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CASE REPORT

Staged curative treatment of a complex direct carotid-cavernous fistula with a large arterial defect and an 'oversized' internal carotid artery

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SUMMARY

This is a case of a high-flow, post-traumatic direct carotid-cavernous fistula with a widened arterial defect and a large-diameter internal carotid artery (ICA). The unique aspect of this case is the oversized ICA, >8mm in diameter, which is both a pathological and a therapeutic challenge, given the lack of available neuroendovascular devices for full vessel reconstruction. We present a planned two-stage embolisation paradigm for definitive treatment. Transarterial coil embolisation is performed as the first stage to disconnect the fistula and normalise flow in the ICA. A 3-month recovery period is then allowed for reduction in carotid diameter. Repair of the large vessel defect and pseudoaneurysm is performed as a second stage in a delayed fashion with a flow-diverting device. Follow-up angiography at 6 months demonstrates obliteration of the fistula and curative ICA reconstruction to a diameter <5mm.

BACKGROUND

Carotid-cavernous fistulae (CCFs) are pathological connections between the carotid artery and the cavernous sinus, which cause venous hypertension, present with proptosis and diplopia, and can lead to visual compromise and intracranial haemorrhage. A direct CCF (Barrow type A) is a high-flow fistula that results from a tear in the cavernous internal carotid artery (ICA) and is often acquired from craniofacial trauma. This contrasts with indirect CCF, Barrow types B–D, which are supplied by dural branches of the ICA, external carotid artery (ECA) or both. The high-flow state results in hypertrophy of the ICA with increased vessel diameter, particularly if the fistula is unrecognised and untreated for a period of time. Both coil embolisation (transarterial or transvenous) and flow diversion are known techniques for endovascular management of direct CCFs.[1–4] However, if the ICA defect is large, then coiling alone is often insufficient for complete vessel reconstruction and healing. Likewise, if the parent vessel is significantly hypertrophied, then the vessel diameter can be too big ('oversized') for proper implantation and wall apposition of a flow-diverting stent. We present a case of delayed presentation of a direct CCF with both large vessel defect and a significantly oversized ICA, representing a therapeutic challenge.

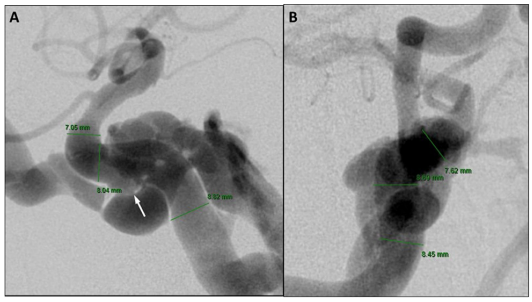

Figure 1 Initial diagnostic cerebral angiogram. (A) Lateral and (B) Anterior-posterior digital subtraction angiography demonstrating a high-flow Barrow type A carotid-cavernous fistula. There is a prominent vessel wall defect (white arrow) on the inferior aspect of the horizontal segment of the cavernous internal carotid artery (ICA). The ICA is hypertrophied, with proximal vessel diameter >8 mm.

CASE PRESENTATION

The patient is a 25-year-old man who initially presented to an outside institution after being ejected from a motorcycle following collision with a truck. He was wearing a helmet, but it broke on impact. He was treated for polytrauma, including right-sided facial fractures, at the outside institution. He subsequently presented to an ophthalmologist at our institution 4 months after the accident with chief complaint of progressive horizontal double vision over a 1-month period. He was diagnosed with a right cranial nerve (CN) VI palsy and mild ipsilateral exophthalmos. Non-contrast CT showed proptosis and a dilated right superior ophthalmic vein suggestive of CCF, confirmed by arterial phase filling of the cavernous sinus on computerised tomography angiography (CTA). Cerebral angiography, which is necessary to plan treatment, further characterised the fistula as robust, high-flow Barrow type A fistula with a large defect approximately 7 mm in length on the inferior wall of the horizontal cavernous ICA and a grossly hypertrophied ICA with diameter >8 mm proximal to the defect (figure 1).

TREATMENT

Initial treatment was directed at closing the fistula connection by transarterial coil embolisation of the cavernous sinus (figure 2). This was accomplished by a transfemoral approach with a triaxial system consisting of a NeuroMax guide catheter

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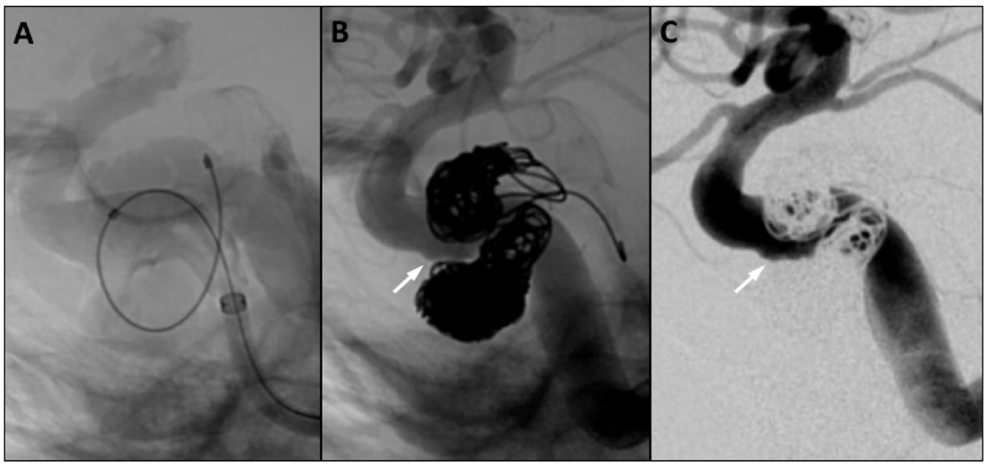

Figure 2 Stage I: transarterial coil embolisation. (A) Lateral native view demonstrating the Navien 072 catheter in the proximal cavernous internal carotid artery and the Echelon 10 microcatheter tracked through the arterial wall defect and tip positioned in the cavernous sinus. Postcoiling (B) native and (C) digital subtraction angiography lateral views demonstrating occlusion of the carotid-cavernous fistula. The arterial defect and residual pseudoaneurysm (white arrow) are clearly visualised.

(Penumbra), Navien 072 distal intracranial catheter (Medtronic) and Echelon 10 microcatheter (Medtronic). A total of 13 coils (2.21 m total length) were deployed with successful disconnection of the fistula; however, the arterial defect of the cavernous ICA remained present with a prominent pseudoaneurysm. After 3 months of recovery, the ICA diameter had reduced to <5 mm in size, and the persistent pseudoaneurysm was treated with placement of a single 4.0 mm diameter by 18 mm length Pipeline device (figure 3). This was performed by a transfemoral approach with a triaxial system consisting of a NeuroMax guide catheter, AXS Catalyst 5 distal intracranial catheter (Stryker) and Via 27 microcatheter catheter (MicroVention).

OUTCOME AND FOLLOW-UP

Follow-up angiography 6 months after Pipeline implantation demonstrated continued occlusion of the fistula as well as complete remodelling of the ICA defect (figure 4). Clinically, his diplopia and exophthalmos had resolved and gaze was conjugate at this time.

DISCUSSION

This case report describes a planned two-stage embolisation paradigm for curative treatment of a complex, direct CCF with a large cavernous ICA defect and an oversized parent vessel diameter. Although coil embolisation and flow diversion are described therapeutic options for the treatment of direct CCFs resulting from trauma or aneurysm, neither were appropriate stand-alone strategies for long-term cure of this patient given the large-diameter ICA on presentation. In appropriate cases, multiple flow diverters can be telescoped to achieve high metal density across the arterial defect in a single-modality cure for direct CCF.[5 6] Flow diversion can also be used for recurrence or contemporaneously with either transvenous or transarterial coiling.[1–4 7]

The sizeable cavernous ICA defect of 7 mm in this case allowed for easy transarterial access to the cavernous sinus for coil embolisation to arrest flow through the fistula. The greatest risk in transarterial coil embolisation of a CCF is coil herniation into the parent vessel, which could cause stroke. In this case,

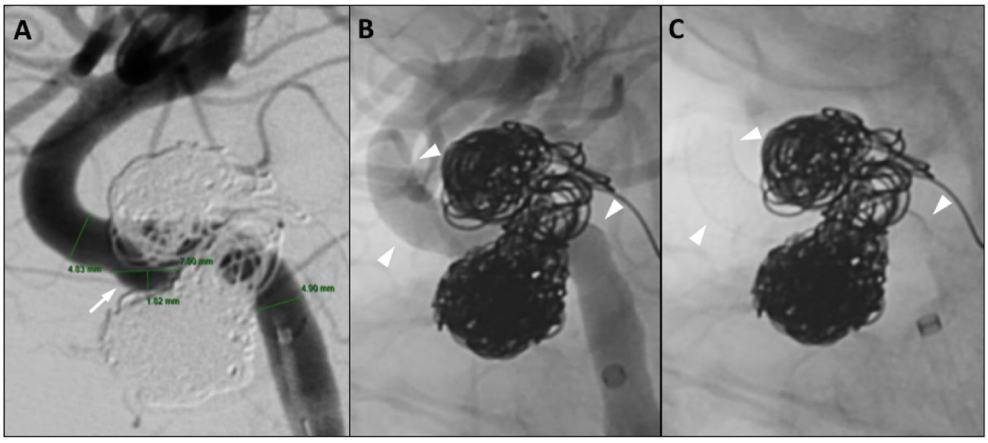

Figure 3 Stage II: pipeline embolisation of pseudoaneurysm. (A) Pre-embolisation lateral digital subtraction angiography demonstrating significant reduction in the diameter of the cavernous internal carotid artery, now <5 mm. The pseudoaneurysm is also well visualised with a neck of 7 mm. (B,C) Native lateral views post-PED implantation (white arrows). (B) Immediate postimplantation angiography already demonstrates decreased filling of the pseudoaneurysm.

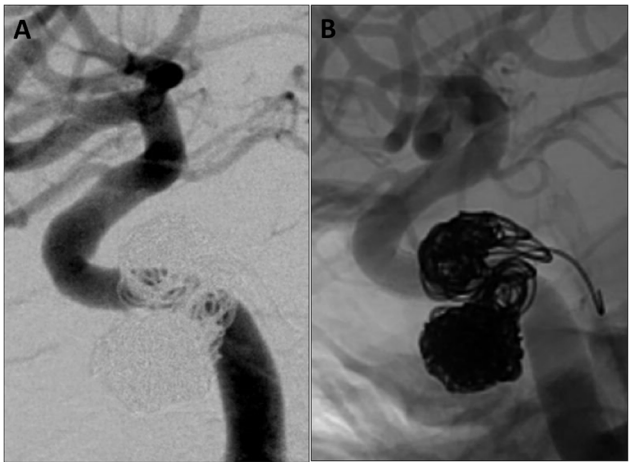

Figure 4 Six-month follow-up angiography. Lateral (A) digital subtraction angiography and (B) native angiography views 6 months afterpipeline embolisation device (PED) implantation demonstrate complete occlusion of the carotid-cavernous fistula and pseudoaneurysm with curative reconstruction of the cavernous internal carotid artery.

ICA diameter >8 mm excluded coil packing up to the wall of the ICA secondary to lack of an appropriately sized NeuroBalloon or traditional vascular reconstruction device to protect against this risk of coil herniation. With a wire in the cavernous sinus, a balloon could have been tracked into or adjacent to the ICA defect for purposes of remodelling and prevention of coil prolapse into the ICA. However, inflation of a balloon in such a position would have potentially risked widening the arterial defect and was not used.

The large ICA diameter also excluded initial treatment by flow diversion because the only FDA-approved flow diverter, the PED, has a maximum available diameter of 5.0 mm. A 5.0 mm diameter PED can be ballooned to approximately 5.25 mm, but this is still significantly undersized for this patient. Were a larger flow diverting stent available, treatment would still have been deferred because stenting an 8 mm ICA while eliminating the CCF as a pressure sink might lead to excessive intracranial blood pressures that would stress and potentially overwhelm the brain's autoregulatory capabilities. Following closure of the fistula with coils, the patient was arbitrarily given 3 months to recover, which proved sufficient for the ICA to decrease in diameter to <5 mm. The patient was then a candidate for stage 2 treatment of the ICA pseudoaneurysm using a PED, which was performed without incident. At 6-month follow-up, angiography demonstrated curative reconstruction of the cavernous ICA.

While long-term follow-up of CCF treated with flow diverters has not been published, other series with average follow-up >4 years have shown that coil embolisation provides durable closure of CCF without recurrence of acute symptoms.[8] For this reason, some interventionalists may have opted to forgo the second-stage PED embolisation in favour of monitoring the pseudoaneurysm. Pseudoaneurysms are common following direct CCF repair, occurring in 30%–90% of patients depending on the series, and they can either remain stable or enlarge to cause recanalisation of the CCF or cranial nerve compression.[9 10] Treatment with a flow diverter required 6 months of clopidogrel and a lifetime of low-dose aspirin, the risk of which we and the patient accepted to attempt a cure of his pseudoaneurysm given its size and the patient's young age.

Learning points

- A high-flow direct carotid-cavernous fistula (CCF) with a widened arterial defect and a large-diameter internal carotid artery (ICA) is an endovascular therapeutic challenge secondary to the lack of available neurospecific devices for reconstruction of this vessel type.
- Such a CCF can be definitively treated with staged embolisations.
- The first stage consists of coil embolisation (transarterial or transvenous) to close the fistula.
- A recovery period is then allowed for normalisation of the ICA diameter. The second stage consists of curative reconstruction of the ICA wall by flow diversion.
- CCF may present after craniofacial trauma with delayed vision loss, conjunctival chemosis, external ophthalmoplegia and proptosis.

Contributors GPC conceived the manuscript. GPC and MTB performed data acquisition. All authors contributed to data analysis, drafting of the manuscript and critical analysis of the manuscript. All authors approved the final version of the manuscript.

Competing interests GPC: Participates in clinical trials for Medtronic, Stryker and MicroVention. Consultant for MicroVention. ALC: Participates in clinical trials for Medtronic and Microvention. Consultant and proctor for Medtronic, Stryker and MicroVention. LML: Participates in clinical trials for Medtronic and Stryker. Proctor for Medtronic.

Patient consent Obtained.

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