Angioarchitectural Evolution of Clival Dural Arteriovenous Fistulas in Two Patients

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Key Words

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Abstract

Dural arteriovenous fistulas (dAVFs) may present in a variety of ways, including as carotid-cavernous sinus fistulas. The ophthalmologic sequelae of carotid-cavernous sinus fistulas are known and recognizable, but less commonly seen is the rare clival fistula. Clival dAVFs may have a variety of potential anatomical configurations but are defined by the involvement of the venous plexus just overlying the bony clivus. Here we present two cases of clival dAVFs that most likely evolved from carotid-cavernous sinus fistulas.

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Introduction

Dural arteriovenous fistulas (dAVFs) involving the clivus are rare: only 10 cases were identified in a 14-year study [1]. Clival dAVFs may present in a similar fashion to other forms of dAVFs such as carotid-cavernous sinus fistulas (CCFs), but as their prognosis is much worse and treatment strategies are very different, clival dAVFs deserve distinct consideration. Like all dAVFs, clival dAVFs have the potential to evolve and cause cortical venous reflux and life-threatening intracranial hemorrhage. In fact, since they are often not associated with any adjacent dural venous sinus, clival dAVFs more often have cortical/cerebral venous

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drainage reflux than other dAVF subtypes. Here we present two cases of clival dAVFs that most likely evolved from CCFs.

Case Reports

Case 1

A 54-year-old male was referred to our clinic for redness OD for the last 2 years and binocular horizontal diplopia. Ten years earlier, he had sustained a concussion and left orbital trauma from a motorcycle accident. He also noted hearing a pulsatile bruit behind his right ear coupled with an intermittent 'red mesh' in his central vision, typically lasting for 15 min. The day before these symptoms had begun 2 years prior, he recalled having a right-sided headache that persisted for hours. An MRI/MRA of his brain and carotid duplex ultrasound conducted 1 year prior to presentation had been unremarkable. His past medical history was otherwise significant for congenital color blindness, hypertension and hypercholesterolemia.

On examination, the patient's best corrected visual acuity (BCVA) was 20/20 OU, and his intraocular pressures (IOP) were 23 mm Hg OD and 19 mm Hg OS. A neuro-ophthalmologist and a glaucoma specialist both agreed that the discs were symmetric and lacked optic nerve head congestion or other glaucomatous changes OU. There was no resistance to repositus or evidence of proptosis. Amsler grid testing showed a central scotoma OS, although tangent screen testing was normal. Abduction OD was limited to 30% of normal with esotropia measuring 8 prism diopters on primary gaze and 30 prism diopters on right gaze, consistent with a right sixth nerve paresis. A pulsatile bruit was auscultated over the region of the right superficial temporal artery anterior to the external auditory canal, and slit lamp examination showed a classic Medusa's head pattern of conjunctival venous arterialization in the right eye.

Brain MRA/MRV showed a small focus of high signal intensity within the posterior aspect of the right cavernous sinus suspected of being a venous structure (fig. 1a). Catheter angiography revealed early opacification of a venous network near the clivus just posterior to the cavernous sinus, consistent with a low-flow clival dAVF supplied bilaterally by branches from both the internal and external carotid arteries (fig. 1c). Venous drainage was via the right inferior petrosal sinus, and there was no evidence of retrograde cortical venous drainage. There was no opacification of the superior or inferior ophthalmic veins, and the majority of the cavernous sinus failed to opacify.

Four days after the angiogram, the patient returned and complained of 2 days of increased right eye redness, swelling and pressure with minimal pain. He also noted resolution of the 'whooshing' sound in his right ear. He denied any changes in his vision or diplopia. No other neurological signs or symptoms were reported. On examination, there was a new onset of proptosis OD with increased conjunctival injection and periorbital edema. Abduction and supraduction OD were slightly restricted compared to baseline. IOPs were 31 mm Hg OD and 23 mm Hg OS. Fundoscopic examination was normal.

These findings were highly suggestive of thrombosis of the CCF secondary to angiogram contrast dye, but the MRI on that day showed no definite evidence of thrombosis of the cavernous sinus and no dilatation of the superior or inferior orbital veins (fig. 1b). The patient was started on acetazolamide 250 mg p.o. b.i.d. for ocular hypertension. Two weeks later, the patient reported diminished swelling OD. On examination, IOP was normal (20 mm Hg OU), and abduction OD had also improved to 80% of normal. A repeat catheter angiogram revealed a dramatic decrease in shunting of contrast through the vasculature to the clivus,

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confirming thrombosis of the clival dAVF (fig. 1d). There was no significant shunting from any of the previously identified contributory vessels, and there was no retrograde filling of the orbital veins.

Case 2

A 60-year-old Caucasian female with Graves' orbitopathy, who had previously had thyroid ablation for hyperthyroidism, presented to our clinic for increased redness and blurring of near vision OS worse than OD, as well as increasing migraines for the last month. She had a history of intractable binocular horizontal diplopia that had improved following both oral prednisone and bimedial rectus recession. Her presentation was consistent with a worsening of her Graves' orbitopathy except for lack of worsening diplopia in the morning and disproportionate swelling of the medial recti compared to the inferior recti on MRI. On examination, BCVA was 20/40 OD and 20/80 OS, and both eyes were tender to touch. Color plate testing showed 2/8 OU, and a 1+ afferent pupillary defect OS was noted. Slit lamp examination showed dilated, arterialized conjunctival vessels, OS greater than OD (fig. 2a, b). Fundus examination showed venous stasis retinopathy OU. These findings were suspicious for a dAVF on top of her existing Graves' orbitopathy, and a brain MRI/MRV was performed, which showed a new area of signal abnormality with mild mass effect involving both the right cerebellar tonsil and dentate nucleus, consistent with a subacute ischemic infarct and edema from associated venous congestion (fig. 3a).

The patient was urgently referred to neurosurgery for catheter angiography, which revealed a dAVF near the clivus supplied by small branches of the right internal and external carotids as well as the left internal carotid (fig. 3c). Venous drainage was exclusively via cortical venous reflux to the right cerebellum. The connecting vessels' small size prevented the fistula from being embolized from a transarterial route, and there was no safe transvenous access through the draining cortical vein. Three days later, the patient suffered a marked decline in visual acuity OU down to BCVA 20/200 OD and 20/400 OS, and IOPs were as high as 28 mm Hg OU. To rule out a possible cerebrovascular event, the patient was hospitalized and found to have extensive macular edema and bilateral central retinal vein occlusions on fundoscopy likely secondary to the dAVF, although repeat MRI/MRA failed to identify the exact location of the fistula. To help control her macular edema, she was treated with Avastin injections once OD and 3 times OS both as an inpatient and during subsequent follow-ups with a retina specialist, where her BCVA gradually improved to 20/40 OD and 20/300 OS over the course of the next 2 months.

During this time, a repeat catheter angiogram successfully identified a right-sided Cognard type III clival dAVF supplied by branches of the meningohypophyseal trunk of the left internal carotid artery. Transvenous fistula access was permitted via the right inferior petrosal sinus and the right anterior condylar vein, even though these had not been initially angiographically visible, and embolization was achieved. The fistula appeared to be completely obliterated on postembolization angiography (fig. 3d).

Following embolization, her conjunctival erythema and engorgement improved (fig. 2c, d), but 1 week later, she developed a complete right sixth nerve palsy that resolved over the next several months. Follow-up brain MRI and MRA 2 months after embolization demonstrated resolving edema in the right cerebellum (fig. 3c). Nine months later, there was no evidence of residual dural fistula on catheter angiogram. The patient continues to see a retina specialist for her macular edema.

Discussion

Clival dAVFs occur in the basilar venous plexus just above the dorsum of the clivus. The complexity of intracranial vasculature in this area affords a wide range of possibilities, but clival dAVFs are usually supplied by small branches from the external and internal carotid arteries and communicate with the dural sinuses of the posterior fossa [2]. Given the proximity and close vascular connections to the cavernous sinus, clival dAVFs are commonly misdiagnosed as the much more common CCFs, which are relatively well documented, rather than clival dAVFs, which are rare [2–7].

Further complicating matters, the ophthalmic presentation of decreased vision, chemosis, proptosis, conjunctival erythema, arterializations and even retinal hemorrhages characteristic of CCFs draining anteriorly to the superior ophthalmic vein may also be seen in clival dAVFs [3–5, 8]. Graves' orbitopathy may present similarly, or concomitantly as our case 2 illustrates [5, 8]. Particular findings that support overlapping clival dAVF and Graves' diagnoses in case 2 included the lack of worsening diplopia and rectus swelling worse in the morning, as well as relative sparing of the inferior recti as would both be expected of solitary Graves'. Interestingly, review of the surgical note from the bimedial rectus recession noted vascular engorgement of the sclera suggesting the two processes were concomitantly developing over several months.

MRI and catheter angiography may not reliably detect clival dAVFs, despite angiography being the gold standard for diagnosis, and while symptoms appear to emerge from drainage patterns, they cannot accurately predict the angioarchitecture of a dAVF [3, 7, 8]. This is partially because vascular pathways may evolve over time while symptoms of previous arrangements may persist. For example, Ernst et al. [9] also described cases where clinical complaints suddenly changed from bruit to chemosis and proptosis. The potential for evolution is consistent with theories regarding dAVF pathogenesis, where venous flow obstruction leads to proliferation of microvascular shunts within the dura [3]. Other possible factors include angiogenic growth factors from local tissue hypoxia and inflammation secondary to trauma or sinusitis [3, 8, 10]. Trauma is a commonly referenced antecedent to dAVF, and the patient in case 1 had a remote history of head trauma [3, 11].

As in case 1, pulsatile tinnitus is another symptom commonly associated with dAVF, which may present as a clival dAVF as well [3, 4, 6, 8]. This symptom is theorized to be due to turbulent flow, which is a criterion for the division of dAVFs into two categories: high-flow, where the fistula is created by a connection of a large artery to a venous sinus, or low-flow, as when the artery and sinus are bridged by a small branch vessel [12]. Our cases were radiographically categorized as low-flow, but demonstrated high-flow symptoms, which could be the result of evolution from a previous arterial arrangement. The Cognard classification scheme better assesses prognosis, and the fistula in case 2 was typed as Cognard class III indicating the presence of retrograde reflux into cortical veins [2, 3, 8, 11, 13]. Surgery is typically reserved for Cognard type III and higher [8, 14].

Endovascular intervention via embolization is the safest approach, but whether a transarterial or transvenous approach is better must be considered on a case-by-case basis due to anatomic variability [4, 15]. Embolization may itself cause further intracranial hemorrhage, cranial nerve palsies and possible recanalization with persistence of the fistula, and dAVFs that communicate with the cavernous sinus are especially predisposed to ocular nerve palsies, as illustrated in case 1 [4, 7, 10, 15]. The nerve palsy can arise either from direct irritation from the embolus, mass effect or arterial ischemia and is a potential risk of endovascular intervention [4, 7].

Following embolization, there may also be a role for anticoagulation. Case reports have described treating patients with underlying thrombophilic conditions such as factor V Leiden mutations with a combination of dual antiplatelet therapy and heparin, with varying results [16–18]. There are no consistent guidelines in the literature for particular anticoagulation regimens after endovascular intervention for dAVFs, but Bink et al. [19] point out that presenting with a dAVF in and of itself may suggest that a patient has an underlying thrombophilia, thereby justifying anticoagulation.

Interestingly, the catheter angiogram in our case 1 also caused spontaneous thrombosis, which resolved the fistula. In rare circumstances, venous hypertension can lead to thrombosis-promoting stasis, but more commonly thrombosis can result from irritation due to angiogram contrast as in case 1 [3].

Conclusions

Clival dAVFs are a unique and rare subtype of dAVF that can take on many different vascular and clinical characteristics depending on their angioarchitecture. This may evolve over time while leaving residual symptoms of past vascular patterns. These symptoms can include ophthalmic complaints, tinnitus and cerebral venous reflux with potentially high risk for intracranial hemorrhage. A high index of clinical suspicion must be maintained by the ophthalmologist to ensure referral for prompt endovascular intervention if necessary.

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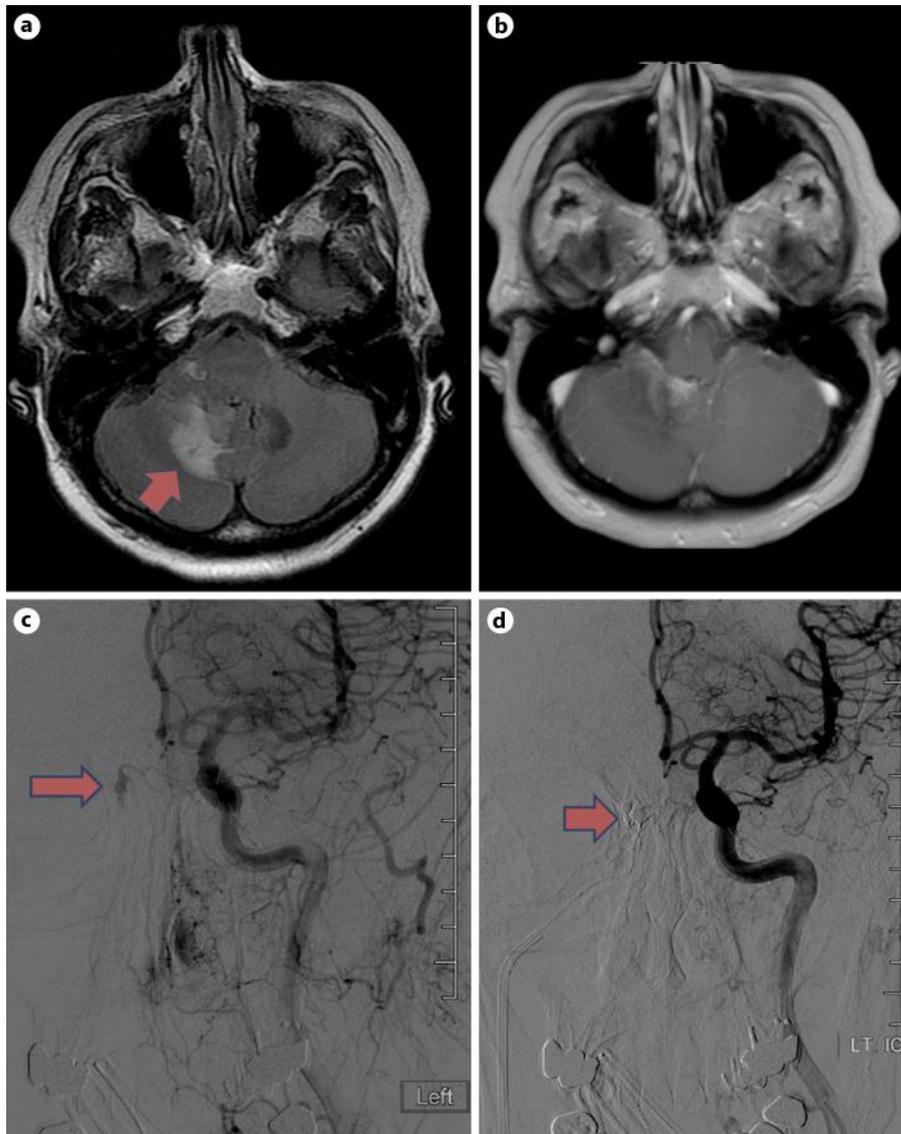
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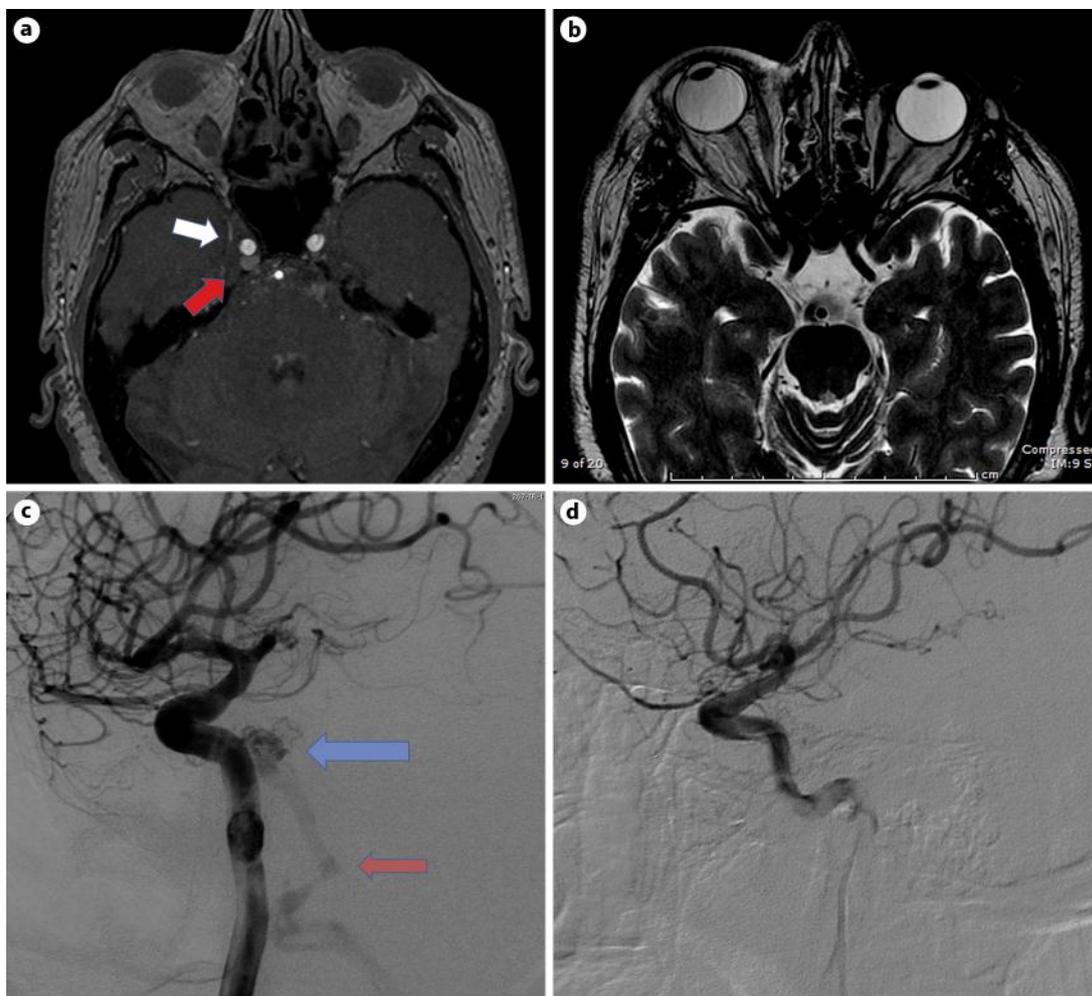
Fig. 1. Case 1. **a** MRA, before angiogram. Note abnormal signal (red arrow) within the posterior aspect of the right cavernous sinus behind the internal carotid artery (white arrow). **b** Axial T2 MRI after spontaneous thrombosis of the fistula. Note new-onset proptosis OD. **c** Right internal carotid angiogram, anterioposterior view, initial workup. Clival dAVF (blue arrow) posterior to cavernous sinus and venous drainage down the inferior petrosal sinus (red arrow). **d** Right internal carotid angiogram, anterioposterior view, after symptoms resolved. No further filling of fistula or petrosal sinus.

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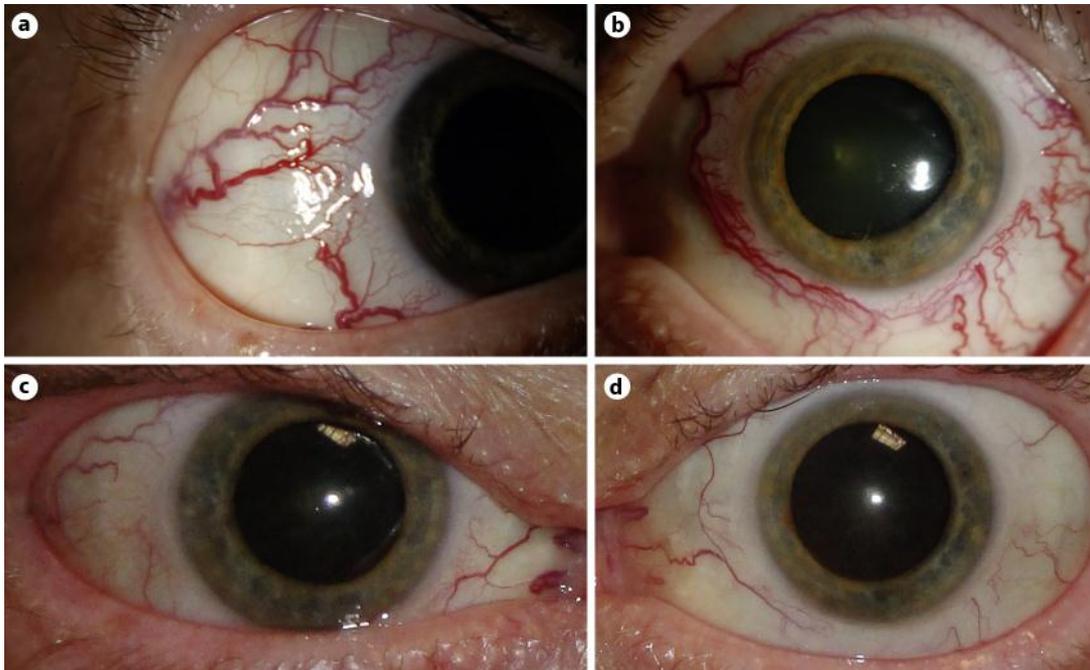
Fig. 2. Case 2. Medusa's head conjunctival venule arterialization. **a**, **b** Before embolization. Note the radial spoke-like appearance of the venules as they course away from the limbus, unique to this disease entity. **c**, **d** After embolization.

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Fig. 3. Case 2. **a** Axial FLAIR MRI, before embolization. Note edema of the right cerebellar tonsil and dentate nucleus (red arrow). **b** Axial T1 MRI with contrast, 2 weeks after embolization. Note decreased cerebellar enhancement. **c** Left internal carotid angiogram, anterioposterior view, initial workup. Early filling of vein draining into right cerebellum (arrow). **d** Left internal carotid angiogram, anterioposterior view, after embolization of the fistula. No further filling of the draining vein. Cast of embolic material in vein (arrow) is subtracted out.