# Indirect carotid cavernous fistula presenting as pulsatile tinnitus

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#### Abstract

This paper reports a case of spontaneous indirect carotid cavernous fistula that presented with pulsatile tinnitus, left-sided temporal headache and left-sided ptosis. The pulsatile tinnitus, its aetiology and investigation are discussed. The importance of pulsatile tinnitus is highlighted, with a discussion of carotid-cavernous fistulas.

This case illustrates that clinically silent cavernous sinus thrombosis can give rise to spontaneous indirect carotid cavernous fistula. Magnetic resonance imaging angiography was used in diagnosis. Treatment ranges from observation, as in our case, to transvenous endovascular techniques.

# Key words: Carotid Artery, Internal; Fistula; Tinnitus

# Introduction

Carotid cavernous fistula is an abnormal arterio-venous communication between the internal carotid arterial system and cavernous sinus. It maybe either spontaneous or traumatic with spontaneous cases usually occurring in the older age group whereas the traumatic form usually occurs in younger age groups. It usually affects the anterior tributaries of the cavernous sinus resulting in exophthalmos, proptosis, ophthalmoplegia, abducent nerve palsy and episcleritis. Less commonly it affects the posterior tributaries of the cavernous sinus and its dural attachments and thus clinically becomes apparent as pulsatile tinnitus and temporal headache.<sup>1</sup>

### **Case report**

A 63-year-old female patient presented to the ENT department with a history of pulsatile tinnitus of sudden onset, left-sided temporal headaches, left retro-orbital pain and no vertigo. ENT, opthalmological and neurological examination revealed only a left-sided ptosis. Past history of note included a right-sided transphenoidal hypophysectomy in 1974. Investigations including pure tone and evoked response audiometry were unremarkable. Full-blood count, urea and electrolytes, ESR, protein S, protein C, anti-thrombin III and anti-cardiolipin were normal.

Magnetic resonance imaging angiography with dual echo axials, T1 sagittals and T1 coronals revealed a degree of cerebral atrophy. There was no sign of exophthalmos and there was nothing to suggest a dilated superior ophthalmic vein. There was no sign of aneurysm. However, there was excess vascularity in the posterior aspect of the left cavernous sinus, the left jugular vein was more prominent than the right and there was prominence of the inferior petrosal sinus. These findings suggested an indirect carotid cavernous sinus fistula arising from the so-called basal angioma that resulted from a thrombosis of the cavernous sinus, as illustrated in Figure 1. The patient settled with no active treatment with resolution of her tinnitus and headache over a period of four to six weeks.

# Discussion

Pulsatile tinnitus is a perception of a rhythmical sound that is synchronous with the heartbeat and is rarely seen in daily practice.<sup>2</sup> The aetiologies are arterio-venous malformations, dissection of internal carotid arteries, fibromuscular dysplasia, intracranial or extracranial aneurysm, carotid-cavernous fistulas, glomus tumours intracranial hypertension,<sup>3</sup> a high or dehiscent jugular bulb and persistent stapedial artery.<sup>2</sup>



Fig. 1

Magnetic resonance imaging angiography revealing an indirect carotid-cavernous fistula (arrow).

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Pulsatile tinnitus can be evident from otoscopic examination, that may reveal a retrotympanic mass as in glomus tumours, a high or dehiscent jugular bulb, or a persistent stapedial artery. Other aetiologies may present with a normal tympanic membrane and pulsatile tinnitus. Some such as arteriovenous shunts and carotid artery stenosis, that are life-threatening should be excluded.

Full ear, nose and throat examination, otoscopy, palpation and auscultation of ears, cervical region and orbits is required.<sup>4</sup> Neurological and ophthalmological referrals are useful. Non-invasive techniques such as ultrasonic examination of cervical vessels and computed tomography (CT) for glomus tumours<sup>5</sup> are advocated firstly. If they are negative or do not clarify abnormal findings, then invasive techniques such as magnetic resonance angiography should be employed.<sup>3</sup>

Hence, patients with pulsatile tinnitus and a normal otoscopic examination should undergo magnetic resonance angiography that can yield significant pathology and can be used as a screening tool in such cases.<sup>2</sup>

Barrow et al.<sup>6</sup> classified four categories of carotid cavernous fistulas. Type A (direct) are direct shunts between the internal carotid artery and the cavernous sinus. Type B (indirect) are between the meningeal branches of the carotid artery and the cavernous sinus. Type C (indirect) are shunts between the meningeal branches of the external carotid artery and the cavernous sinus. Type D (indirect) are fistulas between the meningeal branches of both the internal carotid artery, external carotid artery and the cavernous sinus. Type A is almost always a high flow fistula, traumatic and occurs predominantly in males.<sup>7</sup> Types B, C and D (indirect) are typically spontaneous. The patient described above has an indirect type fistula. The aetiology includes trauma, spontaneous, Ehlers-Danlos syndrome, rupture of an aneurysm and thrombosis.<sup>8</sup>

Anatomically the cavernous sinus is 2 cm long and 1 cm wide, lying on each side of the sphenoid body. The internal carotid artery and the abducent nerve pass through the sinus. The most important anterior tributary is the superior ophthalmic vein; posteriorly, the tributaries are the superior petrosal sinus and inferior petrosal sinuses.<sup>9</sup>

Investigations included are intraocular pressure measurement, B-type ultrasonography, CT and MRI scanning. Sometimes carotid angiography is required if surgical intervention is likely. Treatment ranges from no treatment to surgical intervention in the form of insertion of a detachable balloon using the trans-arterial route in traumatic carotid cavernous fistulas.<sup>10</sup> In spontaneous carotid fistula no treatment may be required,<sup>11</sup> however, external carotid artery occlusion or trans-venous embolization techniques may be used.<sup>12</sup> Carotid jugular compression therapy has been advocated by Higashida *et al.*<sup>13</sup> The risks of active intervention include stroke, in two to four per cent of cases.<sup>8</sup>

#### Conclusion

A case of indirect carotid cavernous fistula has been presented, resulting from cavernous sinus thrombosis

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affecting the posterior tributaries of the cavernous sinus, leading to a sudden onset of pulsatile tinnitus and headaches. This represents an unusual clinical picture. The pulsatile tinnitus, causes and the investigations have been described, with a detailed discussion of carotid cavernous fistulas and their treatment.

#### References

- Jorgenson JS, Gutthoff RF. Twenty-four cases of carotidcavernous fistulas: Frequency, symptoms, diagnosis and treatment. Acta Ophthalmol 1985;63(suppl 173):67–71
- 2 Sanchez TG, Santoro PP, Torres De Medeiros IR, Bittar RS. Magnetic resonance angiography in pulsatile tinnitus. *Int Tinnitus J* 1998;**4**:122–6
- 3 Waldvogel D, Mattle HP, Sturzenegger M, Schroth G. Pulsatile tinnitus – a review of 84 patients. J Neurol 1998;245:137–42
- 4 Tiefert JW. Orbital auscultation. Am Fam Phys 1978;18:117-20
- 5 Reiss M, Reiss G. Pulsating tinnitus. Wien Klin Wochenschr 2000;112:84–91 (German)
- 6 Barrow DL, Spector RH, Braun IF, Landman JA, Tindall SC, Tindall GT. Classification and treatment of spontaneous carotid-cavernous sinus fistulas. J Neurosurg 1985;62:248-56
- 7 Jacobson BE, Nesbit MD, Ahuja A, Barnwell SL. Traumatic indirect carotid-cavernous fistula: Report of two cases. *Neurosurgery* 1996;**39**:1235–7
- 8 Barnwell SJ, O'Neil OR. Endovascular therapy of carotidcavernous fistulas. *Neurosurg Clin North Am* 1994;**5**:485–93
- 9 Gray H. Angiology. In Williams P, Warwick R, Dyson L, Bannister L, eds. *Gray's Anatomy*, 37th edn. Broadway, New York: Longman Group Ltd, 1989;802–3
- 10 Debrun GM, Lacour P, Vinuela F, Fox A, Drake CG, Caron JP. Treatment of 54 traumatic carotid-cavernous fistulas. J Neurosurg 1981;55:678–92
- 11 Phelps CD, Thompson AS, Ossoinig KC. The diagnosis and prognosis of atypical carotid-cavernous fistulas. Am J Ophthalmol 1982;93:423–36
- 12 Vinuela F, Fox AJ, Debrun GM, Peerless SJ, Drake CG. Spontaneous carotid-cavernous fistulas: clinical, radiological and therapeutic considerations. Experience with 20 cases. J Neurol 1984;60:976–84
- 13 Higoshida RT, Halbach VV, Tsai FY, Norman D, Pribram HF, Mehringer CM, et al. Interventional neurovascular treatment of traumatic carotid and vertebral artery lesions: Results in 234 cases. Am J Roentgenol 1989;153:577–82

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