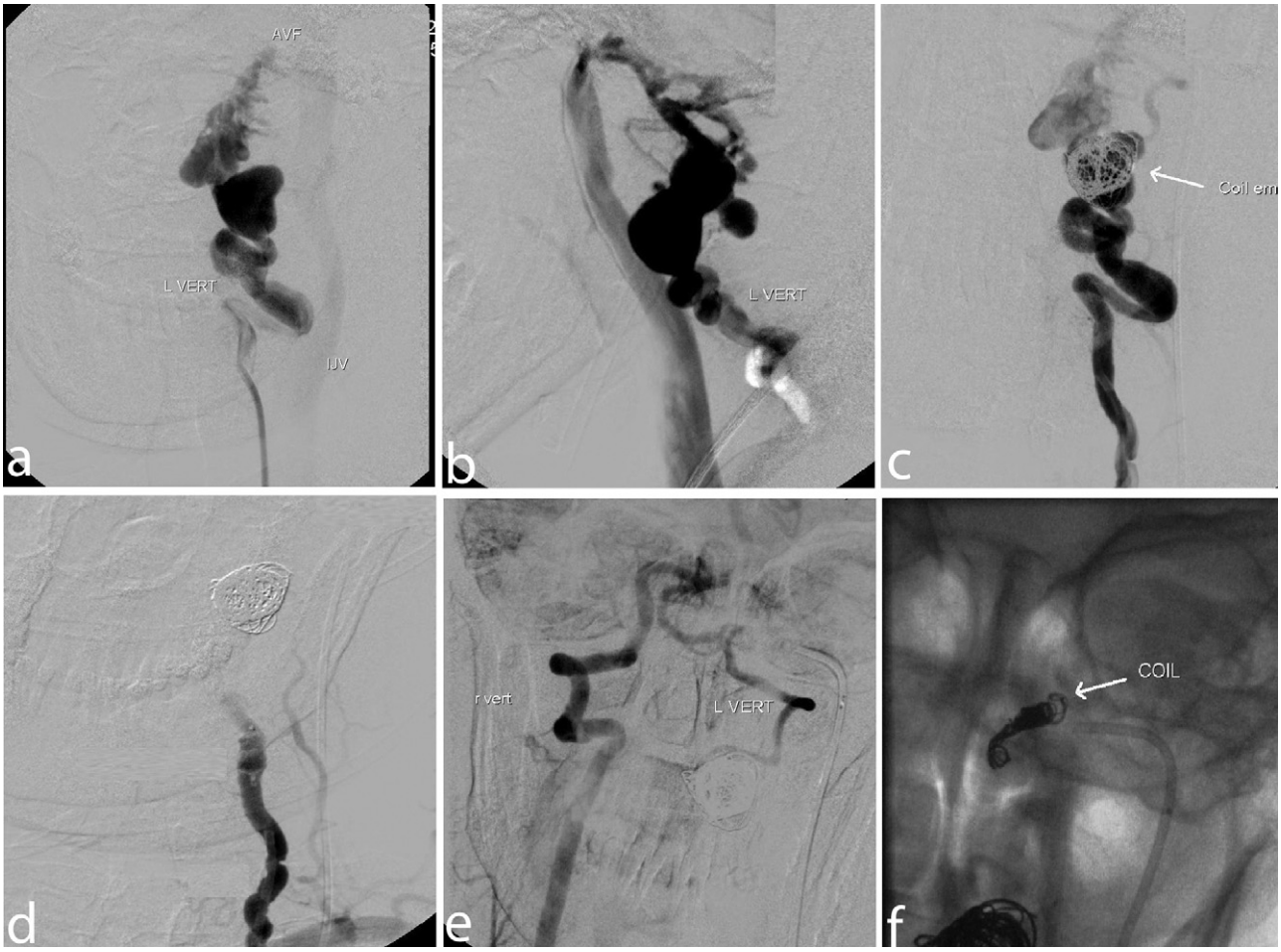

Fig. 2. Digital subtraction angiography revealed a fistula between the left vertebral artery and the internal jugular vein (a). The left V3 segment cannot be shown due to the high flow rate of the fistula (b). Multiple detachable microcoils were first used in an attempt to occlude the fistula (c). The parent artery (left vertebral artery) was successfully occluded (d). Angiograms showing the retrograde filling of the distal portion of the left vertebral artery (e). The venous side of the fistula (left internal jugular vein) was also occluded (f).

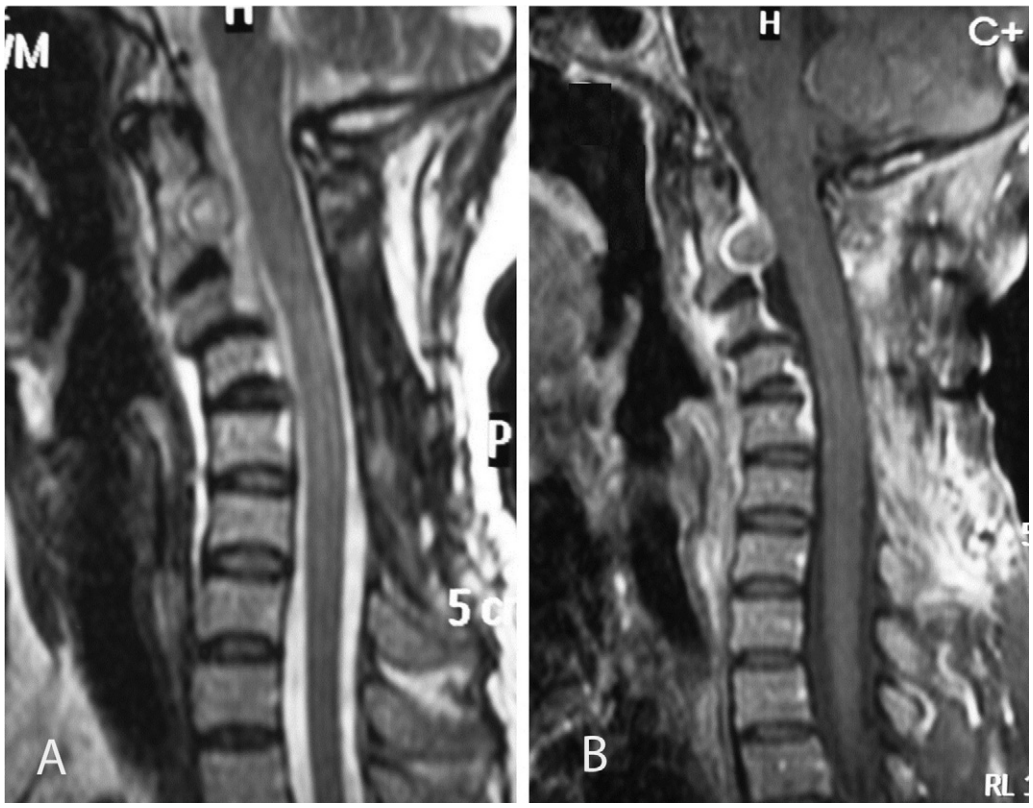

Fig. 3. The postoperative sagittal T2-weighted (A) and T1-weighted (B) images with gadolinium demonstrate rapid resolution of the upper cervical mass within two months and minimal C3–4 dislocation.

MicroVention, Terumo) was placed proximal to the cervical portion of the left VA, and an angled-tip 1.9 F 150 cm microcatheter (Echelon 14, ev3, Plymouth, USA) was placed just proximal to the AVF. We were unable to reach the exact fistula level. We first attempted to embolize the fistula with multiple detachable microcoils (Axium, ev3, Plymouth, USA) (Fig. 2c). Coil embolization failed due to the high flow rate of the fistula. We decided to occlude the parent artery. An 8 mm vascular occlusion device (Amplatzer Vascular Plug 2, AGA Medical Corp., Golden Valley, MN, USA) was deployed in the left VA. The left VA was successfully occluded (Fig. 2d). Selective right VA angiograms were obtained and showed retrograde filling of the distal part of the left VA (Fig. 2e). We therefore decided to embolize the venous side of the fistula. Using a right transfemoral venous approach, a 6 F 90 cm guiding catheter (Chaperon, MicroVention, Terumo) was advanced into the left IJV. A microcatheter (echelon 14, ev3, Plymouth, USA) was placed on the venous side of the fistula, and multiple detachable microcoils (Axium, ev3, Plymouth, USA) were then deployed (Fig. 2f). After the embolization procedure, the AVF was completely occluded.

Two months after the procedure, the patient fell from a height, and a C3–4 dislocation occurred. Posterior cervical stabilization was recommended, but the patient rejected the treatment (Fig. 3A and B). One year after the procedure, the patient was symptomless, and an MRI of the cervical spine revealed no mass lesion.

3. Discussion

The incidence of intradural extramedullary tumors is 0.3 out of 100,000 people, and 22% of these lesions are located on the cervical spine. Histopathological diagnoses include schwannoma in 9.6–35% of total cases, meningioma in 9.6–35%, neurofibroma in 4–23% and metastatic tumors in 6.4–25% [3]. Vascular anomalies, such as AVFs, intradural hematomas and arteriovenous malformations, may infrequently occur as intradural extramedullary anomalies.

Vertebral AVFs are uncommon, consisting of an abnormal communication between the extracranial VA or its branches and the deep venous plexus. Most vertebral AVFs are posttraumatic or iatrogenic in origin [4]. Spontaneous fistulas have been associated with diseases such as neurofibromatosis and fibromuscular dysplasia [2]. More specifically, occurring fistulas between the VA and the IJV, as in our patient are extremely rare, and are almost always secondary to a penetrating trauma [5]. To our knowledge, the presented case is the very first case of a vertebrojugular fistula mimicking an intradural extramedullary spinal tumor without trauma and congenital disease.

The presentation of patients with AVFs may be different than those with spinal tumors both clinically and radiologically. The typical clinical findings of AVFs include cervical bruits, tinnitus and vertigo. Vertebrobasilar insufficiency, radicular pain and spinal cord symptoms are infrequent [1]. Cervical radicular pain is directly related to nerve root compression by dilated veins. Spinal cord symptoms are primarily due to venous hypertension with consequently impaired drainage of the spinal vein leading to myelopathy; pulsatile dilated veins may also generate direct mechanical compression of the spinal cord. In the presented case, the patient suffered from left radicular pain in the arm, which is not very typical for an AVF.

The initial MRI findings for our patient were consistent with an intradural extramedullary lesion, and a meningioma or a schwannoma was suspected. A meningioma that is intradural and extramedullary and grows slowly into the subarachnoid space is identified as a homogenous low-signal intensity mass on T1-weighted images (T1WIs) and as a homogenous high-signal intensity mass on T2-weighted images (T2WIs). Using gadolinium administration, meningiomas appear homogenously and diffusely

enhanced. Using MRI, a schwannoma is observed as iso- or hypointense on T1WIs and hyperintense on T2WIs and are heterogeneously or homogenously enhanced with gadolinium [3].

Because clinical and radiological evaluations diffuse contrast-enhancing lesions mimicking a schwannoma, we preferred surgery. However, surgery should be considered for fistulas unsuitable for treatment by endovascular means or in cases of endovascular treatment failure. As in our case, coil embolization is considered to be an effective treatment for the endovascular occlusion of such fistulas [4].

Thus, AVFs are almost always associated with trauma or iatrogenic manipulations, whereas spinal AVFs should be considered in the differential diagnosis of a spinal intradural mass. In such unusual cases, an angiography should be considered even in the modern era of advanced imaging tools to avoid unexpected complications and the performance of an unnecessary surgical approach.

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