The Journal of Laryngology and Otology
August 1999, Vol. **113**, pp. 744–746

Spontaneous carotico-cavernous fistula presenting as pulsatile tinnitus

A. ROBERTSON, B.Sc., M.B., CH.B., A. R. NICOLAIDES, F.R.C.S., F.R.C.S. (ORL), R. H. TAYLOR, F.R.C.S., F.R.C.OPHTH*

Abstract
A patient with sudden onset pulsatile tinnitus resulting from a spontaneous carotico-cavernous fistula is presented. The case is discussed and illustrated to highlight the clinical features and natural history of a condition rarely seen by ENT surgeons.

Key words: Tinnitus; Intracranial arteriovenous malformations

Introduction

Carotico-cavernous fistulas may occur in a number of clinical settings both spontaneously and following trauma. The ocular manifestations are well described but tinnitus is rarely the main presenting feature of this condition.

Case report

A previously healthy 67-year-old lady presented to the ENT department with a four-week history of sudden onset right-sided pulsatile tinnitus. She described this as a persistent drumming sensation, which in association with some mild earache was causing her considerable discomfort. In addition she complained of double vision. She had no significant medical history, was a non-smoker and was taking no medication.

On examination she had entirely normal tympanic membranes and there were no bruits over the scalp, eyes or neck. Pure tone and impedance audiometry were normal. It was noted, however, that she had a mildly engorged right eye with contralateral (left) lateral rectus palsy. Subsequent ophthalmology review showed an esotropia for distance fixation and raised intra-ocular pressures. Her blood pressure was elevated at 170/90 and she was commenced on aspirin and atenolol. Arrangement was made for magnetic resonance imaging (MRI).

In the intervening four months the patient's tinnitus and diplopia improved. However, her right eye had now developed proptosis with markedly engorged conjunctival vessels of corkscrew-like appearance and chemosis (Figure 1a/1b).

FIG. 1a

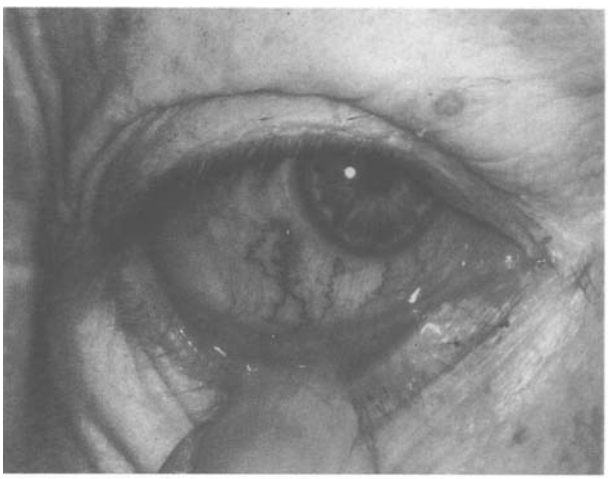

FIG. 1b

FIG. 1
(a) Proptosis of the right eye with swelling of the eyelids. (b) Corkscrew-like conjunctival vessels.

From the Departments of Otolaryngology and Ophthalmology*, York District Hospital, York, UK.
Accepted for publication: 30 March 1999.

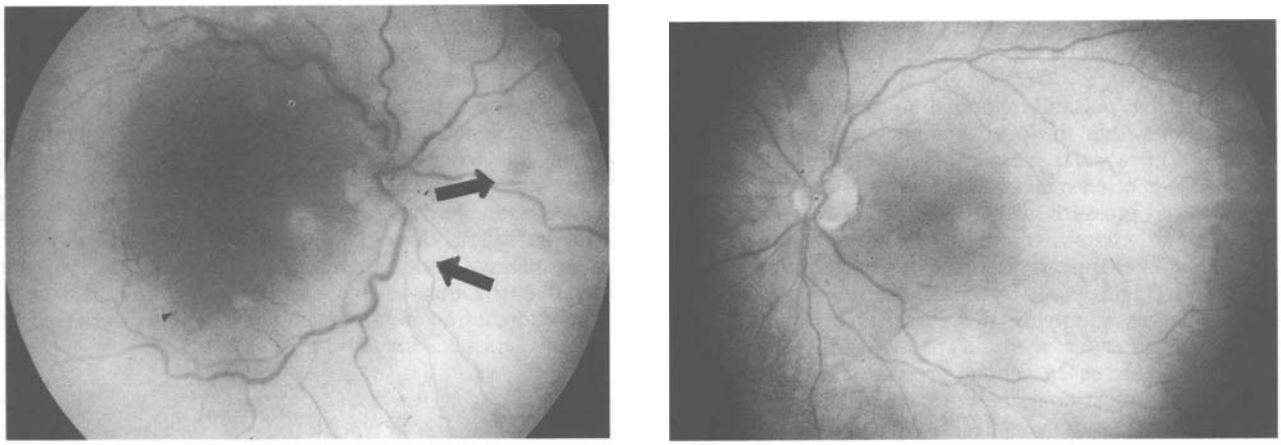

FIG. 2
Normal left retina/Right retinal vein dilatation with retinal haemorrhages (arrows).

She had tortuous vessels in the right retina with associated haemorrhages (Figure 2). MRI of the skull base and magnetic resonance angiography (MRA) of the internal carotids performed at that time revealed no abnormality.

Computerized tomography (CT) scan, with special reference to the orbital and sellar regions, however, showed right axial proptosis with enlarged extra-ocular muscles and superior ophthalmic vein distension. These findings were compatible with the diagnosis of a carotico-cavernous fistula (Figure 3).

Cerebral angiography failed to demonstrate an arterio-venous connection but there was poor filling of the left and no filling of the right cavernous sinus during the venous phase. This suggested thrombosis with resolution of the fistula (Figure 4).

When last reviewed eight months following the onset of symptoms there was still some residual corneal vessel engorgement. All other symptoms and signs including the pulsatile tinnitus had completely resolved.

Discussion

Pulsatile tinnitus is sometimes found in association with conductive deafness or hypertension. Other rarer causes include vascular tumours and carotid artery aneurysms. This case demonstrates a very unusual cause for this symptom namely a carotico-cavernous fistula. Symptoms result from an abnormal communication between the carotid circulation and the cavernous sinus leading to arteriovenous mixing, increased venous pressure and associated changes in the rate and direction of blood flow.

Barrow *et al.* (1985) proposed a four-type classification based on the anatomy of the lesion. Type A fistulas are direct shunts between the internal carotid artery and the cavernous sinus. They usually have a high flow rate and when spontaneous are thought to arise from intra-cavernous carotid artery aneurysm rupture. Because of their relatively high blood flow rates these fistulae tend to produce acute, severe symptoms.

Type B, C and D fistulas are indirect shunts between the meningeal branches of the internal, external or both carotid arteries and the cavernous sinus respectively. Their aetiology is debated. They are thought to arise secondary to rupture of the dural arteries that transverse the cavernous sinus. Other suggested mechanisms include lateral sinus thrombosis with recanalization and the opening up of congenital arteriovenous malformations. High-risk groups of patients include older women, hypertensives, mothers during childbirth and those with atherosclerosis or collagen vascular disease (Barrow *et al.*, 1985). Indirect fistulas tend to have low flow rates and therefore often present more insidiously.

This case beautifully illustrates the clinical features of a low flow indirect carotico-cavernous fistula. Classic features described include pulsatile exophthalmos and conjunctival chemosis. Cranial bruits are also a feature of this

FIG. 3
Right exophthalmos with diffuse enlargement of the extra-ocular muscles.

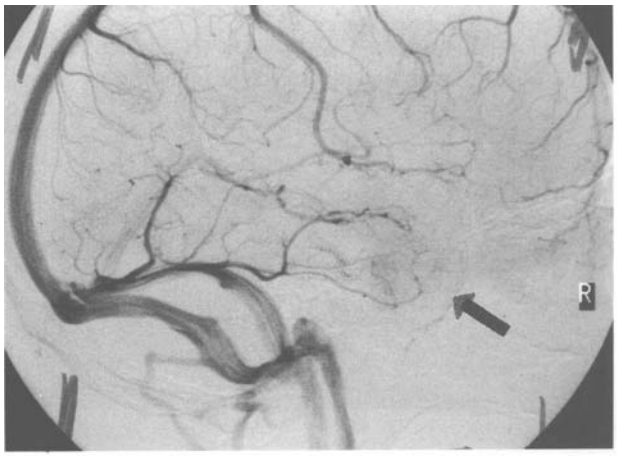

FIG. 4
Right cerebral angiogram showing poor filling of the cavernous sinus (arrow).

https://doi.org/10.1017/S0022215100145086 Published online by Cambridge University Press

condition, however pulsatile tinnitus is rarely the presenting complaint (Morrison, 1989; Jungreis, 1991; Roy *et al.*, 1993).

Progressive pulsatile exopthalmos results from venous congestion within the orbit and may be associated with increased intraocular pressure and swelling of the eyelids. Headache is often mild or absent.

Increased pressure in the conjunctival vessels produces conjunctival chemosis and arterialized vessels with a corkscrew-like appearance, one of the hallmarks of this condition (Figure 1b). Diplopia is often present as the motor nerves supplying the extrinsic eye muscles are damaged either as a result of ischaemia, aneurysmal pressure or increased venous pressure. The sixth cranial nerve is the most commonly affected due to its position within the cavernous sinus. Signs may be ipsilateral, bilateral or contralateral as in the case described due to the anatomical connection between the two cavernous sinuses. Orbital oedema and ocular muscle engorgement may also contribute.

Typical fundoscopic findings include ipsilateral swelling of the optic disc, retinal vein dilatation and intra-retinal haemorrhages (Figure 2). Rarely, pre-retinal or vitreous haemorrhages may be found (Biousse *et al.*, 1998).

Additional evidence for the diagnosis includes CT scanning or MRI that may typically reveal prominence of the superior ophthalmic veins, diffuse enlargement of the extraocular muscles and proptosis (Figure 3). Pseudoaneurysm of the cavernous sinus may also be present (Keltner *et al.*, 1987). Orbital ultrasonography with Doppler, not employed in this case, has also been used to confirm the diagnosis, monitor progression and distinguish between low and high flow rates (Spector, 1991).

Cerebral angiography with selective cannulation of the internal and external carotid arteries and vertebral circulation is helpful to delineate the anatomy and determine flow rates. Spontaneous closure of the defect has been noted following this procedure.

Management strategies vary depending on fistula flow rates and the severity of symptoms. Indirect, low flow fistulas such as the one described here have a tendency for spontaneous resolution in up to 60 per cent of cases, due to complete or partial thrombosis of the cavernous sinus or its tributaries. They are, therefore, initially treated conservatively. Visual loss can occur in up to 30 per cent of patients (Biousse *et al.*, 1998). Intervention may be required in cases of visual deterioration, obtrusive diplopia, intolerable bruit and 'malignant' proptosis (Barrow *et al.*, 1985).

The aim of intervention is selective closure of the fistula without interruption of the internal carotid circulation that may lead to cerebral infarction. Direct surgical approaches include craniotomy with complete excision of the fistula. Newer approaches include selective embolization of the defect using a variety of materials by either an arterial or venous approach or by balloon occlusion. Secondary glaucoma and ischaemic retinopathy may require additional treatment.

High flow fistulas rarely resolve spontaneously and may produce significant morbidity and mortality. Progressive visual loss occurs in up to 90 per cent of cases due to exposure keratopathy, increased intraocular pressure, vitreous haemorrhage, retinal vein stasis, central retinal vein occlusion, choroidal detachment and ischaemic optic neuropathy. They, therefore, usually require prompt treatment. Other serious complications such as venous infarction, intracranial and subarachnoid haemorrhage and massive epistaxis may also arise.

Conclusion

Although carotico-cavernous fistulas are relatively rarely encountered in otolaryngology the diagnosis should be borne in mind in patients presenting with pulsatile tinnitus with associated ophthalmological abnormalities particularly if they are middle-aged, female and hypertensive.

References

Barrow, D. L., Spector, R. H., Braun, I. F., Landman, J. A., Tindall, S. C., Tindall, G. L. (1985) Classification and treatment of spontaneous carotid-cavernous fistulas. *Journal of Neurosurgery* **62:** 248–256.

Biousse, V., Mendicino, M. E., Simon, D. J., Newman, N. J. (1998) The ophthalmology of intracranial vascular abnormalities. *American Journal of Ophthalmology* **125(4):** 527–544.

Jungreis, C. A. (1991) Pulsatile tinnitus from a dural arteriovenous fistula. *Annals of Otology, Rhinology and Laryngology* **100:** 951–953.

Keltner, J. L., Satterfield, D., Dublin, A. B., Lee, B. C. P. (1987) Dural and carotid cavernous sinus fistulas: diagnosis, management, and complications. *Ophthalmology* **94:** 1585–1600.

Morrison, G. A. J. (1989) Pulsatile tinnitus and dural arteriovenous malformation. *Journal of Laryngology and Otology* **103:** 1073–1075.

Roy, D., Lavigne, F., Raymond, J. (1993) Pulsatile tinnitus and dural arteriovenous fistula of the transverse sinus. *Journal of Otolaryngology* **22(6):** 409–412.

Spector, R. H. (1991) Echographic diagnosis of dural carotico-cavernous sinus fistulas. *American Journal of Ophthalmology* **111:** 77–83.

Address for correspondence:
Mr A. R. Nicolaides,
Department of Otolaryngology/Head and Neck Surgery,
York District Hospital,
Wigginton Road,
York YO3 7HE.