Case report

Bilateral sixth nerve palsies from carotid cavernous fistulas with transient worsening following transvenous embolisation

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Accepted 21 October 2019

SUMMARY

A 59-year-old man presented with a 2-week history of binocular horizontal diplopia worse when looking to the left. He was diagnosed with a left sixth nerve palsy (6 NP) and had new bilateral findings at a 2-week follow-up with new dilated episcleral blood vessels. Bilateral carotid-cavernous fistulas (CCFs) were suspected and confirmed with CT angiography and catheter angiography. He underwent successful coil and Onyx embolisation of the left cavernous sinus and immediately noticed worsening of the double vision when looking left due to a worse left 6 NP. Three weeks later, he underwent similar treatment of the right CCF and immediately noticed worsening of the double vision when looking to the right and was found to have a worse right 6 NP. His double vision resolved 6 months later. CCFs are rare causes of bilateral 6 NPs and although there is a good prognosis, symptoms may worsen after treatment.

BACKGROUND

A carotid-cavernous fistula (CCF) is an abnormal connection between the cavernous sinus and the carotid arteries or its branches, and may occur spontaneously or in the context of trauma. CCFs can be classified by Barrow classification according to the lesion that connects the cavernous sinus to the carotid artery. Type A CCFs have a direct high flow connection between the internal carotid artery and the sinus. Type B CCFs are dural shunts between the meningeal branches of the internal carotid artery and the sinus. Type C form from dural branches of the external carotid and type D involve meningeal branches from both the external and internal carotid and the cavernous sinus.¹ Unlike cranial nerves III–V which lie within the dural layers making up the wall of the cavernous sinus, the sixth cranial nerve lies unprotected within the sinus adjacent to the carotid artery. This close proximity to the carotid artery and lack of protection from the wall of the sinus make the abducens nerve vulnerable to compression or ischaemia from carotid artery pathology including CCFs. Bilateral sixth nerve palsies (6 NPs) from non-traumatic CCFs are rare and have been reported in eight previous cases.^{2–7} However, these cases included a patient with a posteriorly draining CCF without ocular or orbital signs,^{3 5} a patient with pronounced orbital signs,³ a bilateral 6 NP with other cranial nerve involvement⁸ or did not describe the treatment or clinical

course.² We present a unique course of sequential, bilateral 6 NPs, without significant orbital findings from bilateral CCFs. We also describe the treatment outcome with transient worsening of the 6 NP after transvenous embolisation.

CASE PRESENTATION

A 59-year-old man with a prior medical history of type 2 diabetes mellitus and dyslipidemia presented with a 2-week history of horizontal binocular diplopia worse when looking to the left. He was seen by the referring ophthalmologist who documented a left abduction deficit and diagnosed a left 6 NP, presumed microvascular ischaemic in origin given his vascular risk factors. A neuro-ophthalmology consultation was requested and he was seen 2 weeks after symptoms onset. At that time, he reported worsening double vision when looking to the right. On exam, he had normal afferent visual function, no proptosis, normal intraocular pressure and a normal dilated fundus examination. He was found to have bilateral abduction deficits (50% of normal in both eyes) and an esotropia measuring 35 prism diopters (PD) in primary position (*figure 1A*). This corresponded with binocular diplopia in all positions of gaze. There was no obvious conjunctival erythema, but when his eyelids were manually elevated, he was found to have corkscrew, dilated episcleral vessels in both eyes (*figure 2*). These findings were very suspicious for bilateral 6 NP secondary to CCFs.

Figure 1 (A) Eye movements at presentation demonstrated mild bilateral abduction deficits. (B) Eye movements 2 weeks after coiling of the left carotid-cavernous fistula (CCF) showed a worsened left abduction deficit. (C) Eye movements after endovascular coiling of the right CCF revealed a worsened right abduction deficit. (D) Six-month follow-up demonstrated full extraocular movements.

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To cite: Kalaichandran S, Micieli JA. *BMJ Case Rep* 2019;**12**:e232191. doi:10.1136/bcr-2019-232191

Figure 2 External photograph of both eyes shows dilated, corkscrew vessels suggestive of elevated episcleral venous pressure and a diagnosis of bilateral carotid-cavernous fistulas.

Investigations

An urgent CT angiography of the head was performed, which showed an enlarged left superior ophthalmic vein, an enlarged cavernous sinus region, left greater than right, and enlarged venous plexi along the dorsum sellae on the left ([figure 3](#)). These findings were highly suggestive of bilateral, left greater than right, CCFs. MRI brain and orbits was also performed and showed no additional findings. The patient underwent a cerebral angiogram that showed bilateral, left greater than right, CCFs with arterial feeders from bilateral ascending pharyngeal arteries, distal branches of the internal maxillary arteries bilaterally, branches from the left accessory middle meningeal artery and bilateral dural branches of the internal carotid arteries, classifying it as a Barrow type D ([figure 4A](#)). There was venous reflux into the left superior ophthalmic vein, but no cortical or deep venous reflux was detected.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis for bilateral 6 NPs includes trauma, raised intracranial pressure, compression from large skull base or cavernous sinus lesions, or brainstem lesions.⁷ Orbital lesions or myasthenia gravis should also be considered in these cases. Trauma was ruled out based on the patient's history and there were no signs of raised intracranial pressure such as papilledema on exam. MRI of the brain and orbits did not show any signs of compression along the course of the sixth cranial nerves or brainstem pathology. The CT angiogram and catheter angiogram confirmed the CCFs.

Figure 3 Axial CT angiography demonstrates prominent left greater than right cavernous sinuses and venous plexi along the dorsum sellae (red arrows) and a prominent left superior ophthalmic vein (yellow arrow).

Figure 4 (A) Lateral cerebral angiography with injection of contrast into the left external carotid artery showing filling of the cavernous sinus (red arrow) and the superior ophthalmic vein (black arrow). There were innumerable small arterial feeders identified from bilateral ascending pharyngeal arteries, distal branches of the internal maxillary arteries bilaterally, branches from the left accessory middle meningeal artery and bilateral dural branches of the internal carotid arteries (Barrow type D). (B) Magnified view postcoiling and Onyx embolisation demonstrating coils in the left cavernous sinus (white arrows). (C) Lateral cerebral angiography with injection of contrast into the left external carotid artery 2 weeks later demonstrating successful embolisation of the left carotid cavernous sinus with the coils (white arrow) positioned in the left cavernous sinus. (D) Anteroposterior view showing the final position of the coils in the cavernous sinuses.

TREATMENT

He underwent successful coil and Onyx embolisation of the left cavernous sinus with 21 coils ([figure 4B](#)). This led to angiographic resolution of the left CCF ([figure 4C](#)). He noticed worsening of the double vision when looking to the left immediately after the procedure. He underwent a repeat MRI/MRA with TRICKS protocol of the brain 2 weeks later, which showed a residual small right CCF and a treated left CCF. The patient attended a neuro-ophthalmology follow-up appointment 2 weeks after treatment and was found to have a larger esotropia in primary position (45 prism diopters) with a larger left abduction deficit compared with his initial presentation (10% of normal; [figure 1B](#)). The impression was that he had worsening of the left 6 NP postcoil embolisation. He underwent a second cerebral angiogram and coiling of the resident right CCF 3 weeks later with four coils and there was no angiographic evidence of residual right CCF after the procedure ([figure 4D](#)). He was seen in neuro-ophthalmology follow-up 3 weeks later and he reported persistent double vision and worsening of the double vision when looking to the right, which started immediately after the procedure. There was improvement of his left abduction deficit to 25% of normal and worsening of the right abduction deficit to 20% of normal ([figure 1C](#)). The impression was that he had a worsened right 6 NP postcoil embolisation and it was recommended that he occlude one eye for symptomatic relief.

OUTCOME AND FOLLOW-UP

He was seen in follow-up 1 month later and his bilateral abduction deficits improved and they completely resolved 6 months later. He had full extraocular eye movements and only a small residual esophoria of 2 PD in primary position at that time (figure 1D).

DISCUSSION

CCFs are a rare cause of bilateral 6 NPs and may occur spontaneously or in the context of trauma. Since the sixth nerve is in close proximity to the internal carotid artery in the cavernous sinus, it is vulnerable when CCFs develop. The mechanism is thought to involve direct compression or vascular steal with cranial nerve ischaemia.² Another potential mechanism of limited eye movements is venous congestion resulting in swelling of the extraocular muscles, but there was no obvious difference in the size of the lateral rectus muscle on imaging in this case, making it a less likely possibility.

Our case was unique in that the 6 NPs were sequential rather than simultaneous and our patient did not have significant orbital findings. The key to the diagnosis was the dilated, episcleral blood vessels, but this was only evident on examination if his eyelids were manually lifted due to his smaller palpebral fissures and dermatochalasis. The bilateral nature of his symptoms and examination findings also made non-arteritic microvascular ischaemia unlikely since this would require two independent ischaemic events. In general, bilateral findings should also prompt a more thorough workup including an MRI of the brain and orbits. The previous cases in the literature differed from ours as the patients had bilateral findings at onset,³ significant conjunctival hyperemia and chemosis at presentation⁴ or had additional cranial nerve involvement.⁸ While an additional case similarly had minimal orbital findings, this was owed to the location of the fistula in the posterior cavernous sinus with drainage to the petrosal sinus, avoiding effect to the orbits.³

Persistence or worsening of the 6 NP following endovascular coiling has been documented in the literature and is a known risk for the procedure.⁹ Potential explanations for the worsening include mass effect from the coils, which can compress and damage the nerve directly, chronic ischaemic caused by the coils or inflammation of the sixth nerve due to a thrombophilic nidus created by the coil mass.^{10–12} Since the worsening of the 6 NPs occurred immediately after the procedure in our case, it is more likely that a direct mass effect on the sixth cranial nerves was the explanation especially since the patient had spontaneous improvement with time. Some cases of 6 NPs after coil embolisation may be delayed and this can be better explained by thrombosis in the cavernous sinus, gradually worsening compression by the coils or a decompensated strabismus.^{10 12} Another possible explanation for a worsening of the 6 NP could be altered blood flow in the dorsal meningeal artery supplying the sixth cranial nerve. This could result in transient ischaemia and paresis. The use of coils has advantages including their radiopacity, ease of use and the ability to redeploy or remove devices as needed.¹³ There is literature to suggest that Onyx alone may be a viable treatment for some dural arteriovenous fistulas¹⁴; however, the study excluded CCFs and it still remains unknown whether this is a good treatment option for CCFs.

In conclusion, CCFs should be considered in the differential diagnosis of bilateral 6 NPs. A detailed examination to assess for

signs of this such as dilated episcleral vessels should be carried out in the context. Patients should understand that the 6 NP may worsen after treatment, but there is a good rate of spontaneous improvement as was seen in this patient who had resolution of double vision after 6 months.

Learning points

- ▶ Carotid-cavernous fistulas (CCFs) should be considered in the differential diagnosis of bilateral sixth nerve palsies (6 NPs).
- ▶ Key examination findings of anteriorly draining fistulas include dilated episcleral blood vessels, elevated intraocular pressure and a carotid bruit.
- ▶ Treatment of CCFs involves endovascular coil embolisation and this may result in transient worsening of the 6 NPs due to mass effect from the coil.

Contributors SK and JAM: conception and design, collection of data, drafting of the manuscript, critical analysis and final approval.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent for publication Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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