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Case reports

Dural Arteriovenous Fistulas - Regarding Two Clinical Cases

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Abstract

Dural arteriovenous fistulas (AVF) are rare intracranial vascular malformations, consisting of communications between dural venous arteries and sinuses, meningeal veins, cortical veins, or combinations thereof. In most cases the etiology is unknown. The clinical presentation is determined by the location and mainly by the drainage pattern. The recognition of this rare clinical entity is essential for an early diagnosis and intervention, to avoid the potentially serious complications of the disease, such as, intracranial hypertension, vascular dementia, hemorrhage, among others.

Keywords: ***Dural arteriovenous fistulas; vascular malformations; pulsatile tinnitus; headache; cranial murmur***

Introduction

Dural arteriovenous fistulas (DAVFs) are rare intracranial vascular malformations, consisting of communications between arteries and dural venous sinuses, meningeal veins, cortical veins, or combinations thereof.

They are responsible for 10-15% of intracranial arteriovenous malformations [1,2]. In most cases, the etiology is unknown [3-5]. Dural AVFs most often affect patients between 50 and 60 years old and there is no gender predilection or genetic component for these lesions [5].

DAVFs may present incidentally or with symptoms related to the location of the fistula and the venous drainage pattern [1,2,5-10]. The venous drainage pattern will also determinate the hemodynamic status of the arteriovenous shunt, which can be translated into venous distention, retrograde flow, ischemic and/or hemorrhagic changes [3,8,11,12]. Although there are many classification schemes to stratify the risk of intracranial hemorrhage (ICH) and nonhemorrhagic neurological deficits (NHND), the presence of retrograde cortical venous drainage (CVD) is an alarming sign that foreshadows a high risk of neurological sequelae [8-10,12].

The recognition of this rare clinical entity is essential for an early diagnosis and intervention, to avoid the potentially serious complications of the disease, such as, intracranial hypertension, vascular dementia, and hemorrhagic manifestations.

The aim of our work is to report two cases of DAVFs, demonstrating their clinical and pathophysiological heterogeneity, which is often not recognized in a timely manner in clinical practice.

Case Report 1

57-year-old woman with a history of type 2 diabetes was diagnosed with bilateral papilledema during screening for diabetic retinopathy. In this context, she was sent to a neurology consultation, where she reported pulsating tinnitus and recurrent intense holocranial migraine-like headache, which worsened with recumbency, with years of evolution. On physical examination, she had a bilateral cranial murmur, predominantly right, associated with thrill, without other major changes. She did not report history of previous head trauma or surgery. From the analytical study carried out, no relevant changes were detected. A head computed tomography (CT) was performed, which was normal. For intracranial hypertension screening, a lumbar puncture was performed, with an opening pressure >50 cmH_2O, and the analysis of the cerebrospinal fluid was normal. Subsequently, a magnetic resonance imaging (MRI) of the head was ordered, which revealed changes suggestive of an arteriovenous fistula in the vicinity of the right lateral sinus. The conventional angiographic study confirmed a DAVF fed bilaterally by external carotid artery (**Figure 1**), internal carotid artery (**Figure 2**), and vertebral arteries. Retrograde

flow in the dural sinuses was documented with evidence of a pseudophlebitic pattern, corresponding to the presence of venous hypertension. An endovascular treatment session was carried out, with embolization with an embolic fluid, under concomitant occlusion of the right lateral sinus with a balloon (**Figure 3**). Due to presence of retrograde flow of the embolic fluid through the collateral branch of the right middle meningeal artery, embolization of this arterial pedicle was interrupted, with partial resolution of the fistulous component to the lateral sinus. In the post-procedure control, a fistulous component persisted to the right lateral sinus, fed by branches of the right external carotid artery and right vertebral artery. The patient showed symptomatic improvement, with a decrease in intracranial pressure, confirmed by lumbar puncture (post procedure opening pressure of 35 cmH_2O).

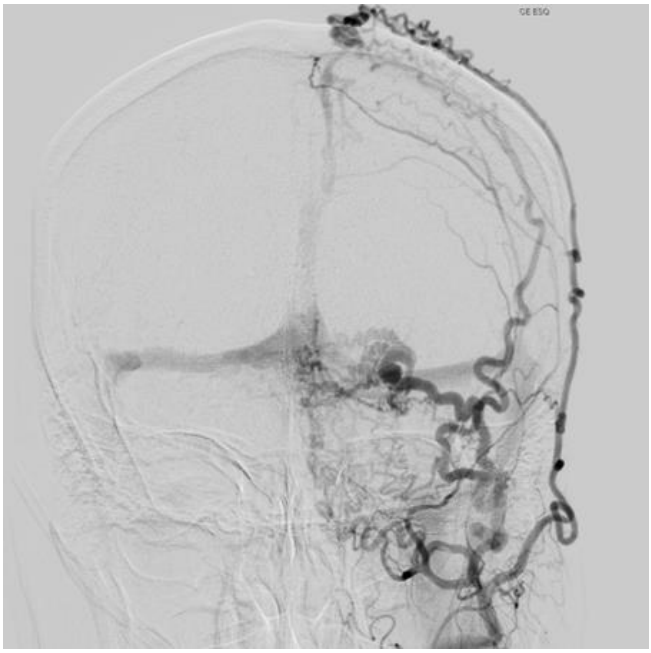

Figure 1: DAVF of the superior sagittal sinus and the torcula fed by meningeal branches of the left external carotid artery.

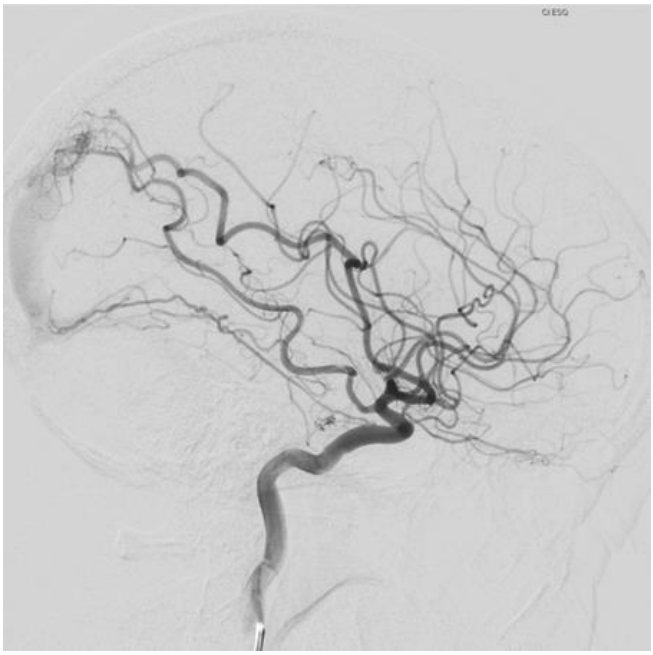

Figure 2: DAVF of the superior sagittal sinus and torcula fed by pial branches of the left internal carotid artery.

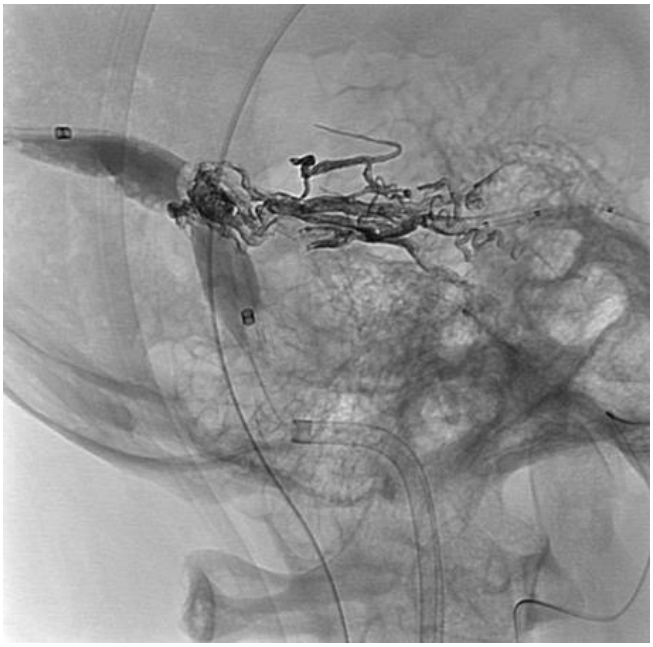

Figure 3: Image of the endovascular treatment of the DAVF of the right lateral sinus, with simultaneous arterial and venous approach: balloon inflated in the right lateral sinus (A) and microcatheter in the right middle meningeal artery (C), through which the embolic fluid (B) is administered to the fistula.

Case Report 2

72-year-old man with a history of arterial hypertension, hypocoagulated due to atrial fibrillation, referred to neurology consultation for orthostatic dizziness. During the consultation, he also reported mild episodic occipital headache, reaching the posterior cervical region, without accompanying alarm signs, with years of evolution. He did not report history of previous head trauma or surgery. On physical examination he had audible bilateral cranial murmur and the neurological exam was normal. From the analytical study carried out, no relevant changes were detected. In order to exclude secondary causes for the symptoms, the patient underwent a head CT scan that revealed a chronic subdural hematoma with left frontoparietal location, without other major findings. The CT angiography reveled probable aneurysmal formation in the insular part (M2 segment) of the left middle cerebral artery, with irregularity in the left internal carotid. Given the suspicion of cerebral arteriovenous malformation, conventional angiography was performed, which excluded this diagnostic hypothesis, and revealed DAVF of the left lateral sinus, fed by external carotid artery (**Figure 4**), internal carotid artery and vertebral artery, with dural and cortical retrograde venous drainage. Venous hypertension with a pseudophlebitic drainage pattern has also been demonstrated (**Figure 5**). After the study, it was concluded that the patient had tension headache and peripheral vertigo, and that the symptoms first referred were not in relation with the FAVD. In view of the spare symptoms and of the complexity of the DAVF which has high risk of unsuccess and complications, and despite the presence of chronic subdural hematoma that may correspond to a previous bleeding from the fistula, it was decided, in a multidisciplinary consultation, not to perform treatment and the patient is currently under careful observation.

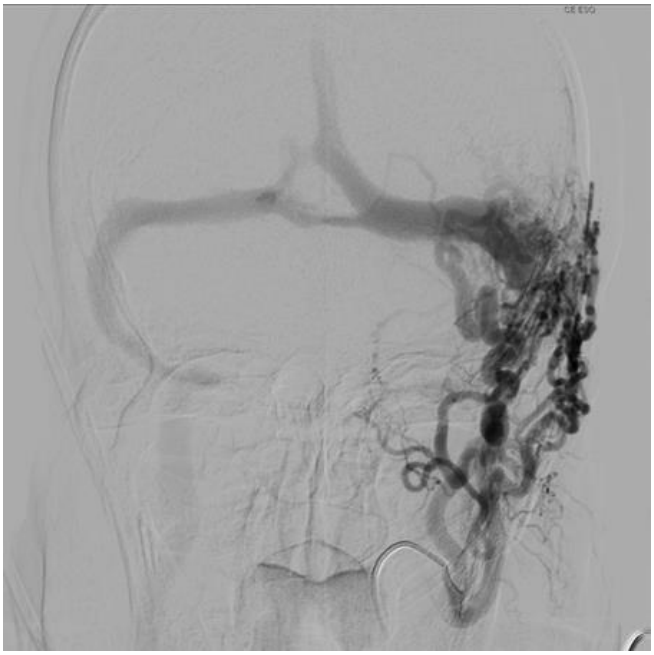

Figure 4: Left lateral sinus DAVF fed by meningeal branches of the left external carotid artery.

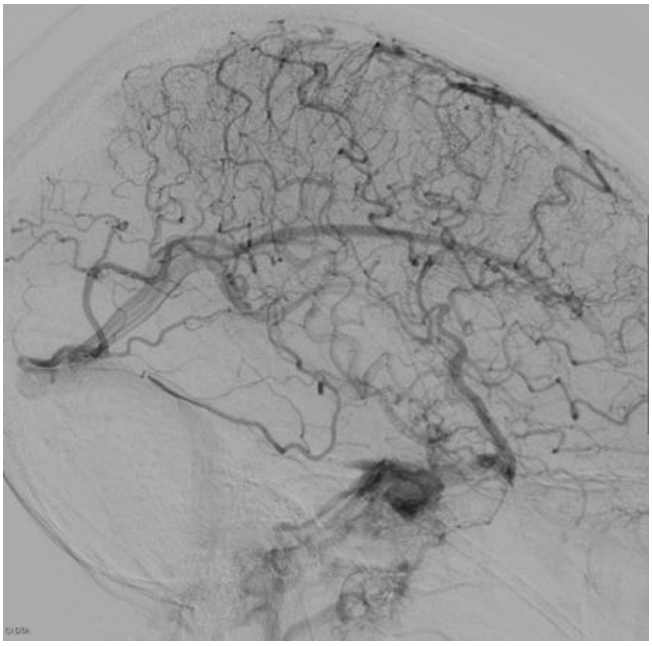

Figure 5: DAVF with retrograde cortical venous drainage and pseudophlebitic pattern.

Discussion

Dural arteriovenous fistulas correspond to a heterogeneous group of vascular malformations with their own symptoms, angioarchitecture and natural history. The fistulous connection is located on the wall of the venous sinus and it is evolved by the dural leaflets. The arterial supply can arise from dural branches of the internal carotid artery, external carotid artery and the vertebrobasilar system. In some cases, pial afferent branches of the anterior or posterior circulation may be found. Venous drainage is performed through the dural sinuses and/or through cortical/ perimedular veins [7].

Dural AVFs are responsible for 10-15% of intracranial arteriovenous malformations: 6% of supratentorial vascular malformations and 35% of infratentorial malformations [1-3,5,7,10]. The location is more commonly supra than infratentorial [3,8,9,12]. DAVFs are distinguished from pial malformations by their arterial supply through vessels that irrigate the dura and by the lack of a nidal component [1,7,10].

In most cases, the etiology is unknown, although a small percentage of patients have a history of craniotomy, head trauma or cerebral venous thrombosis. Still, it is not entirely clear whether the disease corresponds to a congenital anomaly of the dural vessels or if it arises in the context of an acquired injury [3-6]. Currently, it is believed that DAVFs arise from progressive stenosis or occlusion of a dural venous sinus. The significant prevalence of hereditary thrombotic diseases (for example, Factor V Leiden, protein C/S deficiency) in patients with DAVFs supports this hypothesis [7,10,13,14].

The mode of presentation is intrinsically linked to DAVF location and to its venous drainage pattern [1,5-9]. Thus, not rarely, symptoms can be diverse and unspecific, and the neurological examination can be innocent, which can delay the diagnosis of this pathology since it depends on a high level of clinical suspicion [1].

The venous drainage pattern will also determinate the hemodynamic status of the arteriovenous shunt which can be translated into venous distention, retrograde flow, ischemic and/or hemorrhagic changes [3,8,11,12]. Although there are many classification schemes to stratify the risk of an aggressive presentations with ICH and NHND, such as seizures, parkinsonism, cerebellar dysfunction, apathy, and cranial neuropathies; the presence of retrograde CVD is an alarming sign that foreshadows a high risk of neurological sequelae [8-10]. However, even complications like cognitive decline and dementia can improve with prompt treatment [1,2,5,7].

The most prevalent location of DAVFs in adults are in the cavernous, sigmoid, and transverse sinuses. Patients with sigmoid and transverse sinuses involvement usually have pulsatile tinnitus. Cavernous sinus fistulas can manifest with ophthalmoplegia, proptosis, chemosis, retro-orbital pain or decreased visual acuity. Pulsatile tinnitus is more frequent in women with DAVFs then in men, while men generally have more intra-parenchymal and non-traumatic subarachnoid hemorrhages [8].

Dural AVFs correspond to dynamic lesions that can regress or progress spontaneously. For this reason, any change in symptoms is a warning sign, as it may mean a change in the type of venous drainage pattern. The initial radiological evaluation includes CT and MRI images. Non-contrast CT is useful to identify intracranial hemorrhage and edema due to venous congestion. MRI is more useful because it can demonstrate vascular dilations, vascular enhancement, and signs of venous hypertension. Any suspicious flow should prompt additional assessment with CT or MRI angiography, or with conventional angiography. Evaluation by non-invasive imaging techniques can provide useful information for diagnosis, classification, and treatment stratification. However, the gold standard for evaluation of DAVFs remains conventional angiography, which allows the assessment of the hemodynamic aspects of the lesion. With this exam, signs venous congestion can be assessed, not only by the presence of CVD, but also due to the number and pattern of tortuous and engorged leptomeningeal veins (pseudophlebitic pattern) [7,10,12].

The natural history of DAVFs with cortical venous drainage that manifest with aggressive symptoms, such as ICH and NHND, is bad: the annual rate of evolution to intraparenchymal hemorrhage and non-hemorrhagic neurological complications is 7,4% to 19,0%, and the annual mortality rate is 3,8% [10,12]. For this reason, the urgent treatment of these injuries using techniques with the potential to immediately eliminate this risk (endovascular, surgical or combination of techniques) is recommended for most

patients [8]. The natural history of DAVFs with CVD diagnosed incidentally or with more benign symptoms, such as tinnitus or ophthalmic symptoms, is less aggressive, but still significant: the annual rate of intra-parenchymal hemorrhage and non-hemorrhagic neurological complications is 1,4% to 1,5% [7,10,12]. Although treatment is also advisable for most of these patients, endovascular or microsurgical techniques can be performed electively [7].

Given the diverse clinical spectrum and variable natural history, DAVFs require an individualized approach in order to choose the most appropriate treatment and each case should involve discussions among a multidisciplinary team. The risk of the treatment must be weighed against the natural history and expected clinical course of the lesion. Conservative treatment is generally indicated in patients with low-grade fistulas. In case of cortical venous drainage and fistulas located in the posterior fossa a more aggressive approach may be indicated, whoever it is important to consider the severity of the neurological symptoms and the angiographic characteristics of the DAVF [1,8].

Treatment options for DAVFs include transarterial or transvenous endovascular delivery of embolic agents for DAVF obliteration or selective CVD disconnection, microsurgical intervention for DAVF obliteration or selective CVD disconnection, stereotactic radiosurgery for DAVF obliteration, or multimodality therapy [12]. In patients with major comorbidities, short life expectancy or with complex vascular anatomy, there is a higher risk associated with the treatment of DAVF, the best strategy may eventually include stereotactic radiosurgery or careful observation. As previously noted, the ideal approach for each case must be chosen by a multidisciplinary team [1,6,10].

Conclusions

Dural arteriovenous fistulas consist of an abnormal connection, within the dural leaflets, between the meningeal arteries and dural venous sinuses and/or subarachnoid veins. The clinical signs of the disease are heterogeneous, and a high index of suspicion is necessary for the diagnosis. The recognition of this rare clinical entity is essential for an early diagnosis, referral, and intervention, to avoid the potentially serious complications of the disease.

Ethical Disclosures

Conflicts of Interest

The authors declares that there are no conflicts of interest regarding the publication of this paper.

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Confidentiality of Data

The authors declare that they have followed the protocols of their work center on the publication of data from patients.

Patient Consent

Consent for publication was obtained.

References

[1] Gandhi D, Chen J, Pearl M, et al.: Intracranial Dural Arteriovenous Fistulas: Classification, Imaging Findings, and Treatment. AJNR Am J Neuroradiol. 2012, 33:1007–13. 10.3174/ajnr.A2798

[2] Kirsch M, Liebig T, Kuhne D, et al.: Endovascular management of dural arteriovenous fistulas of the transverse and sigmoid sinus in 150 patients. Neuroradiology. 2009, 51:477–83. 10.1007/s00234-009-0524-9

[3] Nabors MW, Azzam CJ, Albanna FJ, et al.: Delayed postoperative dural arteriovenous malformations: report of two cases. J Neurosurg. 1987, 66:768 –72. 10.3171/jns.1987.66.5.0768

[4] Nishijima M, Takaku A, Endo S, et al.: Etiological evaluation of dural arteriovenous malformations of the lateral and sigmoid sinuses based on histopathological examinations. J Neurosurg. 1992, 76:600-606. 10.3171/jns.1992.76.4.0600

[5] Chung SJ, Kim JS, Kim JC, et al.: Intracranial dural arteriovenous fistulas: analysis of 60 patients. Cerebrovasc Dis. 2002, 13:79-88. 10.1159/000047755

[6] Brown RD Jr, Flemming KD, Meyer FB, et al.: Natural history, evaluation, and management of intracranial vascular malformations. Mayo Clin Proc. 2005, 80:269 –81. 10.4065/80.2.269

[7] Elhammady MS, Ambekar S, Heros RC: Chapter 9 - Epidemiology, clinical presentation, diagnostic evaluation, and prognosis of cerebral dural arteriovenous fistulas. Handbook of Clinical Neurology. Spetzler RF, Moon K, Almefty RO (ed): Elsevier, 2017. 143: 99-105. 10.1016/B978-0-444-63640-9.00009-6

[8] Rutkowski MJ, Jian B, Lawton MT: Chapter 10 - Surgical management of cerebral dural arteriovenous fistulae. Handbook of Clinical Neurology. Spetzler RF, Moon K, Almefty RO (ed): Elsevier, 2017. 143: 107-116. 10.1016/B978-0-444-63640-9.00010-2

[9] Zyck S, Jesus O, Gould GC: Dural Arteriovenous Fistula. StatPearls [Internet]. StatPearls Publishing, Treasure Island (FL); 2020. https://www.ncbi.nlm.nih.gov/books/NBK532274/

[10] Reynolds MR, Lanziano G, Zipfel GJ: Intracranial Dural Arteriovenous Fistulae. Stroke. 2017, 48:1424–1431. 10.1161/STROKEAHA.116.012784

[11] Söderman M, Pavic L, Edner G, et al.: Natural history of dural arteriovenous shunts. Stroke. 2008, 39:1735–1739. 10.1161/STROKEAHA.107.506485

[12] Zipfel GJ, Shah MN, Refai D, et al.: Cranial dural arteriovenous fistulas: modification of angiographic classification scales based on new natural history data. Neurosurg Focus. 2009, 26:E14. 10.3171/2009.2.FOCUS0928

[13] Cognard C, Gobin YP, Pierot L, et al.: Cerebral dural arteriovenous fistulas: clinical and angiographic correlation with a revised classification of venous drainage. Radiology. 1995, 194:671–680. 10.1148/radiology.194.3.7862961

[14] Gerlach R, Yahya H, Rohde S, et al.: Increased incidence of thrombophilic abnormalities in patients with cranial dural arteriovenous fistulae. Neurol Res. 2003, 25:745–748. 10.1179/016164103101202101