CASE REPORT

Transverse and Sigmoid Sinus Dural Arteriovenous Fistula Mimicking Idiopathic Intracranial Hypertension and Carotid Cavernous Fistula

Arielle Spitze¹, David Gersztenkorn², Nagham Al-Zubidi¹, Sushma Yalamanchili¹, Orlando Diaz³, and Andrew G. Lee^{1,2,3,4,5,6,7}

¹Department of Ophthalmology, Houston Methodist Hospital, Houston, Texas, USA, ²Department of Ophthalmology, The University of Texas Medical Branch, Galveston, Texas, USA, ³Department of Neurology, Houston Methodist Hospital, Houston, Texas, USA, ⁴Departments of Ophthalmology, Neurology, and Neurosurgery, Weill Cornell Medical College, Houston, Texas, USA, ⁵Baylor College of Medicine, Houston, Texas, USA, ⁶Department of Ophthalmology, The University of Iowa Hospitals and Clinics, Iowa City, Iowa, USA, and ⁷University of Texas MD Anderson Cancer Center, Houston, Texas, USA

ABSTRACT

Intracranial dural arteriovenous fistulas (dAVFs) can produce a variety of symptoms depending on fistula location, size, and venous drainage. Although cavernous sinus fistulas (CCFs) classically present with symptoms of orbital venous congestion due to retrograde venous drainage into the superior ophthalmic vein (i.e. an arterialized “red eye”) (Miller NR. *Neurosurg Focus* 2007;23:1–15), dAVFs not localised to the cavernous sinus rarely present with a “red eye” and instead produce increased intracranial pressure, which can mimic idiopathic intracranial hypertension (IIH). The authors present a unique case of an intracranial dAVF with clinical features suggestive of both CCF and IIH. Clinicians should be aware of this possibility to avoid delayed diagnosis of the intracranial dAVF.

Keywords: Carotid-cavernous fistula, cerebral venous sinus thrombosis, dural arteriovenous fistula, idiopathic intracranial hypertension, red eye

INTRODUCTION

Carotid cavernous sinus fistulas (CCFs) are abnormal connections between the carotid arterial system and the cavernous sinus that can present with a wide variety of symptoms and signs. CCFs may be classified as direct (high flow) or type A, where the internal carotid artery directly connects with the cavernous sinus, or indirect (low flow), which are indirect connections to the cavernous sinus. Low-flow fistulas are then further subdivided into type B if dural branches of the internal carotid artery supply the fistula, type C if dural branches of the external carotid supply the fistula, or type D if both systems supply the fistula.² Although most fistulas fall into one of

these general categories, exceptions are possible depending on the location of the dural connections, flow velocity, and flow direction (anterograde vs. retrograde flow). Generally, CCFs often present to ophthalmologists with a “red eye” due to arterIALIZED episcleral vessels with or without proptosis, ophthalmoplegia, or elevated intraocular pressure (IOP) due to retrograde venous drainage into the superior ophthalmic vein (SOV).³ In contrast, dural arteriovenous fistulas (dAVFs) outside of the cavernous sinus (which arise from abnormal connections between intracranial dural arteries and the venous sinuses or cortical veins) can produce localized intracranial venous congestion and/or increased venous sinus pressure, leading to increased intracranial pressure

Received 18 July 2013; revised 26 July 2013; accepted 26 July 2013; published online 27 January 2014

Correspondence: Andrew G. Lee, MD, Chair, Department of Ophthalmology, Houston Methodist Hospital, 6560 Fannin Street, Scurlock Tower 450, Houston, TX 77030, USA. E-mail: AGLee@tmhs.org

(ICP) due to decreased cerebrospinal fluid (CSF) reabsorption. Due to the rise in ICP, this type of dAVF can produce papilloedema and thus can mimic the clinical presentation to ophthalmologists of idiopathic intracranial hypertension (IIH).⁴ However, non-cavernous dAVFs rarely cause orbital congestion or arterialisation of conjunctival/episcleral vessels.^{5,6} We describe an atypical case of a non-cavernous dAVF presenting initially with papilloedema (suggestive of increased ICP) and later with a “red eye” suggestive of a CCF. We review the literature and compare and contrast the differentiating and defining features of CCFs and non-cavernous dAVFs. To our knowledge, this is the first such case in the English language ophthalmic literature (as determined from an English language, MEDLINE/PubMed search and review for cases containing any of the same keywords as cited by this paper).

CASE DESCRIPTION

A 68-year-old asymptomatic Hispanic male was referred to the neuro-ophthalmology service at Houston Methodist Hospital for bilateral optic disc oedema found on a routine eye examination. Past ocular history was significant for an epiretinal membrane in the right eye (OD) for which he had undergone prior successful membrane peel, pars plana vitrectomy, and endolaser photocoagulation. He had a remote history many years prior to presentation of prior presumed non-arteritic anterior ischaemic optic neuropathy (NAION) in the left eye (OS), with stable visual acuity and visual field defect. He had hyperlipidaemia and chronic, poorly controlled hypertension with secondary hypertensive-induced renal failure for which he was on haemodialysis 3 times a week. The patient had hyperphosphataemia secondary to renal osteodystrophy. The patient reported a 10-pack-year tobacco history but had quit smoking many years prior. Current medications were lisinopril, hydralazine, amlodipine, sevelamer (binds dietary phosphate to prevent absorption), and cinacalcet (a calcimimetic).

On examination, the patient’s systemic blood pressure was 160/70 mm Hg. Best-corrected visual acuity was 20/30 OD and 20/40 OS. IOP measurements were 19 mm Hg OD and 20 mm Hg OS and the applanation mires were not pulsatile. Slit-lamp biomicroscopy revealed a nuclear sclerotic cataract consistent with 20/30 OD and 20/40 OS. The pupils measured 4 mm in both eyes (OU) and there was no anisocoria or relative afferent pupillary defect (RAPD). Motility examination was normal OU. Automated perimetry (Humphrey visual field [HVF] 24-2) showed small central and paracentral scotomas OD and a mild inferior arcuate nerve fibre layer defect OS (believed to be due to prior NAION OS).

There was bilateral optic disc oedema and residual macular epiretinal membrane OD but the remainder of the fundus examination was normal OU. There were no signs or symptoms of giant cell arteritis and an erythrocyte sedimentation rate and C-reactive protein were both within normal limits. A presumptive diagnosis of papilloedema was made and neuroimaging was recommended but the patient failed to return for follow-up.

Six months after his initial presentation, the patient returned with a new, 2–3-month history of progressive blurred vision OU and a “red” right eye. He described a new pulsatile feeling and constant boring pain behind the right eye, which he rated a 5/10 in severity. He also complained of persistent redness, irritation, eyelid oedema, and epiphora OD; worsening blurred vision upon bending over, new intermittent binocular diplopia; and spontaneous episodes of blurred vision lasting seconds at a time OU. On examination, best-corrected visual acuity was now 20/200 OD and 20/60 OS. IOP measured 26 mm Hg OD and 24 mm Hg OS, with very pronounced, wide Goldmann applanation mire pulsations on tonometry, OD greater than OS. There was a mild abduction and infraduction deficit OD and the alignment showed esophoria in primary gaze but a small-amplitude incomitant esotropia present in right gaze. Automated perimetry (HVF with level V stimulus) showed generalised depression with a central scotoma OD and an enlarged blind spot with cecocentral scotoma OS. There was a right RAPD but no anisocoria. No ocular or cranial bruits were auscultated. Slit-lamp examination OD showed 2+ conjunctival injection with prominent and tortuous arterialisated conjunctival and episcleral vessels OD (see Figure 1). Dilated fundus examination revealed a hazy view in OD greater than OS secondary to nuclear sclerotic cataracts. There was persistent optic disc oedema and superimposed optic atrophy OD, and moderate to severe disc oedema with multiple peripapillary haemorrhages OS (see Figure 2). There was moderate retinal vessel tortuosity observed OU. Mild macular oedema was also present OD. The patient was admitted to the hospital and started on brimonidine drops twice daily OU. Magnetic resonance imaging (MRI) of brain and orbits showed mild, periventricular T2 signal hyperintensity consistent with chronic small vessel ischaemia. There was also posterior flattening of both globes, cerebrospinal fluid signal in both retrobulbar optic nerve sheaths, and mildly dilated superior ophthalmic veins (SOVs) (see Figure 3). The cranial MR venography (MRV) showed decreased flow in the right transverse and sigmoid sinuses compared with the left side, and no flow in the right internal jugular vein (IJV). However, no MRI/MRV evidence of CCF, dAVF, or cerebral venous sinus thrombosis was identified (see Figure 4).

FIGURE 1 External view (A) showing orbital congestion OD greater than OS. A greater magnified, slit-lamp photo of the right eye is also shown (B), demonstrating arterialed episcleral and conjunctival vessels with a corkscrew appearance.

FIGURE 2 Dilated fundus examination with disc photos of OD (A) and OS (B). The images are blurred due to visually significant cataracts OU (OD greater than OS), but disc oedema is clearly present OU. The oedema is more severe in OS with peripapillary flame haemorrhages, indicating the oedema is more acute than that seen in OD.

FIGURE 3 MRI of brain and orbits read by radiology as significant for mild, periventricular hyperintensities likely consistent with chronic small-vessel ischaemia. With detailed history and re-review with neuroradiology, the MRI also demonstrated posterior flattening of both globes (B, top arrow), fluid in both retrobulbar optic nerve sheaths (B, bottom arrow), disc oedema, and equal-sized, but mildly dilated SOVs (see arrows in A pointing to the right and left SOV). (A) T1-weighted axial MRI FLAIR (fluid attenuation inversion recovery) sequence, whereas picture (B) is a T2-weighted axial propeller sequence.

FIGURE 4 Cerebral MRV, demonstrating a smaller right IJV (bottom arrow) than left, with no flow within the right IJV. Less flow was also noted within the right transverse (top arrow) and sigmoid sinuses (left arrow) as compared with the left side. The flow abnormalities were initially thought to be secondary to congenital asymmetry of the right transverse sinus, sigmoid sinus, and IJV as compared with the left, but were later discovered to be secondary to venous sinus thrombosis.

FIGURE 5 Angiogram image pre-Onyx coil. Right external carotid injection demonstrates an extensive dAVF of the right sigmoid sinus supplied by dural branches of the right occipital and right middle meningeal artery.

Cerebral angiography demonstrated diffuse and extensive dAVFs involving the right transverse and sigmoid sinus (see Figures 5–7). There was moderate stenosis at the level of the proximal right transverse sinus and torcula. The dAVF showed drainage into the right sigmoid sinus and retrograde drainage into the right transverse sinus representing a type IIA Cognard classification⁷ dAVF. The delayed venous phase showed slow flow into the right cavernous sinus, which was subsequently draining into the right superior ophthalmic vein. The right IJV was thrombosed in its most proximal segment. There was no

FIGURE 6 Angiogram image pre-Onyx coil of the early venous phase demonstrating filling of the right inferior petrosal sinus (see arrow).

FIGURE 7 Angiogram image pre-Onyx coil showing the inferior petrosal sinus (line arrow) draining into the right cavernous sinus (middle arrow) and right superior ophthalmic vein (far left arrow).

drainage of the cerebral veins during the venous phase into the right transverse sinus, indicating a non-functional status. The left IJV was occluded distally at the cervical level, with no communication to the left brachiocephalic vein. Instead, the left IJV was draining into the brachiocephalic trunk via a cervical collateral circulation. Embolisation of the right transverse and sigmoid sinus AVF was performed with Onyx, resulting in complete obliteration of the dAVF.

FIGURE 8 External photo 5 months post-embolisation. There is a small sensory exotropia OD. All of the previously noted vascular conjunctival congestion has resolved.

Unfortunately, post-embolisation, the patient developed a rapidly expanding right temporal lobe intraparenchymal haematoma associated with a small right temporal subdural haematoma and 1.1 cm subfalcian herniation. An emergency craniotomy was performed with drainage of the haemorrhage. The patient was discharged to a rehabilitation facility and made a slow recovery. Post-operative imaging showed resolution of the subfalcian herniation, slow reabsorption of the intracranial haemorrhage, and residual encephalomalacia of the right temporo-occipital region.

The patient was discharged from rehabilitation and returned to the neuro-ophthalmology clinic for follow-up examination 5 months later. His vision was 20/200 OD and 20/60 OS. His abduction and downgaze deficits had resolved, although he had a residual 7 prism dioptre exotropia OD, which was thought to be sensory due to full extraocular muscle function OU. Anterior segment examination was normal without any evidence of conjunctival congestion or chemosis (see Figure 8). A left homonymous hemianopsia was evident on automated visual field testing. On fundus examination, there was resolution of the previous optic disc oedema with secondary optic atrophy OU.

DISCUSSION

Patients with symptoms and signs of either increased ICP (e.g. papilloedema) or a CCF (e.g. “red eye”) often present to ophthalmologists first. Typically it is not difficult to make the clinical and radiographic diagnosis of each condition separately, but when there is overlap between the presenting findings for presumed IIH and CCF, a single unifying diagnosis of dAVF should be considered. Anteriorly draining CCFs produce the classic symptoms of orbital congestion, such as chemosis, proptosis, ophthalmoplegia, and arterialised conjunctival injection.¹ Orbital venous pressure is also increased in proportion to the severity of the fistula; thus, with progressively higher resistance to venous drainage, secondary pathologic processes can occur, including glaucoma (secondary open or closed angle), choroidal effusions, retinal detachment, retinal vessel tortuosity and sluggish

flow with associated branch or central retinal vein occlusion, macular oedema, disc oedema, or ischaemic optic neuropathy.^{9,10} Our patient presented with papilloedema suggestive of increased ICP (e.g. IIH) but then developed delayed signs of elevated intraorbital venous pressure, including right-sided arterialised, corkscrew-shaped episcleral veins (see Figure 1), elevated IOP, large-amplitude mire pulsations, and disc oedema. His indolent, chronic course, including worsening disc oedema over a 6-month period, the development of a red eye, and signs consistent with high venous pressure in the SOV and/or inferior ophthalmic veins (but without audible bruit), all suggested a dAVF of the cavernous sinus, even though the bilateral optic disc oedema was more suggestive of increased ICP.¹¹

Cerebral angiography revealed a dAVF involving the right transverse and sigmoid sinus also associated with right transverse sinus, sigmoid sinus, proximal right IJV, and distal left IJV thrombosis. Abnormal connections were present between the transverse and sigmoid sinus and multiple dural branches originating from the right middle meningeal artery and transmastoid branch of the right occipital artery (see Figures 5–7). Due to multiple foci of high venous resistance, this dAVF was slowly draining into the right cavernous sinus. The increased flow within the cavernous sinus was then directed into the SOV, which was responsible for the signs mimicking a CCF.

Intracranial dAVFs are abnormal connections between dural arteries and any portion of the intracranial venous sinuses, meningeal veins, or cortical veins,¹² and represent 10–15% of all intracranial vascular abnormalities. These dAVFs most commonly occur in the transverse or cavernous sinus (i.e. CCF), but can be located anywhere within the intracranial dura.¹³ Intracranial dAVFs can have secondary cavernous sinus involvement in 10–57% of cases.^{14–17} However, unlike fistulas directly or indirectly connected to the cavernous sinus, in our case, there was no involvement of the cavernous sinus by the fistula itself. Rather, the fistula was ultimately draining into the cavernous sinus, giving a falsely localizing sign of a CCF.

Ocular symptoms related to disc oedema are not uncommon in non-cavernous intracranial dAVFs, causing papilloedema due to generalised venous hypertension; similarly, cavernous dAVFs commonly cause ocular symptoms related to localised orbital venous congestion. However, ocular symptoms and signs of both increased ICP and localised orbital venous congestion have rarely been described in a non-cavernous dAVF. In a review by Oh et al.,¹² only 2 of 95 patients with dAVFs had “ocular symptoms” and 3 of 95 patients presented with “visual symptoms.” In another review of patients with multiple dAVFs not involving the cavernous sinus, one patient presented with ocular symptoms (exophthalmos).¹⁸

Another review by Chung et al.¹⁷ examined 60 patients with dural intracranial arteriovenous malformations. Ocular symptoms were present in the majority of patients with CCFs but were rare in all other fistula types (one patient with a transverse sinus fistula had exophthalmos and one patient with a dural convexity fistula had exophthalmos and conjunctival injection). In yet another review of 17 intracranial arteriovenous malformations, no ocular symptoms were present.¹⁴

The distinction between CCF and non-cavernous dAVF is important because fistulas involving the cavernous sinus are much less likely to cause severe neurologic sequelae (i.e. haemorrhage, seizure) unless they have cortical venous drainage.¹⁸ In contrast, intracranial dAVFs involving the transverse-sigmoid sinuses usually require treatment due to the unfavourable natural history of untreated lesions (e.g. intracranial haemorrhage, non-haemorrhagic progressive neurologic deficits, or non-specific signs secondary to increased ICP).^{13,14}

We propose two possible hypotheses for the ocular vascular congestion seen in our case: (1) increased ICP secondary to venous sinus thrombosis with high overall intracranial venous pressure (decreased outflow of CSF), or (2) high venous flow transmitted in a retrograde fashion anteriorly through the petrosal or sphenoid sinuses from the transverse and/or sigmoid sinus arterial fistulas. More likely, our patient's presentation was a combination of both mechanisms.

The pathogeneses of fistula and/or venous sinus thrombosis formation are not identified in all cases, but it is known that both disease processes are often found associated together; thus, it is thought that one condition predisposes to the other (although which one is the initiating process remains controversial).¹⁸ Venous hypertension may also play a major role in the pathogenesis of dAVF formation through up-regulation of angiogenic mediators such as fibroblast growth factor and vascular endothelial growth factor (secondary to tissue ischaemia), exacerbating the fistula.²⁰ Small-scale studies show that about a third of dAVFs have a congenital hypercoagulable abnormality,² with thrombi frequently found in dAVFs, often downstream from the fistula.²¹ Pregnancy and other conditions with a propensity to cause hypercoagulation can also increase this risk, and should heighten the suspicion for sinus thrombosis in a patient presenting with symptoms of elevated ICP. Our patient had severe, persistent hypertension in the setting of renal failure and chronic haemodialysis, a factor that certainly could have increased the risk of both fistula formation and thrombosis.²²

In summary, dAVFs can involve the cavernous sinus directly or indirectly and ocular findings may occur due to elevated ICP or venous sinus thrombosis. Thus, a dAVF can mimic IIH by producing papilloedema from increased ICP, or mimic a CCF by

producing localised arterialised orbital signs and symptoms. To our knowledge, this is the first described case of a dAVF producing both papilloedema and an arterialised "red eye" in the English language ophthalmic literature. Most importantly, the diagnosis could require catheter angiography, because conventional computed tomography or MRI studies (even with MRV), as highlighted in this case, may not reveal the diagnosis of dAVF. Thus, clinicians should be aware of this unique combination of ocular findings, which may be the only clues to the presence of an intracranial, life-threatening dAVF.

Declaration of interest: The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

This work was supported in part by an unrestricted grant from Research to Prevent Blindness.

Note: Figures 1, 2 and 8 of this article are available in colour online at www.informahealthcare.com/oph

REFERENCES

- [1] Miller NR. Diagnosis and management of dural carotid-cavernous sinus fistulas. *Neurosurg Focus* 2007;23:1–15.
- [2] Benndorf G. Etiology, prevalence and natural history of dural cavernous sinus fistulas (DCSFs). In: Benndorf G, editor. *Dural Cavernous Sinus Fistulas. Diagnosis and Endovascular Therapy*. Berlin and Heidelberg: Springer, 2010;65–83.
- [3] Das JK, Medhi J, Bhattacharya P, Borah N, Bhattacharjee K, Kuri GC, Deka H, Goswami BJ. Clinical spectrum of spontaneous carotid-cavernous fistula. *Indian J Ophthalmol* 2007;55:310–312.
- [4] Cognard C, Casasco A, Toevis M, Houdart E, Chiras J, Merland JJ. Dural arteriovenous fistulas as a cause of intracranial hypertension due to impairment of cranial venous outflow. *J Neurol Neurosurg Psychiatry* 1998;65:308–316.
- [5] Kiyosue H, Hori Y, Okahara M, Tanoue S, Sagara Y, Matsumoto S, Nagatomi H, Mori H. Treatment of intracranial dural arteriovenous fistulas: current strategies based on location and hemodynamics, and alternative techniques of transcatheter embolization. *Radiographics* 2004;24:1637–1653.
- [6] Meyers PM, Halbach VV, Dowd CF, Lempert TE, Malek AM, Phatouros CC, Lefler JE, Higashida RT. Dural carotid cavernous fistula: definitive endovascular management and long-term follow-up. *Am J Ophthalmol* 2002;134:85–92.
- [7] Davies MA, TerBrugge K, Willinsky R, Coyne T, Saleh J, Wallace MC. The validity of classification for the clinical presentation of intracranial dural arteriovenous fistulas. *J Neurosurg* 1996;85:830–837.
- [8] Chaudhry IA, Elkhamry SM, Al-Rashed W, Bosley TM. Carotid cavernous fistula: ophthalmological implications. *Middle East Afr J Ophthalmol* 2009;16:57–63.
- [9] Grumann AJ, Boivin-Faure L, Chapot R, Adenis JP, Robert PY. Ophthalmologic outcome of direct and indirect carotid cavernous fistulas. *Int Ophthalmol* 2012;32:153–159.
- [10] Stiebel-Kalish H, Setton A, Nimii Y, Kalish Y, Hartman J, Huna Bar-On R, Berenstein A, Kupersmith MJ. Cavernous

- sinus dural arteriovenous malformations: patterns of venous drainage are related to clinical signs and symptoms. *Ophthalmology* 2002;109:1685–1691.
- [11] Benndorf G. Neuro-ophthalmology in dural cavernous sinus fistulas (DCSFs). In: Benndorf G, editor. *Dural Cavernous Sinus Fistulas. Diagnosis and Endovascular Therapy*. Berlin Heidelberg: Springer, 2010;85–96.
 - [12] Oh JT, Chung SY, Lanzino G, Park KS, Kim SM, Park MS, Kim HK. Intracranial dural arteriovenous fistulas: clinical characteristics and management based on location and hemodynamics. *J Cerebrovasc Endovasc Neurosurg* 2012;14:192–202.
 - [13] Gupta A, Periakaruppan A. Intracranial dural arteriovenous fistulas: a review. *Indian J Radiol Imaging* 2009;19:43–48.
 - [14] Awad IA, Little JR, Akarawi WP, Ahl J. Intracranial dural arteriovenous malformations: factors predisposing to an aggressive neurological course. *J Neurosurg* 1990;72:839–850.
 - [15] Malek AM, Halbach VV, Higashida RT, Phatouros CC, Meyers PM, Dowd CF. Treatment of dural arteriovenous malformations and fistulas. *Neurosurg Clin N Am* 2000;11:147–166, ix.
 - [16] Satomi J, van Dijk JM, Terbrugge KG, Willinsky RA, Wallace MC. Benign cranial dural arteriovenous fistulas: outcome of conservative management based on the natural history of the lesion. *J Neurosurg* 2002;97:767–770.
 - [17] Chung SJ, Kim JS, Kim JC, Lee SK, Kwon SU, Lee MC, Suh DC. Intracranial dural arteriovenous fistulas: analysis of 60 patients. *Cerebrovasc Dis* 2002;13:79–88.
 - [18] Ha SY, Kwon YS, Kim BM, Kim DI, Kim DJ. Clinical and angiographic characteristics of multiple dural arteriovenous shunts. *AJNR Am J Neuroradiol* 2012;33:1691–1695.
 - [19] Cha KC, Yeon JY, Kim GH, Jeon P, Kim JS, Hong SC. Clinical and angiographic results of patients with dural arteriovenous fistula. *J Clin Neurosci* 2013;20:536–542.
 - [20] Shin Y, Uranishi R, Nakase H, Sakaki T. Vascular endothelial growth factor expression in the rat dural arteriovenous fistula model [abstract only]. *No To Shinkei* 2003;55:946–952.
 - [21] Mironov A. Pathogenetical consideration of spontaneous dural arteriovenous fistulas (DAVFs). *Acta Neurochir (Wien)* 1994;131:45–58.
 - [22] Smits JH, van der Linden J, Blankestijn PJ, Rabelink TJ. Coagulation and haemodialysis access thrombosis. *Nephrol Dial Transplant* 2000;15:1755–1760.