Case report

Dural carotid cavernous fistula following hypertensive emergency

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SUMMARY

A 58-year-old woman was referred to the outpatient ophthalmology clinic with progressive bilateral eye redness and vision loss. She had presented 2 weeks earlier with an episode of hypertensive emergency. CT angiography revealed bilateral superior ophthalmic vein (SOV) dilation, prompting further workup with a cerebral angiogram. Subsequent imaging revealed an indirect (type D) carotid-cavernous fistula (CCF) with venous drainage into both SOVs and cavernous sinuses. Successful treatment of the CCF with coil embolisation required interdisciplinary teamwork between ophthalmologists and interventional neuroradiologists. The patient made a substantial visual recovery following treatment.

BACKGROUND

Carotid-cavernous fistula (CCF) occurs in the presence of abnormal vascular connections between the internal or external carotid arteries (ECAs) and branches and the cavernous sinus.¹ These vascular shunts are commonly secondary to trauma, but can also present spontaneously in the setting of systemic disease that results in damage to arterial walls.^{2,3} Herein, we present the first known case of a dural CCF following an episode of hypertensive emergency. This case not only highlights a rare disease presentation but is also clinically important for physicians since it reinforces the high index of suspicion that should be maintained for a diagnosis of CCF when patients present with bilateral conjunctivitis refractory to traditional treatment.

CASE PRESENTATION

A 58-year-old previously healthy Caucasian woman presented to the emergency department (ED) with 2 weeks of progressive headache and intermittent visual changes. CT angiography of the head was reported as normal. Her blood pressure measured 200/105, and she was diagnosed with hypertensive emergency. After her blood pressure was stabilised in the ED, she was started on with oral antihypertensive medications, and with close monitoring by her family physician, her blood pressure remained well controlled. Her previous surgical history was significant for a left total hip replacement. She had no personal or family history of ophthalmologic disease.

She was then seen in the emergency ophthalmology department 2 months later for evaluation of progressive, bilateral eye redness that began 2

weeks after her prior visit to the ED. These ocular symptoms were refractory to olpatadine and loteprednol eye drops two times per day in both eyes, as prescribed by her family doctor. Ophthalmological examination revealed a best-corrected visual acuity (BCVA) of 20/60 in the right eye and 20/40 in the left eye. Intraocular pressure (IOP) measured 26 mm Hg in the right eye and 27 mm Hg in the left eye. Pupils were 4 mm bilaterally with no afferent pupillary defect. Anterior segment examination showed bilateral upper and lower lid oedema, conjunctival injection and chemosis, and engorged, tortuous vessels (figures 1 and 2). Hertel exophthalmometry revealed measurements of 24 mm and 22 mm in the right eye and left eye, respectively. Testing of extraocular movements demonstrated a -3 abduction deficit in the right eye. Colour vision testing with Ishihara plates was 10/11 bilaterally. Ophthalmoscopy revealed a cup to disc ratio of 0.4 bilaterally, with a flame-shaped haemorrhage noted superiorly to the right optic disc. The retinal vasculature and periphery appeared within normal limits.

INVESTIGATIONS

The initial CT angiography head performed during her episode of hypertensive emergency was reviewed. Axial images at that time showed early filling of both cavernous sinuses and left greater than right enlargement of the superior ophthalmic veins (SOVs) (figure 3). Given the clinical history and radiographic findings, we maintained a high index of suspicion for a CCF as the cause for the patient's current symptoms and urgently referred her to interventional neuroradiology for diagnostic angiography and embolisation. The cerebral angiogram showed an indirect (type D) left CCF, with arterial supply from the left middle meningeal and ascending pharyngeal artery branches, and dural branches of the left meningohypophyseal trunk; venous drainage was also seen crossing the circular sinus into the right ophthalmic vein (figure 4). There was no evidence of cortical venous drainage.

TREATMENT

Successful transvenous coil embolisation of the left cavernous carotid fistula was performed, with coils placed in the left and right SOVs and cavernous sinuses (figure 5). No residual arteriovenous (AV) shunting was noted at the end of the procedure.

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Figure 1 External photograph of the patient with marked bilateral injection, chemosis and vessel tortuosity.

OUTCOME AND FOLLOW-UP

The patient was seen in follow-up 2 weeks after coil embolisation with complete resolution of conjunctivitis and diplopia. At time of examination, her visual acuity was 20/25 in both eyes. She continues to be seen by the ophthalmology service at regular intervals.

DISCUSSION

CCF occurs when there is an abnormal communication between the carotid artery and the cavernous sinus. CCFs can be classified based on haemodynamics (high flow vs low flow), anatomy (direct vs indirect/dural) or aetiology (traumatic vs spontaneous). Previous reports have identified that greater than 75% of all CCFs are secondary to trauma.²⁻⁴ The mechanism is thought to be due to shear forces, or by direct damage sustained from bony injuries. Conversely, dural CCFs arise secondary to systemic diseases including hypertension, Ehlers-Danlos, fibromuscular dysplasia and pseudoxanthoma elasticum.⁵⁻⁶ The Barrow classification system further organises CCFs into four types based on angiographic features: type A CCFs are the most common and present as direct, high-flow communications between the internal carotid artery (ICA) and cavernous sinus¹²; type B, C and D dural CCFs are characterised as indirect and low-flow communications. Dural CCFs occur when connections are formed between the cavernous sinus and smaller arterial branches of the internal or ECA. These fistulas classically involve the various meningeal branches of the ICA (type B) and ECA (type C), or both (type D).¹

Based on the interventional angiography findings, our patient's CCF can be categorised as a type D given its supply from both ICA and ECA branches. In our search of the existing literature, we were unable to find any previously documented cases of type

Figure 3 Axial CT head scan with contrast. Red arrows demonstrate bilateral (left greater than right) superior ophthalmic vein dilation.

D dural CCF following hypertensive emergency. It is possible that the patient's untreated hypertension and subsequent hypertensive emergency contributed to the formation of a cavernous sinus thrombosis, precipitating the formation of fistulae between the dural arteries and cavernous sinus.¹⁷⁻⁸ Another consideration could be that the hypertensive emergency resulted in the rupture of thin-walled dural arteries, allowing pre-existing dural arterial anastomoses to shunt collateral blood into the cavernous sinus.⁹⁻¹⁰

Symptoms associated with CCFs include blurred vision, headache, tearing, diplopia or an audible bruit described as a 'whooshing' noise.¹²⁻⁴ The manner of presenting signs, however, depends on whether it is a direct or indirect fistula. Direct,

Figure 2 Anterior segment photographs of the right (top) and left (bottom) eye.

Figure 4 Indirect type left carotid cavernous fistula. Venous drainage is seen across the circular sinus to the right cavernous sinus.

Figure 5 Postcoil embolisation of right and left superior ophthalmic veins and cavernous sinuses.

high-flow CCFs will commonly present with rapidly progressive chemosis, proptosis, increased IOP, conjunctival vascular tortuosity, strabismus secondary to cranial nerve palsy or orbital congestion, and in severe cases central retinal vein occlusion.^{9 11 12} Indirect, low-flow CCFs are more difficult to diagnose given their indolent nature. As with our patient, these cases often present with an unremitting conjunctivitis as the only symptom. It is only after chronic venous engorgement that other signs develop, such as conjunctival arterialisation, chemosis, proptosis and elevated IOP.⁴

To confirm the presence of a CCF, various imaging modalities can be employed. Radiographic findings on non-contrast and CT angiographic or MRI include orbital oedema, proptosis, enlargement of extraocular muscles due to venous congestion and enlarged SOVs.⁹ However, cerebral angiogram remains the gold standard imaging modality in confirming a diagnosis and is required whenever there is a high index of suspicion for a CCF.

Treatment approaches for CCFs can be conservative, endovascular or surgical. Conservative management and observation are typically reserved for low-flow fistulas that have been demonstrated to close on their own. Al-Mufti *et al*³ also found that carotid artery compression has some demonstrable efficacy in the treatment of unilateral, low-flow CCFs. However, these approaches are generally not recommended if there are secondary ocular complications from the CCF, or if there are high-risk vascular features such as ICA dissection.¹³ Endovascular embolisation remains the first-line therapy for fistula closure. Techniques include transarterial embolisation with flow-diverting stents, balloons, detachable coils, or embolic agents such as n-butyl cyanoacrylate.³ Transvenous approaches can also be employed in the management of CCFs and have shown high success rates, since this method avoids navigation through arterial feeder vessels and instead occludes the fistula from a more approachable pathway via the venous system.^{9 14} It is currently unknown whether the response to treatment is equivocal between unilateral and bilateral CCFs,² although a

Patient's perspective

I originally went to the ER for headache and was only told that my blood pressure was a high. I never would have thought that my eyes would end up being affected by this. It was especially concerning when I saw multiple doctors for the red eyes but it still was not getting better. I am grateful for the expedited care I received from both eye doctors, and their help in arranging follow-up with the radiologists to look into my issue to identify the real cause.

Learning points

- ▶ Carotid-cavernous fistula (CCF) is an important diagnosis to consider in patients with unilateral or bilateral conjunctivitis that does not respond to traditional management.
- ▶ A thorough ophthalmologic examination is essential in the initial diagnosis of CCF as well as monitoring the patient's response to treatment.
- ▶ A multidisciplinary team approach is required for appropriate management of CCF. These complex cases require careful planning among the ophthalmologist, interventional neuroradiologist and neurosurgeon if needed in order to achieve optimal patient outcomes.

stepwise approach has been advocated for the latter in order to reduce the risk of haemorrhagic reperfusion.⁹ A diagnosis of CCF should be made clinically, and in conjunction with a referral to interventional neuroradiology for diagnostic imaging and treatment if necessary.

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