

Intracranial Dural Arteriovenous Fistula with Perimedullary Venous Drainage: A Case Report

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Abstract

Case Report

Intracranial dural arteriovenous fistulas with perimedullary venous drainage are indeed very rare cerebral vascular malformations. They can present with symptoms such as rapidly progressive ascending myelopathy associated with dysautonomia, complicating the diagnostic process and delaying therapeutic intervention. We present the case of a 26-year-old patient admitted to the neurosurgical department for an evolving medullary syndrome caused by an intracranial dural fistula with perimedullary venous drainage. The diagnosis was suspected on a cerebro-medullary MRI, which visualized an arteriovenous shunt at the level of the left superior petrosal sinus and at the level of the ipsilateral internal cerebral vein. The MRI also showed signs of edematous distress of the pontine and tiered medullary regions.

Keywords: Dural arteriovenous fistula, Dura mater, Spinal cord edema, Cerebral hemorrhage, Tinnitus, Embolization, Endovascular treatment."

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INTRODUCTION

A dural arteriovenous fistula (DAVF) is an acquired abnormal arteriovenous connection within the layers of the dura mater, with a wide range of clinical presentations and natural history.

CASE PRESENTATION

A 26-year-old patient presents with heaviness in the lower limbs and progressively worsening sphincter disturbances for the past 6 months. A brain and spinal MRI was performed.

DISCUSSION

Physiopathology

While the trigger for the formation of arteriovenous communications in the dura mater remains unknown, the pathophysiology of the consequences of these arteriovenous communications is now well understood. When the venous drainage of a dural arteriovenous fistula (DAVF) only occurs through the lumen of a sinus, there is no risk of parenchymal complication, as the dura mater wall can withstand arterial flow. However, when the venous drainage of the DAVF involves a leptomeningeal vein, there is a risk of

hemorrhage or parenchymal edema in the territory of this vein. This critical rule applies regardless of the DAVF's location. The arterialization of a leptomeningeal vein, whose wall is much more fragile than that of the dura mater, can lead to its rupture, resulting in intradural hemorrhage. This hemorrhage can occur in any location: parenchymal, subdural, or subarachnoid.

Clinical manifestations of DAVFs

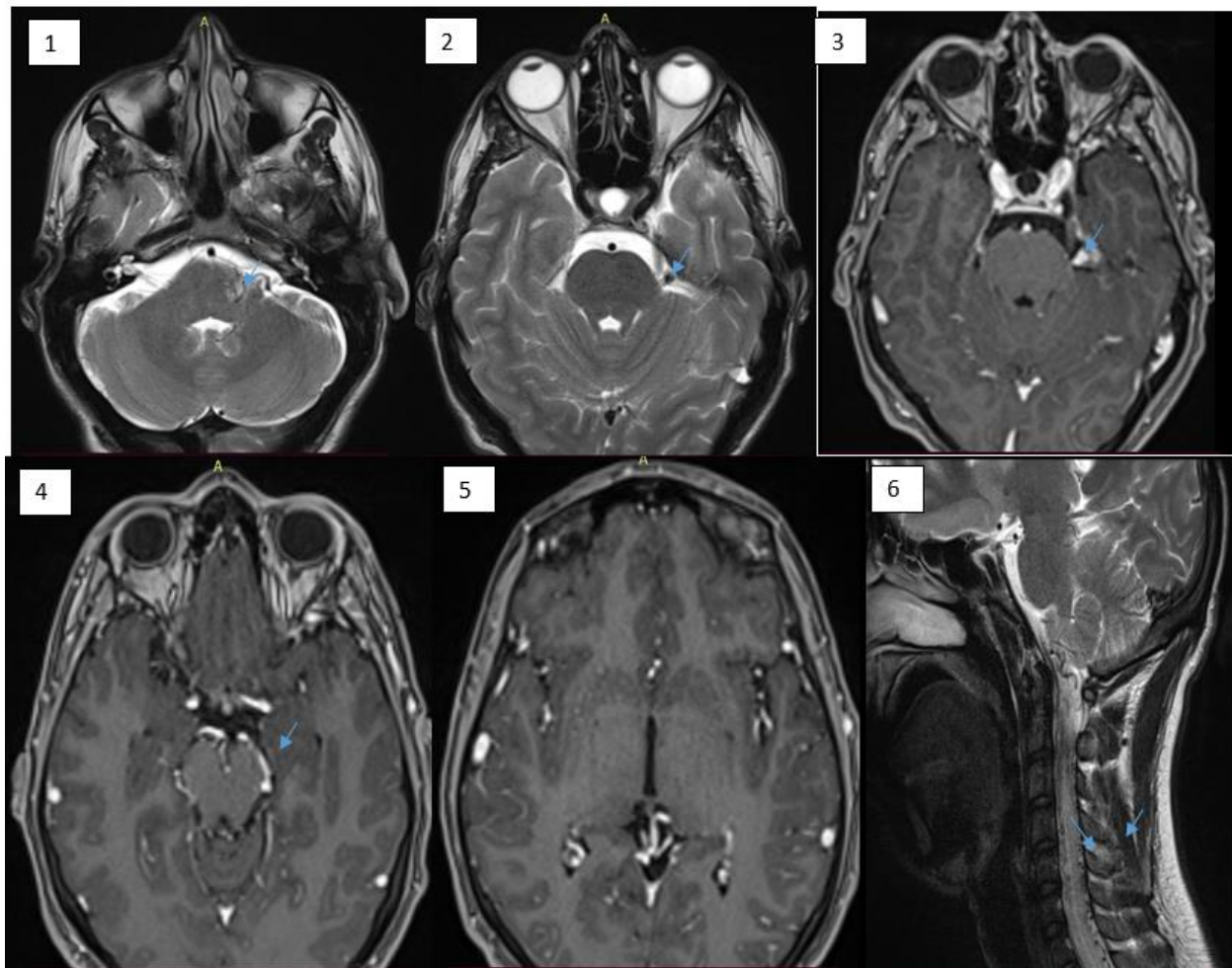
It should be considered in the presence of a clinical history of neurological worsening over several days, with gait disturbances, sphincter dysfunction, and sensory deficits. These clinical signs of myelopathy progress in a caudocranial manner: affecting the lower limbs in all cases, followed by sphincters, upper limbs, and possibly cranial nerves.

Radiological diagnosis of DAVFs

Conventional cerebral angiography is, of course, the reference examination in the radiological diagnosis of dural arteriovenous fistulas (DAVFs). It is systematically performed in the assessment of a DAVF with two objectives: to evaluate the type of venous drainage of the DAVF and to define the strategy for possible treatment. However, it is a specialized

examination that is only undertaken as a first-line procedure in cases of cerebro-meningeal hemorrhage. Outside of this situation, the essential non-invasive

examination is the MRI in three-dimensional (3D) time-of-flight (TOF) sequence.



The MRI shows an enlarged spinal cord with a slightly hyperintense central signal on T2, suggesting spinal cord edema (figure 6). Abnormally enlarged vessels are identified at the left cerebellopontine angle (figure 1) and along the cervical spinal cord (Figure 6). These vessels correspond to vascular structures, specifically the anteroinferior cerebellar artery communicating with the dilated left Dandy vein, which drains into the left superior petrosal sinus and the ipsilateral internal cerebral veins (Figure 3 et 4). This indicates spinal cord edema complicating a dural fistula located in the left cerebellopontine angle region.

The Cognard *et al.*, and Merland classification distinguishes five types according to the mode of venous drainage. In our case, the posterior fossa dural fistula exhibited cervical perimedullary venous drainage, leading to clinical manifestations associated with cervical spinal cord involvement.

Imaging will reveal myelopathy initially corresponding to edema, which is first reversible and easily identifiable on T2-weighted sequences (spinal cord edema due to venous congestion), and then to venous infarction. The draining veins enhance after gadolinium injection, with possible enhancement within the spinal cord due to venous stasis, which should not be confused with a tumor lesion. TOF MR angiography or gadolinium injection allows visualization of the

arteriovenous shunt located at the left cerebellopontine angle in our case.

Treatment of DAVFs

The treatment of dural arteriovenous fistulas (DAVFs) is primarily endovascular. There have been two major developments in the endovascular treatment of DAVFs: the use of venous access with coils initiated in the 1980s, and more recently, the use of arterial access with liquid embolic agents.

CONCLUSION

Intracranial dural arteriovenous fistulas (DAVFs) are arteriovenous lesions whose frequency is underestimated. Their diagnosis still requires the use of conventional cerebral arteriography. Their treatment is

endovascular in the vast majority of cases and should be handled by a specialized interventional neuroradiology center.

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