CASE REPORT

Posterior fossa dural arteriovenous fistula presenting clinically as a carotid–cavernous fistula treated by a direct access cavernous sinus approach

Ai Peng Tan, Manish Taneja, Francis Hui

Department of Neuroradiology,
National Neuroscience
Institute, Singapore

Correspondence to

Dr Ai Peng Tan,
National Neuroscience
Institute, 11 Jalan Tan Tock
Seng, Singapore 308433;
tan_ai_peng@hotmail.com

SUMMARY

Dural arteriovenous fistulas (dAVFs) represent approximately 10–15% of all cerebral vascular malformations. Although dAVFs can occur anywhere in the brain, they occur most frequently in the cavernous and transverse-sigmoid sinuses. Posterior fossa dAVFs presenting clinically as carotid–cavernous fistulae (CCF) are rarely encountered in clinical practice. We discuss and illustrate an unusual case of a left posterior fossa dAVF that presented clinically with chemosis and early visual impairment, similar to that of CCF. This was subsequently treated by a direct access cavernous sinus approach. We describe the technique used to access the cavernous sinus directly in cases where conventional transvenous and transarterial routes have been exhausted.

did not reveal pathological findings in the cup:disc ratio. The visual field was also preserved. No other focal neurological deficit was detected. There was also no evidence of raised intracranial pressure.

INVESTIGATIONS

MRI of the brain revealed prominent vessels within the subarachnoid space of the left posterior fossa. A diagnostic angiogram confirmed the presence of a posterior fossa dAVF supplied by the neuromeningeal branch of the left ascending pharyngeal and meningeal branches of the left vertebral arteries. The venous drainage pattern was along the paraclival veins into the cavernous sinus and superior ophthalmic vein (figure 1A,B). The superior

BACKGROUND

Dural arteriovenous fistulas (dAVFs) represent approximately 10–15% of all cerebral vascular malformations. Although dAVFs can occur anywhere in the brain, they occur most frequently in the cavernous and transverse-sigmoid sinuses.^{1–3} We present an unusual case of a left posterior fossa dAVF that presented clinically with chemosis and early visual impairment similar to that of CCF, which was treated by a direct access cavernous sinus approach.

This case is important because posterior fossa dAVFs presenting clinically with venous congestive symptoms similar to that of carotid–cavernous fistulae (CCF) are rare. In addition, this case is unique because the posterior fossa dAVF was treated by a direct access cavernous sinus approach as all conventional transvenous and transarterial routes had been exhausted. The direct access cavernous sinus approach has been described for the treatment of CCF but, in this case, it was used as an access route to treat the left posterior fossa dAVF.

CASE PRESENTATION

A patient presented with a 3-month history of left eye redness and swelling which progressively worsened over a 2-week period. There was no history of prior head injury. The patient was not on any medications and had no significant family history. A comprehensive review of the systems was also non-contributory.

Clinical examination of the left eye revealed orbital bruit with dilated conjunctival vessels and mild exophthalmos. Extraocular muscle motility was preserved apart from a mild limitation in left eye adduction. Visual acuity was 4/6 in the left eye and 6/6 in the right eye. Ophthalmoscopic examination

Figure 1 Digital subtraction angiography of (A) the left vertebral artery and (B) the ascending pharyngeal artery confirm the presence of a dural arteriovenous fistula in the left posterior fossa, draining anteriorly into the cavernous sinus and superior ophthalmic veins, contributing to the ophthalmic congestive symptoms.

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ophthalmic vein was occluded in its anterior portion, in the mid part of the left orbit, while the left inferior ophthalmic vein (IOV) was completely occluded. No other superficial or deep venous drainage was seen.

DIFFERENTIAL DIAGNOSIS

The initial clinical impression was that of a left CCF, although the onset of symptoms was more insidious which was unusual for CCF. Prominent vessels seen within the subarachnoid space of the left posterior fossa on MRI of the brain raises the possibility of either a posterior fossa dAVF or previous cavernous sinus thrombosis. On diagnostic angiography, these prominent vessels were found to be related to a posterior fossa dAVF with arterial feeders from the neuromeningeal branch of the left ascending pharyngeal and meningeal branches of the left vertebral arteries.

TREATMENT

Two attempts at transarterial embolization using Onyx had been performed at two other institutions over the preceding 2 months. Adequate nidal penetration could not be achieved and the fistula persisted through the left vertebral artery side branches in its V3 segment. The patient was referred to us for endovascular treatment of the posterior fossa dAVF. Repeat angiography confirmed the presence of feeders from the left vertebral and ascending pharyngeal arteries with drainage into the cavernous sinus and superior ophthalmic vein. The superior ophthalmic vein was occluded in its anterior portion while the left IOV was completely occluded, contributing to the orbital congestion. The vertebral artery pedicles were too small and not suitable for transarterial embolization using n-butyl-2-cyanoacrylate (NBCA) or Onyx. In the absence of direct transarterial and transvenous access, the decision was made to proceed with direct access into the cavernous sinus (figure 2).

General anesthesia was administered and access to the right common femoral artery was obtained. A 4 F vertebral catheter was advanced into the left vertebral artery and an angiogram was performed. Using fluoroscopic guidance and roadmap angiographic technique, a micropuncture needle was introduced through the floor of the orbit and direct access into the

cavernous sinus was obtained. On frank aspiration of venous blood, contrast was injected to verify the position of the cavernous sinus. Care was taken to keep a safe distance from the cavernous segment of the internal carotid artery. Over a 0.018 inch Terumo guidewire, a 3 F micropuncture dilator was advanced into the venous system. Subsequently, a 0.018 inch Mandril guidewire was introduced and a 4 F micropuncture sheath/dilator system was advanced into the cavernous sinus. Through this, a SL-10 preshaped 45° microcatheter and Synchro preshaped guide wire (Stryker, Freemont, USA) were used to go through the paraclival draining vein to the point of the fistula. The fistula was then occluded using multiple target coils. Coils were also deployed into the entire length of the draining vein up to the mid part of the ipsilateral cavernous sinus. The access site into the cavernous sinus was occluded using 33% NBCA that was injected during withdrawal of the 4 F sheath. Minor pressure was applied to the access site enabling complete hemostasis. Post-embolization angiography showed complete occlusion of the dAVF (figure 3A, B).

Figure 2 The arrow denotes the access site of the 4 F sheath which was introduced into the left cavernous sinus through the floor of the left orbit.

Figure 3 Embolization of the fistulous point was performed with deposition of detachable platinum coils via a direct cavernous sinus approach. Post-embolization angiograms of (A) the left vertebral artery and (B) the ascending pharyngeal artery show complete occlusion of the left posterior fossa dural arteriovenous fistula.

OUTCOME AND FOLLOW-UP

The patient did extremely well on follow-up and the congestive symptoms of the left eye improved dramatically within a week of the intervention.

DISCUSSION

Intracranial dAVFs can occur anywhere within the dura mater.^{1–3} Signs of pulsating exophthalmos, conjunctival chemosis, motility restriction and reduced visual acuity are usually attributed to recruitment of venous drainage from either the superior or inferior ophthalmic veins.⁴ Subarachnoid hemorrhage may result if venous drainage is primarily through cortical veins or brainstem edema and cranial neuropathies if venous drainage is through the basilar plexus.⁵ It is now generally accepted that the venous drainage pattern is the most predictive factor of aggressive dAVF symptoms.⁶ The most common symptoms of CCF are ocular symptoms caused by anterior venous drainage.⁷ Rarely, ocular symptoms can occur in other types of non-CCF dAVF. Several types of non-CCF dAVFs have been associated with ocular symptoms and these include posterior fossa dAVFs draining via the inferior petrosal sinus and cavernous sinus to the ophthalmic vein,⁴ as in our case.

Recent developments in catheter interventions since the introduction of Onyx allow most patients to be cured with transcatheter embolization.² The aim of treatment is the obliteration of the fistulous point, approached from either the arterial or venous side. Occasionally, as in our case, the tributaries of the ophthalmic veins fail to provide access for treatment. Feeding artery embolization of external carotid branches with particles is easily performed and can reduce shunt flow. However, complete cure is difficult to achieve with this method because of the existence of feeding arteries that cannot be catheterized and the recruitment of a blood supply from collateral arteries.²

This case presented unique challenges to the interventional neuroradiologist as no conventional transarterial or transvenous routes were available. Direct IOV access under ultrasound guidance was also attempted without success. A direct percutaneous puncture of the IOV was shown to be a safe alternative route for CCF access and embolization when the inferior petrosal sinus and superior ophthalmic vein are inaccessible.⁸ As more conventional transvenous and transarterial routes had been exhausted, a direct cavernous sinus approach was attempted with the hope of achieving catheterization of the fistulous connection. Direct cavernous sinus access is well described in the literature although, to our knowledge, all the cases described were for the treatment of CCF.⁹

Our case was unique in that the cavernous sinus was used as access for the treatment of a posterior fossa dAVF. We also found NBCA to be a useful agent for tract occlusion. We conclude that, in challenging cases where conventional transvenous and transarterial routes have been exhausted, a direct cavernous

sinus approach can potentially be used for treatment of complex neurovascular disease.

Learning points

- Posterior fossa dural arteriovenous fistulas (dAVFs) presenting clinically as a carotid-cavernous fistula (CCF) are rarely encountered in clinical practice.
- The most common symptoms of CCF are ocular symptoms caused by anterior venous drainage. Rarely, ocular symptoms can occur in other types of non-CCF dAVFs such as posterior fossa dAVFs draining via the inferior petrosal sinus and cavernous sinus to the ophthalmic vein.
- The direct cavernous sinus approach can potentially be used for the treatment of complex neurovascular disease where conventional transvenous and transarterial routes have been exhausted.

Contributors APT: drafting of manuscript, revision of manuscript. MT, FH: conception of case and image interpretation, review of manuscript.

Competing interests None.

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