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Medullary Venous Hypertension Secondary to a Petrous Apex Dural Arteriovenous Fistula: A Case Report

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Key Words
Dural arteriovenous fistula · Venous hypertension · Petrous apex · Onyx embolization

Abstract
Background: Dural arteriovenous fistulae (dAVF) are common intracranial vascular lesions typically becoming symptomatic with cortical venous hypertension and possible hemorrhage. Here, we present a case illustration of a petrous apex dAVF with marked medullary venous hypertension and a unique clinical presentation.
Methods: Case report.
Results: A 72-year-old female, whose clinical progression was significant for altered mental status and progressive weakness, presented with diplopia, right leg paresis, and ataxia. Magnetic resonance imaging revealed edema involving the medulla. On digital subtraction cerebral angiogram, the patient was found to have a petrous apex dAVF, Cognard type IV. Following treatment with Onyx embolization, her symptoms rapidly improved, with complete resolution of diplopia and drastic improvement of her ataxia.
Conclusion: The importance of this case is in the presentation and deterioration of the clinical exam, resembling an acute ischemic event. Further, this case illustrates that dAVF may cause venous hypertension with rapid onset of focal neurologic symptoms not exclusive to cortical locations.

Introduction

Intracranial dural arteriovenous fistulae (dAVF) are vascular lesions that account for 10–15% of intracranial vascular malformations. They are characterized by multiple arteriovenous fistulae connections within the leaflets of the dura mater [1]. The natural

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history of these lesions is dependent upon both the anatomical location and the venous drainage patterns [1]. Three classification systems have employed these variables in delineating low-risk from high-risk lesions. Low-risk lesions are more likely to follow a benign course, while high-risk lesions may result in hemorrhage, rebleeding, and higher mortality rates [2]. The variety of characteristics of dAVF results in a spectrum of clinical presentations, from clinically asymptomatic to profound neurologic deficits on presentation [3].

The following is a case of a petrous apex dAVF, Cognard classification type IV [4], presenting with rapidly evolving symptoms and marked medullary venous hypertension.

Case Presentation

A 72-year-old female with a past medical history significant for hyperlipidemia, hypertension, diabetes mellitus type II, and breast cancer status post left mastectomy was admitted to an outside facility after presenting to her local emergency department with a 4-day history of right leg paresis, ataxia, and vertical binocular diplopia. The patient reported first noticing the diplopia with distance gaze, and subsequently developing right-sided paresis and balance difficulties, with a propensity to fall to the right. Her symptoms persisted and she presented to a local emergency department with worsening right-sided weakness and ataxia. On examination, she was noted to have slurred, slow speech, vertical binocular diplopia, right lower extremity paresis, and difficulty maintaining her posture.

Computed tomography (CT) revealed subtle medullary edema and abnormal enhancement at the left petrous apex. CT angiogram was nondiagnostic. Magnetic resonance imaging (MRI) revealed diffuse edema involving the medulla. Cerebral angiogram confirmed a dAVF at the left petrous apex with arterial supply from the anterior temporal branch of the left middle meningeal artery. The venous drainage ran inferior along the ventral margin of the cervicomedullary junction (**fig. 1**).

During her admission, she developed worsening right hemiparesis as well as altered mental status. She was transferred to our institution 9 days after admission for further management. On examination she was alert and oriented, with normal mental status. She was noted to have vertical nystagmus and diplopia with upgaze. Her motor exam demonstrated marked weakness in the right lower extremity, more prominent proximally as well as mild weakness in the right upper extremity. She was also noted to have hyperactive reflexes on her right lower extremity with an ipsilateral Babinski sign.

She subsequently underwent Onyx embolization of the left petrous apex dAVF with complete angiographic obliteration (**fig. 2**). On postembolization day 1 she noted improvement of the right-sided weakness and diplopia. The diplopia had resolved by the following day and her motor exam had improved to the point that she could ambulate with the assistance of a walker. She was discharged on postembolization day 6 to an acute rehabilitation facility with a scheduled follow-up angiogram in 6 months.

Discussion

The anatomy of dAVF can be described as one or more arterial pedicles that can be traced to a single arteriovenous fistula site. These pedicles coalesce to a distinct lesion located in the dura – this is in contrast to a pial arteriovenous fistula or shunt, which involves the brain parenchyma [3]. The transverse-sigmoid sinuses account for 40–65% of intracranial dAVF [3].

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Though also called dural arteriovenous malformations, this nomenclature is not accurate, as a 'malformation' denotes a congenital lesion. The majority of, if not all, dAVF are acquired lesions. The pathogenesis of the fistulae is thought to be a multistep phenomenon instigated by thrombosis and resultant obstruction of a dural sinus. The inciting thrombosis may be secondary to a hypercoagulable state induced by infection, trauma, or an iatrogenic stimulus such as surgical manipulation, for example, in the case of a resection of a lesion involving the sinus [2]. There are two hypotheses to explain the formation of dAVF following dural sinus thrombosis. It has been suggested that sinus occlusion causes local hypoxia secondary to venous congestion. Hypoxia triggers the production of angiogenic factors which in turn stimulate the formation of aberrant vessels with abnormal direct arteriovenous shunts [5, 6]. Alternatively, it has been suggested that impaired venous drainage leads to increased dural sinus pressure which in turn causes compensatory dilatation of physiologic shunts connecting the thrombosed sinus and the extracranial arterial system [5, 7]. The increasing pressure leads to a recanalization of the dural sinus with arterial shunting. The sustained increased pressure can translate to increased pressures in cortical veins leading to ischemic injury secondary to impaired outflow and/or hemorrhage – both pathophysiologic mechanisms resulting in the onset of neurologic deficits [5, 7].

Multiple classification systems have been employed to describe dAVF; they all feature categorical divisions based on drainage patterns, as this characteristic of the lesions is the most prognostic with respect to risk of hemorrhage and corresponding neurologic deficits. The first classification system, established by Djindjian and Merland in 1978 [8], was based on the initial drainage of the fistulae; Borden et al. [9] and Cognard [4] introduced 2 angiographic-based classification systems that also incorporated the venous drainage. Borden et al.'s [9] system is based on the site of cortical venous drainage as well as whether there are single or multiple fistulae present in the nidus. The Cognard system [4] is more directly an adaptation of Djindjian and Merland's [8] classification utilizing venous drainage, but also incorporating dural sinus flow directionality (retro- or anterograde), and venous flow architecture into the classification system [10]. These classification systems were also applied in risk stratification with respect to the propensity for hemorrhage in dAVF.

Classification schemas were derived to correlate the anatomy, drainage, and flow patterns with clinical presentation, indicated treatment, and prognosis. Cognard et al. [4] describe the expected incidence of intracranial hypertension and hemorrhage with each classification. A low-risk lesion, type IIa, with retrograde flow into the sinus only, has potential risk for intracranial hypertension; however, if asymptomatic, intervention is not indicated and serial clinical follow-up is recommended [4]. Satomi et al.'s [11] series of 68 patients noted the benign natural history of those dAVF without cortical venous drainage. Only 1 out of 68 patients suffered a hemorrhage over a mean follow-up of approximately 28 months, and angiographic progression marked by the development of cortical venous drainage occurred in only 2 out of 50 patients with follow-up cerebral angiography [11]. Conversely, in high-risk lesions such as types III and IV the risk of hemorrhage is quoted at 40 and 65%, respectively [4]. Duffau et al.'s [12] review of 20 cases of dAVF with cortical venous drainage revealed a 35% rate of rebleeding in those patients who presented with intracerebral hemorrhage. The rebleeding occurred in the 20-day mean period between initial diagnosis and therapeutic intervention [12]. Type V lesions, with drainage into the spinal

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premedullary veins, lead to myelopathy in 50% of cases [4]. These higher-risk lesions, type IIb and higher, merit closer consideration for surgical intervention. Evolution from a low-risk to a high-risk lesion may occur in 2–4% of cases [4].

Clinical presentation of dAVF ranges from an incidental finding in an asymptomatic patient to severe neurologic deficits secondary to hemorrhage, ischemia, or venous congestion [13]. The spectrum of presentations depends on many pathophysiologic factors including the degree of AV shunting and increased venous pressure, and the presence of ischemia and recruitment of parenchymal venous drainage – all of which are related to the anatomical features that govern the flow and drainage patterns of the fistula. Headache, pulsatile tinnitus, hemorrhage, seizures, and focal deficits may occur [4].

The natural history largely depends on the venous drainage; thus the utility of the classification systems set forth by Djindjian and Merland [8], modified by Cognard [4], and expanded upon by Borden et al. [9], are utilized for both prognosis and therapeutic decision-making [14]. Conservative treatment is indicated in asymptomatic patients with no evidence of retrograde cortical venous drainage. Intervention-based treatments include stereotactic radiosurgery; transcatheter embolization, which may be transvenous or transarterial; and open surgery. With advancement in endovascular techniques, embolization (either through a transvenous or a transarterial approach) has become the mainstay of therapy. Radiosurgery can be considered for 'benign' fistulae without retrograde cortical venous drainage (such as many transverse-sigmoid sinus dAVF or indirect cavernous sinus fistulae) or for aggressive fistulae refractory to other treatments [15]. Open surgery is often employed when embolization methods have failed or are not feasible.

Conclusion

The case presented here exemplifies some of the diagnostic and therapeutic challenges as well as the clinical and radiological diversity of intracranial dAVF. The patient described suffered from a potentially dangerous dAVF with exclusive retrograde parenchymal venous drainage causing severe and progressive neurological compromise. Several pathophysiologic factors led to the patient's clinical presentation and subsequent deterioration. Increased arteriovenous shunting and venous pressures from the petrous apex dAVF caused venous hypertension and congestion in the medulla, explaining the clinical manifestations and progression. Prompt and complete endovascular exclusion of the fistula resulted in dramatic improvement of her symptoms. This case illustrates the potential for clinical progression over a brief period of time. The rapid evolution of symptoms is similar to presentations observed in cases of acute ischemic events that result from thrombosis or hemorrhage. Similar to other acute ischemic insults, there are a variety of dAVF and a corresponding continuum of presentations and indicated interventions.

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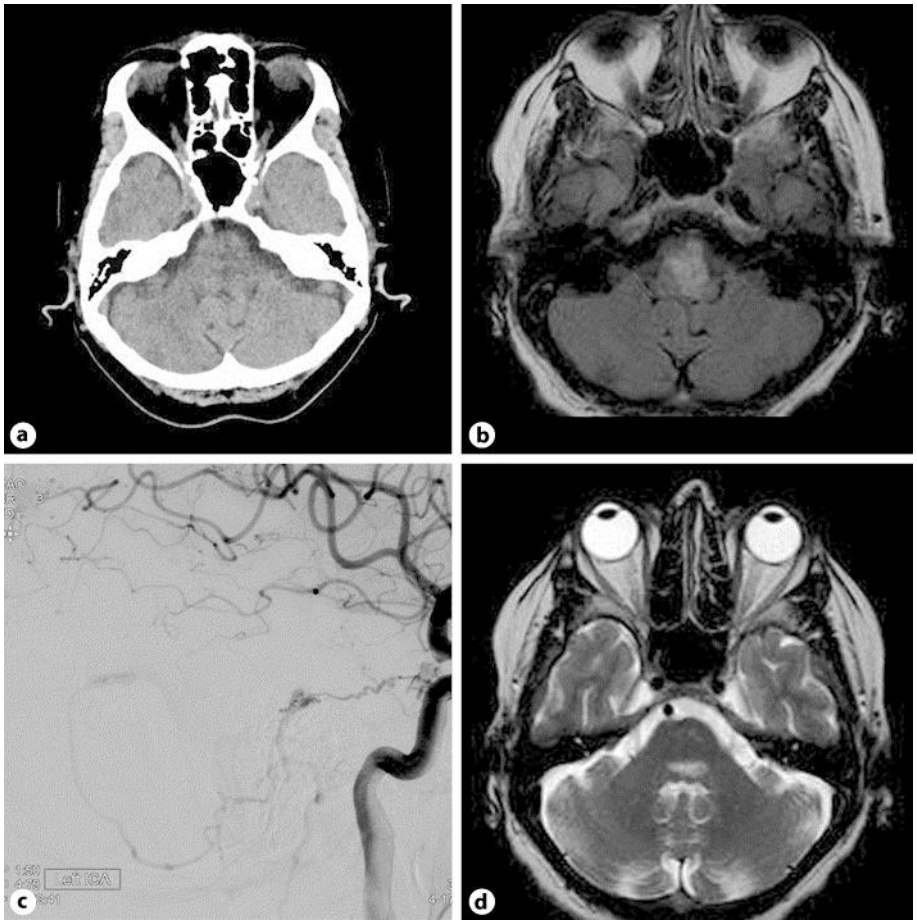

Fig. 1. Pre-embolization imaging. **a** Non-contrast head CT showing subtle medullary edema, evident following review of MRI examination. **b** Flair sequence with hyperintensity showing medullary edema. **c** Conventional digital subtraction angiogram demonstrating left ICA injection demonstrating a left tentorial apex dAVF. The fistula was noted to have multiple feeder vessels including the left meningohypophyseal trunk and the petrous branch and posterior division of the left middle meningeal artery. The venous drainage ran inferior along the ventral margin of the cervicomedullary junction. **d** Axial T2 MRI with hyperintensity showing pontine edema.

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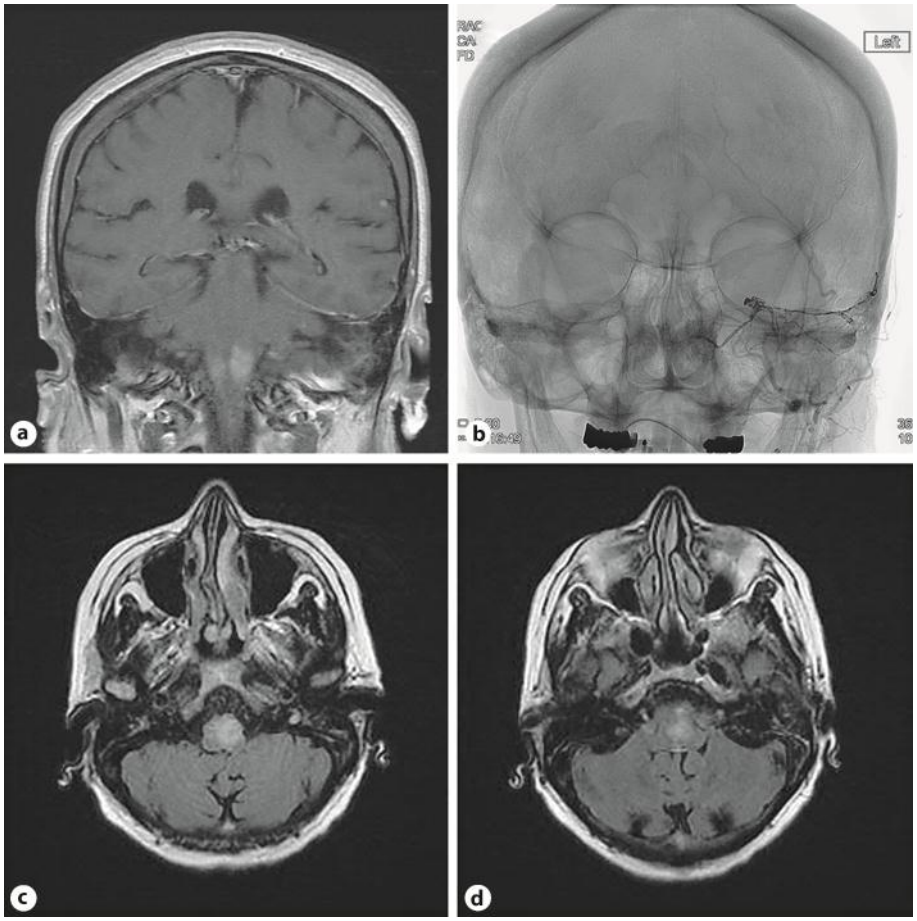

Fig. 2. Status following Onyx embolization. **a** MRI coronal post-gadolinium T1 image showing medullary edema. **b** Left tentorial apex dAVF as visualized in anterioposterior plane, unsubtracted, demonstrating the Onyx embolization at the petrous apex. **c**, **d** Axial flair sequences showing medullary edema.

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