INTRACRANIAL DURAL ARTERIOVENOUS FISTULA MANIFESTING AS VISION DISTURBANCES IN A 36-YEAR-OLD PATIENT

Wewnątrzczaszkowa przetoka tętniczo-żylna opony twardej objawiająca się zaburzeniami widzenia u 36-letniego pacjenta

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Abstract

Intracranial dural arteriovenous fistulas are a heterogeneous group of vascular pathologies characterized by abnormal arteriovenous shunts within the dura mater. These pathological connections can occur between arteries of the dura or pia mater and veins or venous sinuses of the dura, cortex, or arachnoid. While some dural arteriovenous fistulas may remain asymptomatic, others can lead to significant clinical consequences. They can manifest in various ways, including ophthalmological symptoms. Non-invasive neuroimaging studies are crucial in their diagnosis, with endovascular therapy being the primary treatment modality. This report presents the case of a 36-year-old patient referred to the neurology department due to bilateral optic disc edema and progressive visual acuity deterioration, later diagnosed with an intracranial dural arteriovenous fistula. Given the rarity of the condition, this case aims to enhance clinicians' understanding of dural arteriovenous fistulas, their diagnostic processes, and treatment strategies.

Streszczenie

Wewnątrzczaszkowe przetoki tętniczo-żylne opony twardej to heterogeniczna grupa patologii naczyniowych charakteryzujących się nieprawidłowymi połączeniami tętniczo-żylnymi w obrębie opony twardej. Te patologiczne połączenia mogą występować między tętnicami opony twardej lub miękkiej a żyłami lub zatokami żylnymi opony twardej, kory mózgowej lub pajęczynówki. Podczas gdy niektóre przetoki mogą pozostać bezobjawowe, inne mogą prowadzić do poważnych konsekwencji klinicznych. Mogą powodować różne objawy, w tym okulistyczne. Nieinwazyjne badania neuroobrazowe mają kluczowe znaczenie w ich diagnozie, a podstawową metodą leczenia jest terapia wewnątrznaczyniowa. W niniejszej pracy przedstawiono przypadek 36-letniego pacjenta skierowanego na oddział neurologii z powodu obustronnego obrzęku tarczy nerwu wzrokowego i postępującego pogorszenia ostrości wzroku, u którego później zdiagnozowano wewnątrzczaszkową przetokę tętniczo-żylną opony twardej. Biorąc pod uwagę rzadkość tego schorzenia, przedstawiony opis przypadku ma na celu pogłębienie wiedzy lekarzy na temat wewnątrzczaszkowych przetok tętniczo-żylnych opony twardej, ich procesów diagnostycznych i strategii leczenia.

Keywords: central nervous system; dural arteriovenous fistula; vascular malformations

Słowa kluczowe: centralny układ nerwowy; przetoka tętniczo-żylna opony twardej; malformacje naczyniowe

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Introduction

Intracranial dural arteriovenous fistulas (DAVFs) are a heterogeneous group of vascular anomalies characterized by abnormal arteriovenous shunts originating from dural vessels. These vascular abnormalities involve pathological connections between arteries of the dura or pia mater and veins or venous sinuses within the dura, cortex, or arachnoid [1–3]. DAVFs account for 10–15% of all intracranial vascular malformations [3], with an incidence ranging between 0.15 and 0.29 per 100,000 persons per year [3]. While DAVFs are frequently located near dural venous sinuses, they can develop anywhere within the intracranial dura mater [2]. The most common sites include the junction of the transverse and sigmoid sinuses, the cavernous sinus, and the superior sagittal sinus [1, 3, 4]. The clinical presentation of DAVFs varies significantly depending on the fistula's location [1, 3], with symptoms ranging from mild headaches or pulsatile tinnitus to severe complications such as seizures or intracranial hemorrhage [3, 5]. The most common symptom is pulsatile tinnitus, present in 60% of cases [6], but other manifestations, such as intracranial hypertension, speech or language disturbances, cranial neuropathies, and visual impairment, as seen in our patient, are also notable [3]. Studies indicate that 83% of patients with cavernous sinus DAVFs present with proptosis or visual disturbances [7]. More aggressive severe lesions may present with progressive dementia or parkinsonism [6]. Conversely, some DAVFs may remain asymptomatic or even regress spontaneously [1–3, 8].

Case presentation

A 36-year-old male with no history of chronic illnesses was referred to the Neurology Department at the hospital in Zielona Góra from an ophthalmology clinic due to bilateral optic disc edema without evidence of optic nerve inflammation and progressive bilateral visual acuity deterioration over the past month. Neurological examination on admission revealed no meningeal or pathological signs, aside from the noted visual disturbances. An extensive laboratory workup, including oncological, virological, bacteriological, autoantibody, and thyroid panels, returned negative results.

A series of neuroimaging studies, including orbital magnetic resonance imaging (MRI), cranial computed tomography angiography (CTA), head MRI, and cranial MR angiography (MRA), were performed. Orbital MRI showed a slightly tortuous course of the optic nerves and bilateral dilation of the optic nerve sheaths up to 6 mm, measured in the sagittal plane, and 3 mm posterior to the eyeballs. Additionally, there was bilateral flattening of the posterior poles of the eyeballs at the optic nerve level, suggesting increased intracranial pressure.

In the initial magnetic resonance imaging using SWI/SWAN (Susceptibility Weighted Imaging/Susceptibility Weighted Angiography) sequences, the lesion was not visible, probably due to its location in the immediate vicinity of the skull vault bones. However, given the high sensitivity of these sequences, even small changes of this type may be visible, which may lead to a preliminary diagnosis of a dural arteriovenous fistula.

CTA revealed a tortuous distal course of the left external carotid artery and small, twisted blood vessels in the posterior cranial fossa near the left sigmoid sinus. Early contrast filling of the sigmoid and left transverse sinuses suggested a DAVF (fig. 1).

MRA confirmed the diagnosis, revealing tortuous arteries with segmental dilatation of the left posterior branch of the external carotid artery and branches of the posterior auricular artery within the dural wall at the level of the left sigmoid sinus (fig. 2).

The patient was subsequently referred for neurosurgical consultation and scheduled for fistula embolization. Diagnostic digital subtraction angiography (DSA) confirmed an arteriovenous fistula located on the dural wall near the junction of the left transverse and sigmoid sinuses. The arterial supply was from the external carotid

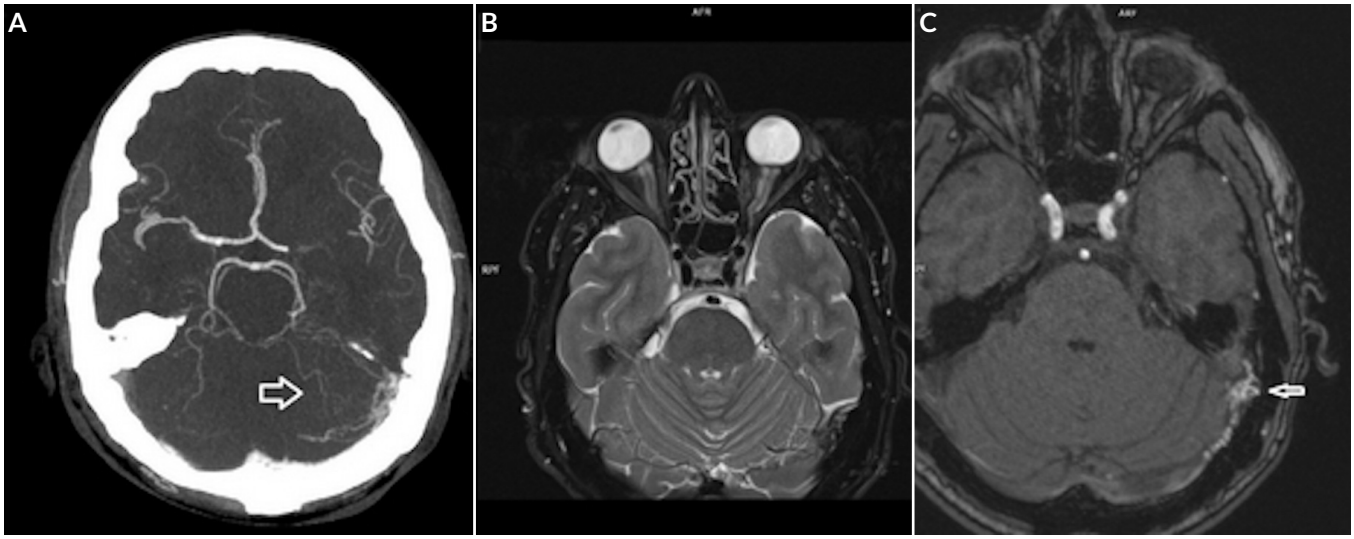

Figure 1. Angio-CT and MR scans. **A.** Abnormal vessels in the posterior cranial fossa on the left side during an angio-CT examination of cerebral arteries. **B.** Tortuous course of the optic nerves in a T2-weighted MR image of the orbits. **C.** Arteriovenous fistula in the posterior cranial fossa in the area of the left sigmoid sinus in a TOF (time of flight) angio-MR image

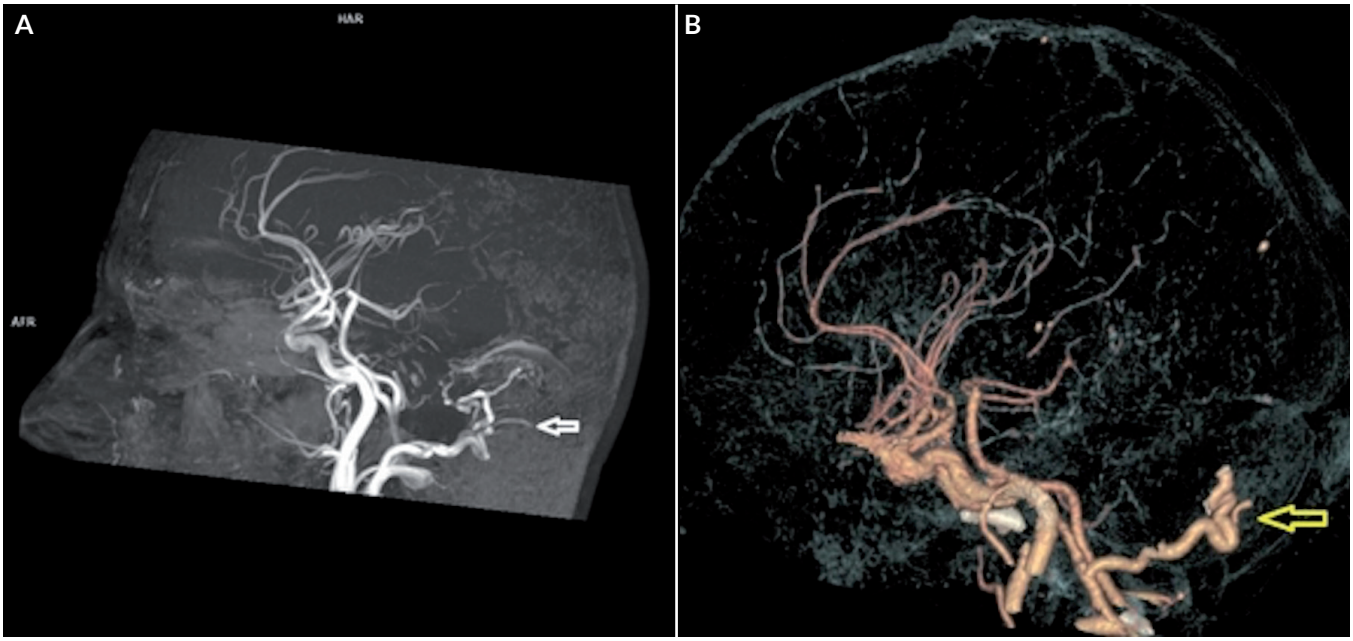

Figure 2. Angio-MR and CT angiogram scans. **A.** Image of an arteriovenous fistula in the sagittal plane in the maximum intensity projection (MIP) of an angio-MR (time of flight) TOF examination. **B.** Dilated, tortuous left external carotid artery in a virtual reality (VR) reconstruction of a CT angiogram

artery, small meningeal-hypophyseal arteries, and the left anterior inferior cerebellar artery. Retrograde venous outflow into the transverse sinus and single cortical outflows were noted, classifying the fistula as a Cognard type 2a+b. The fistula was successfully treated with the placement of eight coils, resulting in complete exclusion from circulation. Follow-up angiography demonstrated normal arterial flow. Postoperative ophthalmological assessment showed improved visual acuity and resolution of optic disc swelling.

Discussion

Dural arteriovenous fistulas (DAVFs) are a rare cause of neurological symptoms that can lead to life-threatening conditions. Although they can occur at any age, DAVFs are more prevalent in men aged 40 to 60 years [3]. Adult-onset DAVFs are typically acquired, while those diagnosed in childhood are considered congenital [6]. The exact etiology remains unclear, but factors such as prior craniotomy, trauma, thrombotic diseases, venous sinus occlusion, tumors, or systemic thrombotic activity have been implicated [1–3]. Elevated levels of cytokines related to neovascularization, including vascular endothelial growth factor (VEGF) and basic fibroblast growth factor (bFGF), may also contribute to DAVF pathogenesis [9]. As inherited thrombophilias are mentioned as risk factors for the development of dural arteriovenous fistulas, a familial history of fistulas may rarely be present [10]. The Cognard and Borden classifications are widely used to predict DAVF progression [1, 3]. Below, we present the Cognard classification, which is based on venous drainage morphology and allows for the estimation of the risk of serious fistula complications (tab. 1).

Spontaneous regression is possible even in more aggressive lesions [1, 8]. However, the type 2 a+b DAVF described in this case carries a 66% risk of cerebral hemorrhage [3].

CTA or MRA are invaluable for the initial diagnosis of DAVF, with typical findings including dilated, tortuous vessels, sinus enlargement, or obstruction [3]. However, definitive diagnosis and treatment planning require digital subtraction angiography, the gold standard for DAVF diagnosis [2, 4].

Table 1. Cognard classification of dural arteriovenous fistulas [1, 3]

Type	Description	Risk of complications
I	Located in the main sinus, with antegrade flow, without cortical drainage	Mild clinical course
IIa	Located in the main sinus, with reflux into the sinus, without cortical drainage	Intracranial hypertension in 20% of cases
IIb	Located in the main sinus, with reflux into cortical veins	Hemorrhage in 10% of cases
IIa+b	Located in the main sinus, with reflux into cortical veins and into the sinus	Hemorrhage in 66% of cases
III	Direct drainage into a cortical vein without venous ectasia	Hemorrhage in 40% of cases
IV	Direct drainage into a cortical vein with venous ectasia	Hemorrhage in 65% of cases
V	Direct drainage into spinal perimedullary veins	Progressive myelopathy in 50% of cases

The optimal treatment goal is a complete disconnection of the fistula from its venous drainage. While asymptomatic and low-grade lesions may be managed conservatively with serial monitoring, endovascular therapy (EVT) remains the mainstay of treatment, achieving long-term complete obliteration in 70–90% of cases [5]. EVT can be performed via transarterial or transvenous approaches [3]. Alternative therapeutic options include open surgery and stereotactic radiosurgery [3], with open surgery typically reserved for cases where endovascular approaches have failed or are unfeasible, such as lesions involving the anterior cranial fossa [3]. Stereotactic radiosurgery is particularly effective for cavernous sinus lesions and can be used alone or in combination with endovascular or surgical treatment [7].

Conclusion

Dural arteriovenous fistulas are complex conditions with diverse symptomatology that are often diagnosed through non-invasive imaging modalities such as CTA or cranial MRA. Endovascular therapy is a highly effective treatment method, often leading to full recovery, except in cases complicated by hemorrhagic stroke.

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