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Case Report

A case report of a multi-arterial dural arteriovenous fistula ☆

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ABSTRACT

Dural arteriovenous fistulas are rare intracranial vascular malformations with a propensity for hemorrhage. The Cognard classification system is the most widespread classification system wherein type IIB through V must be promptly treated to avoid the risk of hemorrhage. The case presented herein reports a 71-year-old male presenting with vague non-hemorrhagic neurologic deficits found to have a Cognard type III dural arteriovenous fistula with multiple arterial feeders. Although quite obvious in retrospect, a DAVF can be missed even by an astute radiologist. This should be considered a "never miss" diagnosis as it carries a risk of intracranial hemorrhage and death.

Introduction

Dural arteriovenous fistulas (DAVFs) are pathologic shunts between dural arteries and dural venous sinuses, meningeal veins, or cortical veins. DAVFs account for 10%-15% of intracranial arteriovenous malformations [1]. Some of the more common intracranial vascular malformations include arteriovenous malformations (AVMs), pial arteriovenous fistulas (PAVFs), developmental venous anomalies (DVAs), and capillary telangiectasias. An AVF is characterized by the absence of a parenchymal nidus (which is pathognomonic for an AVM).

The case presented herein reports a patient presenting with vague non-hemorrhagic neurologic deficits found to have a Cognard type III DAVF with a primary right occipital arterial feeder including smaller arterial feeders from the right middle meningeal and left occipital arteries. The arteries fistulized into the transverse dural sinus which had thrombosed proximally and distally with a resultant (non-dilated) cerebellar cortical venous drainage into a vein of Galen.

☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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Case report

A 71-year-old male with a past medical history of a reported stroke three months prior without any residual deficits presented to the emergency department by emergency medical services after the patient was involved in a minor traffic collision where he sideswiped two cars without airbag deployment or damage to windshield or steering column. The patient was noted to have mild dysarthria, tangential thoughts, confusion, and difficulty answering questions. The patient was not able to answer if there was loss of consciousness. Given these findings, a code stroke was initiated. The patient's National Institutes of Health Stroke Scale (NIHSS) score was 3 for partial hemianopsia, mild fluency loss, and slurred but intelligible speech. Additional history obtained days later after improvement of mental status revealed two instances of loss of consciousness over a lifetime including a rock dropped on his head as a child and a kick to the posterior head about 50 years prior.

Unenhanced Computed Tomography (CT) of the head revealed no abnormalities. Computed Tomography Angiography (CTA) revealed an enlarged right occipital artery with a transosseous course through the posterolateral aspect of the right occipital bone and into the right transverse sinus, however, the transverse sinus was discontinuous at the proximal and distal aspect and a prominent (non-dilated) cerebellar cortical vein was noted which drained superiorly into the vein of Galen (Fig. 1). These findings were suggestive of a Cognard type III dural arteriovenous fistula. This was, in all likelihood, related to one of the two significant head traumas in the patient's history.

Unenhanced magnetic resonance imaging (MRI) demonstrates abnormal serpentine T2 flow voids and abnormal loss of flow void at the proximal and distal aspects of the right transverse sinus (Fig. 2A). In addition, there was a small area of edema at the junction of the occipotemporal lobe (Fig. 2B).

The patient was then taken to the neurointerventional suite for digital subtraction angiography (DSA) of the bilateral internal and external carotid arteries. Selective angiography of the left occipital artery (Fig. 3A) revealed a dilated tortuous left occipital artery with a transosseous course into the dural sinus. An angiogram distal to the occipital artery ostium revealed no arterial feeder vessels (image not shown). A magnified super selective angiogram of the distal left occipital artery demonstrated well-defined early opacification of the dural sinus and a draining cortical vein (Fig. 3B). A right common carotid angiogram demonstrated four separate arterial feeder vessels from the occipital artery, ascending pharyngeal artery, and petrosal and petrosquamosal branches of the right middle meningeal artery (Fig. 4A).

The patient was treated with transarterial embolization utilizing liquid embolization. The first stage included total embolization of the left occipital artery (Fig. 3C) and marked reduction of supply from the right-sided arterial feeders (image not shown). The larger of the two right occipital arterial feeders was embolized, however, the multiple smaller tortuous arterial feeders were not amenable for embolization (Fig. 4C). As there were no signs of high-risk features of cortical venous drainage, the decision was made to have the patient follow up in the outpatient setting with the neurointerventional service in three months for a second stage embolization.

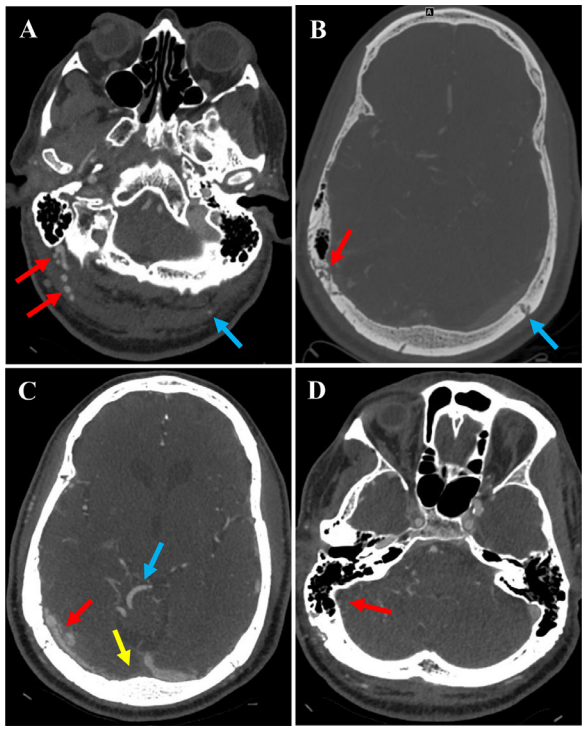

Fig. 1. A-D – Select axial contrast enhanced CTA images of the brain are shown. *Panel A* demonstrates a dilated tortuous right occipital artery (red arrows) at the level of the occipital condyles relative to the non-dilated left occipital artery (blue arrow). *Panel B* demonstrates the transosseous course of multiple small branches of the right occipital artery (red arrow). Note the disruption of the inner table compared to the normal appearance of the left lambdoid suture (blue arrow). *Panel C* demonstrates an irregular appearing right transverse dural sinus (red arrow), with absence of the medial aspect of the transverse sinus (yellow arrow), and an abnormal non-dilated cortical vein traversing the superior-most aspect of the posterior cranial fossa (blue arrow). *Panel D* demonstrates the absence of the right sigmoid sinus (red arrow).

Discussion

DAVFs are predominantly idiopathic although a small percentage of patients have a history of previous craniotomy, trauma, or dural sinus thrombosis [2]. The pathogenesis of DAVFs remains unclear. In patients with a documented antecedent cause, most occur as the result of neovascularization induced by a previously thrombosed dural venous sinus [3].

The majority of patients with DAVFs present in the fifth and sixth decades with signs and symptoms related to lesion location and the presence of complications. Cavernous DAVFs can present with ophthalmoplegia, proptosis, chemosis, retroorbital pain, or decreased visual acuity. Transverse

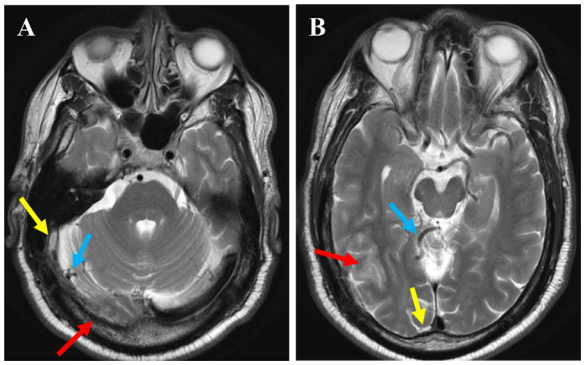

Fig. 2. (A-B) – Two axial T2-weighted MR images of the brain are shown. *Panel A* demonstrates a thin irregular-appearing right transverse sinus (red arrow), abnormal loss of flow void at the lateral-most aspect of the right transverse sinus (yellow arrow), and the proximal aspect of a non-dilated cortical vein branching from the right transverse sinus (blue arrow). *Panel B* demonstrates the distal aspect of the cortical vein draining superiorly towards the midline (blue arrow), abnormal loss of flow void at the medial-most aspect of the right transverse sinus (yellow arrow), and a small area of parenchymal edema (red arrow) at the junction of the occipital and temporal lobes

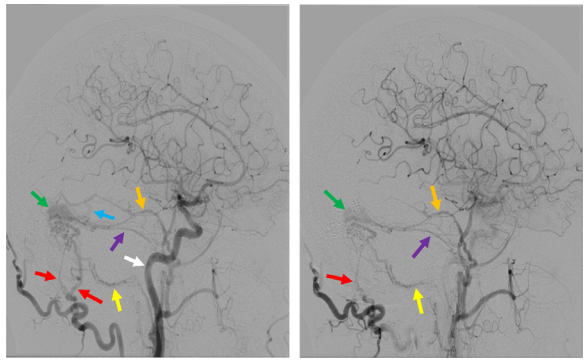

Fig. 4. (A-B) – Lateral right common carotid angiogram (*Panel A*) demonstrates multiple arterial feeders to the dural sinus (green arrow) with a faint draining cortical vein (blue arrow). Arterial feeders include the occipital artery (red arrows), ascending pharyngeal artery (yellow arrow), petrosal branch (purple arrow), and petrosquamosal branch (orange arrow) of the middle meningeal artery. The right internal carotid artery (white arrow) is shown for orientation. Lateral postembolization angiogram (*Panel B*) demonstrates absence of the larger occipital arterial feeder and persistence of the remaining smaller tortuous arterial feeders (the same colored arrow labels apply). Note the mildly decreased opacification of the dural sinus and absence of the draining cortical vein.

and sigmoid sinus DAVFs commonly present with pulsatile tinnitus. The most common complication of a DAVF is hemorrhage which can result in more severe neurologic signs and symptoms.

The venous drainage pattern, however, is often the determining factor for the severity of symptoms and provides the foundation for the Cognard and Borden classification systems.

The Borden classification organizes lesions based on the site of drainage and the presence of cortical venous drainage [4]. The Cognard system was the original and more widely used classification scheme, which also considers the direction of flow [5]. Type I lesions drain into the dural sinus, have ante-

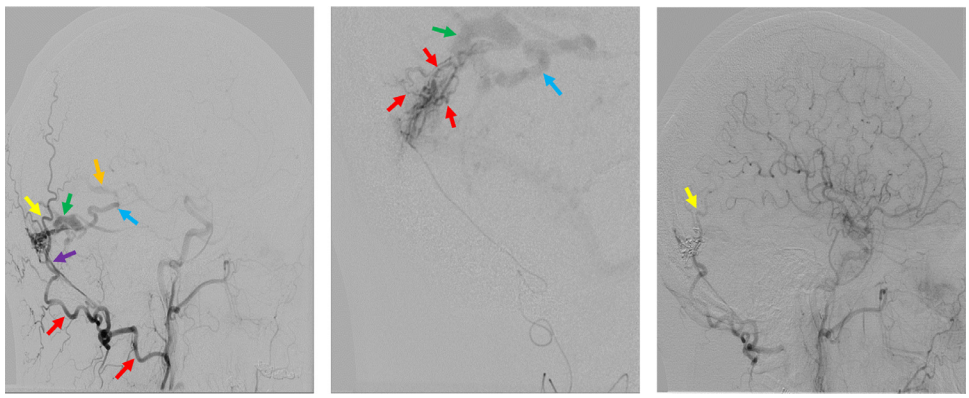

Fig. 3. (A-C) – Lateral selective angiogram of the left occipital artery (*Panel A*) demonstrates an enlarged tortuous left occipital artery (red arrows) with a transosseous course (purple arrow) into the dural sinus (green arrow) which has a prominent but non-dilated draining cortical vein (blue arrow) coursing superiorly toward the Vein of Galen (purple arrow). Note the ascending extracranial left occipital artery (yellow arrow). A magnified lateral super selective angiogram of the distal left occipital artery (*Panel B*) demonstrates multiple very small arterial feeders (red arrows). The abnormal fistulous connection to the dural sinus (green arrow) and draining cortical vein (blue arrow) is more well-defined. Note the extracranial occipital artery is not opacified (yellow arrow of Panel A). Lateral post-embolization angiogram of the left common carotid (*Panel C*) demonstrates non-opacification of the previous note dural sinus and draining vein. The extracranial occipital artery is again noted (yellow arrow).

grade flow direction, and lack cortical venous drainage. Type II lesions are further subdivided into A, B, and A+B. Type IIA lesions drain antegradely into a dural sinus without cortical venous drainage. Type IIB lesions drain antegradely into a dural sinus with cortical venous drainage. Type IIA+B drains retrogradely into a dural sinus with cortical venous drainage. Type III and IV lesions drain into a non-dilated or dilated cortical vein (without any dural sinus flow), respectively. Type V lesions drain into a spinal vein (without any dural sinus flow).

Approximately 20%-33% of DAVFs present with intracranial hemorrhage [6]. The risk of intracranial hemorrhage from Cognard type I and IIA (which lack cortical venous drainage) is extremely low. [7] Moreover, the risk of type I or IIA converting into an AVF with cortical venous drainage is low reported to be approximately 2% [8]. Spontaneous resolution of a DAVF is rare but also possible [9]. Whereas the presence of cortical venous drainage (Cognard IIB-V) is an aggressive feature which constitutes an annual mortality rate of 10.4% and an annual risk of intracranial hemorrhage of 8.1% [10]. Following an initial hemorrhage, rebleeding risk can be as high as 35% in the first two weeks [11]. Further subdividing Cognard types IIB-V into symptomatic and asymptomatic cases, demonstrates a significant difference in the annual risk of intracranial hemorrhage (7.4% vs 1.5%, respectively) [12]. In patients with retrograde cortical venous drainage, parenchymal edema is seen in up to half of patients; the addition of enhancement in areas of edema are indicative of an aggressive fistula with a high rate of hemorrhage [13].

Conventional digital subtraction angiography (DSA) remains the gold standard for detection and classification of DAVFs, however it is not without risk as 2.6% complication rates have been reported [14]. Fortunately, the increasing technological applications of CT and MRI, has greatly improved pre-angiography detection and classification. It is also important to note that a full DSA, including angiography of the bilateral internal, external, and vertebral arteries and superselective evaluation of the distal feeder branches are required to fully evaluate all possible arterial supplies.

The risks of treatment should always be weighed against the natural history of the patient's DAVF. Conservative treatment and close clinical follow-up are indicated for Cognard I and IIA lesions and any change in symptoms warrant repeat imaging. High grade lesions (Cognard IIB-V) should be treated early to avoid the risks of hemorrhage and non-hemorrhagic neurologic deficits. Transarterial embolization utilizing liquid embolization embolic agents have become the mainstay of DAVF treatment to completely eliminate the arteriovenous shunt [15]. Incomplete treatment allows for recruitment of collateral vessels and persistent risk of intracranial hemorrhage. Transvenous embolization was a pertinent aspect of treatment prior to the advent of liquid embolization as only half of cases were technically successful with arterial embolization alone [16]. However, there are several scenarios in which transvenous approaches are often preferred such as: when a DAVF is supplied by small tortuous arteries excluding safe transarterial access to the fistulous aspect, when the DAVF is only supplied by branches directly from the internal carotid or vertebral artery, when the DAVF is supplied by arteries with dangerous extracranial to intracranial anastomoses, or when the DAVF is supplied by nutrient arteries of cranial nerves [17]. For cases in which endovascular approaches have failed or are not feasible, a variety of surgical options are available including direct intraoperative embolization, resection of abnormal dura, packing of diseased sinus, disconnection of the cortical venous drainage, and skeletonization of the dural sinus [18]. As a last salvage option, stereotactic radiosurgery can also be utilized to induce endothelial cell damage and thrombosis. However, similar to treatment of AVMs, obliteration of DAVFs can take months and during that time the risk of hemorrhage remains [19].

Conclusion

The case presented herein represents a patient presenting with vague non-hemorrhagic neurologic deficits found to have a Cognard type III DAVF with four arterial feeders (left occipital, right occipital, right ascending pharyngeal, and right middle meningeal arteries) with fistulization into the transverse dural sinus and a (non-dilated) cerebellar cortical venous drainage into a vein of Galen. This was treated with transarterial embolization of multiple arterial feeders with resultant complete obliteration of the left occipital and near-complete obliteration of multiple small tortuous right-sided arterial feeders. Unfortunately, the patient was lost to follow up, however, there was the complete elimination of the cortical venous drainage thus neutralizing the high risk/aggressive factor.

Although quite obvious in retrospect, a DAVF can be missed even by an astute radiologist. This should be considered a "never miss" diagnosis, as the annual risk of hemorrhage and mortality is significant. The Cognard classification is the most widespread classification system of DAVFs which helps determine how aggressive it is. Moreover, it is essential for the angiographer to perform a complete evaluation of the entire vasculature, including the bilateral internal and carotid arteries as well as the bilateral vertebral arteries as DAVFs can have multiple feeders that are not distinctly present on CTA. Early endovascular treatment is the mainstay for Cognard types IIB-V, although surgical and stereotactic radiosurgery options can also be utilized for certain cases.

Informed consent statement

Informed written consent was obtained from the patient for publication of this case report and all imaging studies. Consent form on record.

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