Asian J Ophthalmol. 2016;14:118-123

Embolization of dural carotid cavernous fistula via the superior ophthalmic vein approach

Chen Siang Nigel Lim[1], Hui-En Hazel Anne Lin[2], Kok Kee Tang[3], Cheng Kang Ong[4], Gangadhara Sundar[2]

[1]National University Hospital, National University Health System, Singapore; [2]Department of Ophthalmology, National University Hospital, National University Health System, Singapore; [3]KK Tang Adult and Paediatric Neurosurgery, Mount Elizabeth Hospital, Singapore; [4]Parkway Radiology, Mount Elizabeth Hospital, Singapore

Abstract: *In this case report, we present a patient with Type B dural carotid-cavernous fistula (CCF), who had failed cannulation via the transfemoral route and subsequently underwent CCF occlusion via the anterior orbital approach through the superior ophthalmic vein (SOV). Successful occlusion of CCF was achieved, with excellent visual and cosmetic outcomes postoperatively. When all venous routes have been exhausted, the SOV approach is an excellent and viable alternative in the treatment of dural CCFs. Close cooperation between the orbital and neuro-interventional teams in a hybrid operating theatre setting is essential in ensuring success of the operation.*

Key words: *Carotid cavernous sinus fistula, hybrid operating theatre, superior ophthalmic vein, cavernous sinus coiling*

Introduction

Carotid cavernous fistula (CCF) is an abnormal vascular shunt, which connects the intracranial carotid artery and cavernous sinus. With the advancement of liquid embolic agents[1] and metallic coils,[3] endovascular obliteration is now the treatment modality of choice for closure of CCFs. The transvenous approach is recommended for indirect CCF closure, and this is conventionally achieved transfemorally via the inferior petrosal sinus (IPS). If the IPS is inaccessible due to occlusion or lack of visualization, the cavernous sinus may be approached via the facial vein. However, this method is often unsuccessful due to tortuosity of the facial venous system. When all venous routes have been exhausted, surgical exposure and direct cannulation of the superior ophthalmic vein (SOV) is an excellent alternative to access the cavernous sinus, with good results and minimal complications reported in previous studies.[3] In this case report we present a patient with Type B dural CCF, who was treated successfully using the SOV approach.

Correspondence: Gangadhara Sundar DO, FRCS (Ed), FAMS, Head & Senior Consultant, Orbit & Oculofacial Surgery, Department of Ophthalmology, National University Hospital 1E, Kent Ridge Road, NUHS Tower Block, Level 7, Singapore 119228
E-mail: gsundar1@yahoo.com

Case report

A 47-year-old Malay lady with a past medical history of diabetes and hypertension presented with a six-month history of left eye prominence, associated with pain, swelling, tearing, retro-orbital headache and intermittent diplopia (Fig. 1).

Fig. 1. Mild proptosis (4 mm) and eyelid edema of the left eye at presentation.

A CT-scan (Fig. 2A) and MRI (Fig. 2B) revealed a dilated superior ophthalmic vein and enlarged extraocular muscles in the left eye. Preoperative intracranial angiogram showed a Type B dural CCF supplied by many fine feeder vessels from the internal carotid circulation (Fig. 3). Conservative management did not relieve her symptoms of pressure and discomfort behind the left eye and headaches even after six months. An attempted closure of her CCF via the transfemoral approach was unsuccessful resulting in referral to the Orbit and Oculofacial Service.

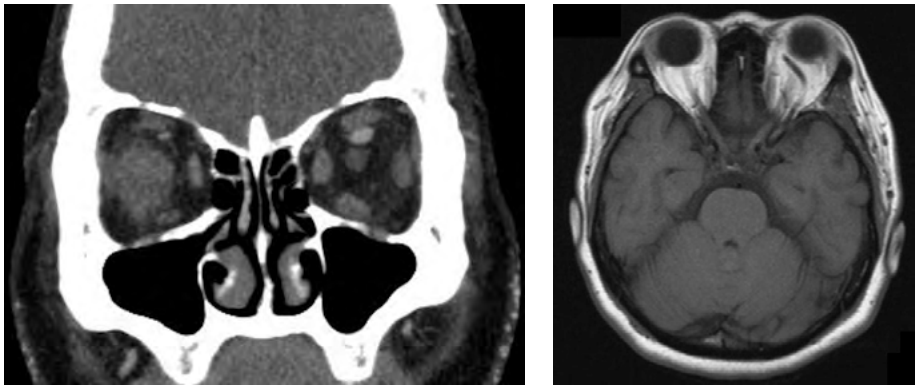

Fig. 2A(left) & 2B(right). Pre-operative imaging revealed a dilated superior ophthalmic vein and enlarged extra-ocular muscles in the left orbit. (2A) CT scan (2B) MRI (axial view).

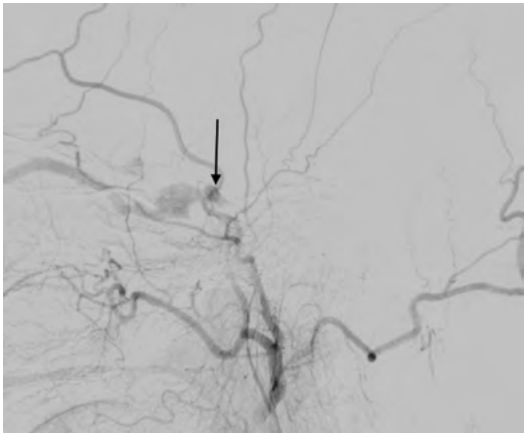

Fig. 3. Pre-operative intracranial angiogram showed a left dural CCF, supplied by many fine feeder vessels from the internal carotid circulation.

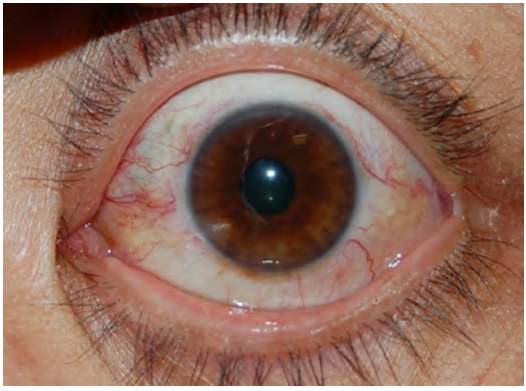

Fig. 4. Left eye demonstrating tortuous corkscrew-type conjunctival blood vessels

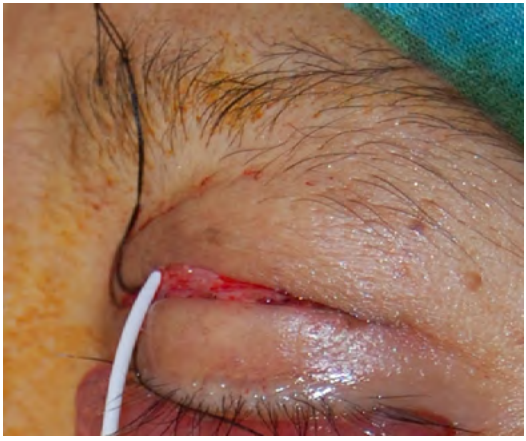

Fig. 5. Intra-operative photograph demonstrating the superior ophthalmic vein cannulation.

On examination, tortuous corkscrew-type blood vessels were seen in her left eye (Fig. 4). Best-corrected visual acuity (BCVA) was 6/6 bilaterally. Ocular motility was full in the right eye (OD), and there was mild global limitation in the left eye (OS). Pupillary reaction was sluggish, with a grade-2 relative afferent pupillary defect (RAPD) and proptosis of 4 mm in the left eye. Confrontational and static, automated visual field testing and color vision was normal. Intraocular pressure (IOP) was 14 mmHg (OD) and 21 mmHg (OS). Fundus examination was normal in the right eye, and showed dilated retinal veins in the left eye. After obtaining informed consent, she underwent a left anterior superior orbitotomy through an upper eyelid crease incision, in a Hybrid Operating Theatre under general anesthesia. The orbital septum was opened and dissection and exploration performed to identify the dilated SOV, which was then cannulated with a 4Fr Angiocatheter (Fig. 5). After confirmation of patency by intra-operative angiography, CCF obliteration was carried out using detachable HyperSoft neuro-coils (Microvention). Once adequate closure of the fistula was confirmed by angiography (Fig. 6A, 6B), the catheter was withdrawn and the wound closed. Postoperatively, she recovered well. BCVA was 6/6 bilaterally, with restoration of full ocular motility, no RAPD and normal color vision. In addition, IOP was normal and her proptosis resolved completely.

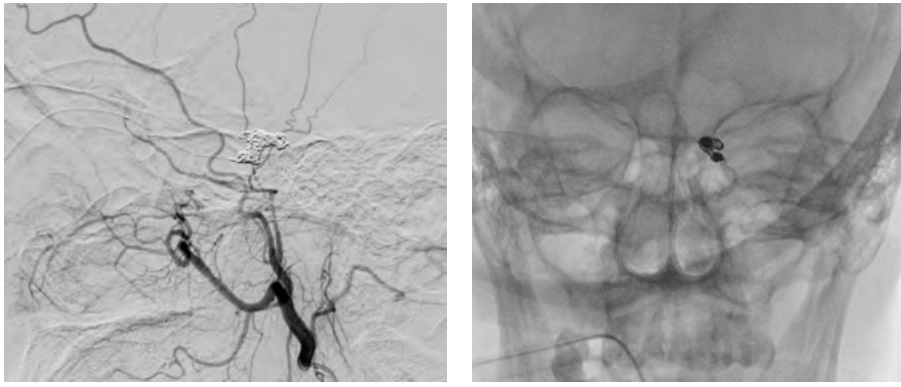

Fig. 6A(left) & 6B(right). Postoperative angiogram revealed successful obliteration of CCF by coiling (A) Lateral view; (B) Anterior view.

Discussion

CCFs may be anatomically and hemodynamically classified into four types based on the Barrow classification.[4] Type A CCFs are direct, high-flow lesions, which result from a direct connection between the intracavernous carotid artery and cavernous sinus. Type B, C and D CCFs are indirect, low-flow lesions that vary anatomically – the arterial supply of Type B lesions originate from meningeal branches of the internal carotid artery (ICA), Type C from meningeal branches of the external carotid artery (ECA), and Type D from meningeal branches of both the ICA and ECA.

Compared to direct CCFs, dural CCFs tend to have an insidious onset, with fewer and milder symptoms. Meyers *et al.*[5] noted that the commonest presenting signs and symptoms of dural CCFs were conjunctival injection (93%), chemosis (87%), proptosis (81%), diplopia (68%), cranial bruit (49%), retro-orbital headache (34%), raised IOP (34%) and decreased visual acuity (31%). Dural CCFs pose a major diagnostic dilemma and are often initially misdiagnosed as allergic or infective conjunctivitis. Accurate diagnosis is usually only made once the patient develops more severe symptoms such as visual loss or diplopia. This delay in diagnosis can be seen in our patient, who was misdiagnosed by several doctors and was unresponsive to conservative treatment initially. As dural CCFs may potentially lead to devastating complications such as stroke or intracranial hemorrhage secondary to retrograde cortical venous flow, all physicians should maintain a high index of suspicion in cases refractory to initial treatment. CT/MRI scans should be conducted to look for findings suggestive of dural CCF such as proptosis, expansion of the cavernous sinus, dilated SOV and enlargement of extraocular muscles. Digital subtraction angiography is used as the gold standard for diagnosis, and also to identify feeding vessels and drainage patterns.[5]

Although 20-60% of patients with dural CCF have spontaneous fistula closure and may be treated conservatively, endovascular obliteration is indicated in patients with progressive visual decline, refractory elevation of IOP, cortical venous drainage with neurological symptoms, and intractable headache/ocular pain.[6] As our patient experienced worsening symptoms and a decreased quality of life, she opted for surgical treatment. The conventional transfemoral approach via the IPS was carried out, but was unsuccessful due to occlusion of the IPS. As such, the patient underwent a surgical cut down approach with direct cannulation of the SOV. There were no intra-operative complications noted and the patient had excellent functional and cosmetic outcomes postoperatively. As interventional radiological and surgical techniques become more precise, with delivery of good outcomes with minimal morbidity and minimal invasiveness, such procedures may be considered in refractory cases of carotid-cavernous fistula even without disabling and severe proptosis and visual loss.

In most cases of carotid cavernous fistula, regardless of chronicity, the superior ophthalmic vein is dilated and patent. The superior ophthalmic vein is formed by the fusion of the supraorbital vein and the angular vein of the face, behind the trochlea of the superior oblique tendon. It then courses intraconally beneath the superior rectus-levator complex from medial to lateral and then enters the cavernous sinus through the annulus tendinus of Zinn.

We believe that our illustrated case report is unique in several ways. Unlike previously published case reports elsewhere where such patients had the initial surgical access performed in the operating theatre, then wheeled to the interventional suite under general anesthesia for intraluminal coiling before finally returning to the operating theatre for completion of the procedure, the availability of the Hybrid Operating Theatre with high-risk cardiac anesthesia support made this possible, highlighting a patient-centered approach rather than a systems-based approach.

Finally, all of this was possible because of the excellent understanding and cooperation between the various teams involved, common goal of patient's best interest, and the ability to perform a delicate and skillful procedure in a facility that permits the above with least inconvenience to the patient.

In conclusion, dural CCFs may potentially lead to severe visual dysfunction and should be diagnosed and treated promptly. When all venous routes have been exhausted, the transorbital approach via the superior ophthalmic vein remains an excellent and viable alternative to access the fistula. Close cooperation between the orbital, anesthetic and radiological teams is essential in ensuring success of the operation.

References

1. Suzuki S, Lee DW, Jahan R, Duckwiler GR, Viñuela F. Transvenous treatment of spontaneous dural carotid-cavernous fistulas using a combination of detachable coils and Onyx. AJNR Am J Neuroradiol 2006;27:1346-1349.
2. Luo CB, Teng MM, Lin CJ, Chang FC, Chang CY. Transarterial embolization of traumatic carotid-cavernous fistulae by Gugliemi detachable coils. A seven-year experience. Interv Neuroradiol 2008;14:5-8.
3. Lee JW, Kim DJ, Jung JY, Kim SH, Huh SK, Suh SH, Kim DI. Embolization of indirect carotid-cavernous sinus dural arterio-venous fistulae using the direct superior ophthalmic vein approach. Acta Neurochir (Wien) 2008;150:557-561.
4. Barrow DL, Spector RH, Braun IF, Landman JA, Tindall SC, Tindall GT. Classification and treatment of spontaneous carotid-cavernous sinus fistulas. J Neurosurg 1985;62(2):248.
5. 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.
6. Ellis JA, Goldstein H, Connolly ES Jr, Meyers PM. Carotid-cavernous fistulas. Neurosurg Focus 2012;32:E9.