The Journal of Laryngology & Otology
February 2004, Vol. **118,** pp. 153–155

Jugulotympanic paraganglioma (glomus tumour) presenting with recurrent epistaxis

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Abstract
A case is presented where a left jugulotympanic paraganglioma (JTP) extended to the nasopharynx and the patient presented with recurrent epistaxis. Although initial biopsy of an aural polyp had been suggestive of the diagnosis several years previously, the diagnosis was not confirmed until the patient presented with recurrent epistaxis and severe anaemia. To the best of our knowledge, this is the first case reported of such a presentation of JTP.

Key words: Glomus Jugulare; Epistaxis

Introduction

Jugulotympanic paragangliomas are benign vascular tumours arising from the region of the jugular foramen at the skull base. The term jugulotympanic paraganglioma (JTP) has largely replaced the old term, glomus jugulare, as the cell of origin has been shown to be the chief cells of paraganglionic tissue rather than the pericytes in the glomus complexes as was once believed.[1] The three main types of jugular foramen paragangliomas are: 1) tympanic, which mainly originates from the middle ear, 2) jugular, which originates in the jugular foramen, and 3) vagal, which is located below the skull base. In large tumours, however, it is almost impossible to detect the actual origin of the tumour.[2]

Although JTPs are not encountered frequently, they represent the most common neoplasm of the middle ear and are second in frequency only to vestibular schwannomas within the temporal bone.[3] Clinically, the condition should be suspected if the patient complains of pulsatile tinnitus and otoscopic examination may be suggestive of the diagnosis. Optimal imaging with computerized tomography (CT) and magnetic resonance imaging (MRI) often displays characteristic appearances of the neoplasm and allows differentiation from vascular anomalies such as a high jugular bulb or an aberrant carotid artery.[4]

We present a case of JTP that was neglected by the patient until severe anaemia caused by recurrent epistaxis persuaded her to be admitted and investigated. The cause of epistaxis was found to be a nasopharyngeal extension of the paraganglioma.

Case report

A 50-year-old housewife had presented with a history of left pulsatile tinnitus 16 years previously. The rate of pulsation of her tinnitus increased with exercise and emotional stress. There was a noticeable hearing loss at that time and examination showed a red polyp and purulent discharge from the ear. A biopsy of the polyp was performed at another institution and this was followed by brisk bleeding, which was controlled with packing of the ear. The histopathology of the removed tissue showed an ulcerated epithelium, numerous vascular channels and a stroma infiltrated with fibroblasts and acute and chronic inflammatory cells (Figure 1). There was no histological evidence of malignancy, and reviewing the available material could not provide conclusive evidence of a glomus tumour.

The patient was lost to follow-up until the first of November 2001, when she presented to our institution complaining of recurrent anterior and postnasal epistaxis. Examination of the right ear showed no abnormality. However, the left ear canal was filled by a red polyp with a smooth surface and surrounded by an inoffensive mucopurulent discharge. Tuning fork tests were suggestive of a total loss of hearing in the left ear, and this was confirmed by pure tone audiometry. Endoscopy of the nose and nasopharynx revealed a red soft tissue mass in the left side

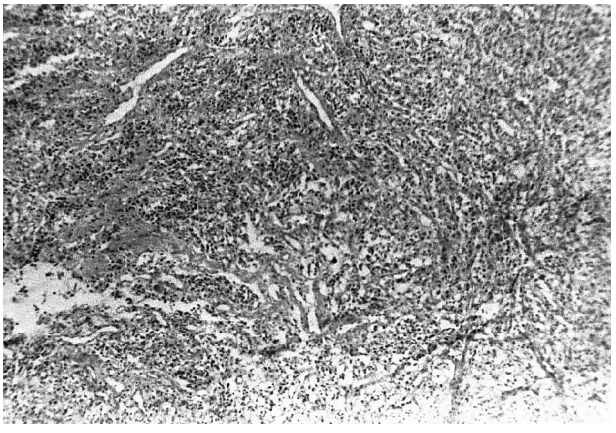

Fig. 1
Histopathology of aural polyp revealing vascular channels in inflammatory cellular infiltrate (H & E; ×40)

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Accepted for publication: 14 November 2003.

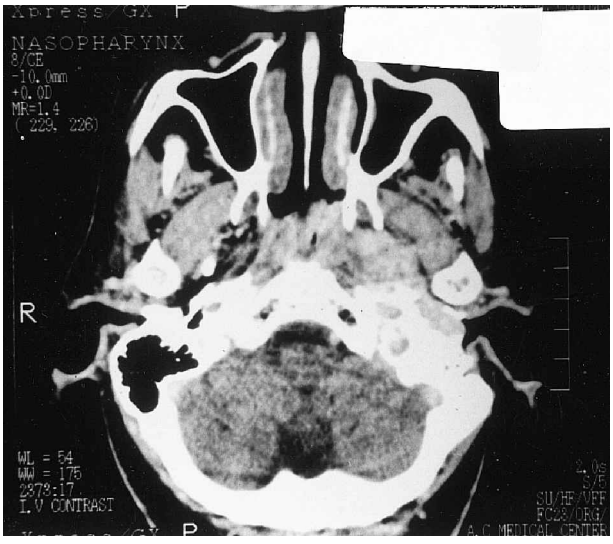

FIG. 2
Axial CT scan showing destruction of left temporal bone.

of the nasopharynx approximately 1.5 × 2 cm, arising from the left lateral nasopharyngeal wall. The mass partly occluded the left posterior choana, and its surface showed evidence of fresh bleeding. Fibre-optic laryngoscopy revealed a normal larynx. Cranial nerve examination revealed no deficits apart from cranial nerve VIII deficit. Examination of the neck revealed no swelling and auscultation over the mastoid revealed no bruits. Haematological investigation showed a haemoglobin concentration of 6.5 gm%; bleeding and clotting times were normal and serum biochemistry showed normal values.

- **This is a case report of a nasopharyngeal extension of a glomus jugulare presenting with epistaxis**
- **This association has not been reported previously**

A contrast-enhanced CT scan showed a 4 × 2 cm mass involving the left petrous temporal bone, with adjacent bone destruction and involvement of the left pterygopalatine fossa (Figure 2). The mass was seen to protrude into the nasopharynx and oropharynx.

An MRI scan showed a hyperintense mass filling the left jugular fossa with displacement of left levator and tensor palati muscles below the skull base to the contralateral side and extension to the nasopharyngeal cavity at the left fossa of Rosenmuller (Figure 3). Also evident, was destruction of the left inner ear and encroachment of the tumour through the foramen ovale intracranially. The nasopharyngeal component was shown by MRI to be the most vascular area of the tumour. MR angiography showed increased vascularization of the branches of the left maxillary artery. The patient received five units of blood and she had the left external carotid artery ligated under general anaesthesia. Subsequently, she underwent a full course of therapeutic external radiotherapy to the lesion with no serious after-effects. At the last follow-up on 15 June 2003, she reported no complaints apart from the hearing loss in the left ear. Examination of the left ear showed a pale residual mucosal swelling emanating from a dry tympanic membrane perforation. The nasopharyngeal swelling resolved completely. Her haemoglobin concentration was 11.8 gm%.

Discussion

Jugulotympanic paragangliomas are quite uncommon. It is estimated that about 1 in 30 000 head and neck tumours is a paraganglioma.[5] Although reports of malignant tumours have been described, most paragangliomas are benign, slow-growing vascular tumours. Histologically, the tumour cells (called chief cells or Zellballen) contain vesicles that store catecholamines.[6] Although all paragangliomas have neurosecretory granules, only one to three per cent of the tumours are considered functional.

The most common symptom of JTPs is pulsatile tinnitus. Their effect on the mobility of the tympanic membrane and ossicles results in a conductive hearing impairment.[7] However, because of their slow growth, they may not be detected until they are quite large; thus causing symptoms such as bleeding from the ear, otalgia, vertigo, facial paralysis and hoarseness. As they grow, they tend to extend along paths of least resistance. Thus, they may extend to the nasopharynx along the eustachian tube, to the petrous apex along peritubal cells, or along the foramina of the skull base and vascular channels to spread intracranially. They may also cause significant destruction of the temporal bone.[4] Optimum imaging allows differentiation from vascular anomalies (such as a dehiscent high jugular bulb) and it is essential for staging of the tumour.[3] CT scanning demonstrates the amount of bone destruction and reveals the relation of the tumour to large vessels, whereas MRI is more useful for characterization of the lesion. T2-weighted MR images often display the characteristic salt and pepper appearance typical of paraganglioma (Figure 3), whereas T1-weighted MR images display the actual vascularity of the tumour. Magnetic resonance angiography (MRA) can determine vascular involvement and the directionality of blood flow. Selective angiography is used to discover the blood supply of these tumours, and in some instances, permits embolization of large vascular tumours.

The management of JTPs consists of either surgery or radiotherapy.[8] Surgical removal is the only effective means of achieving a complete cure, however, the risks of surgery are significant and must be evaluated over the natural progression of growth of a benign, slow growing tumour.[9] Radiotherapy has no effect on the actual tumour cells. It acts by inducing a vascular thrombosis inside the tumour.[10]

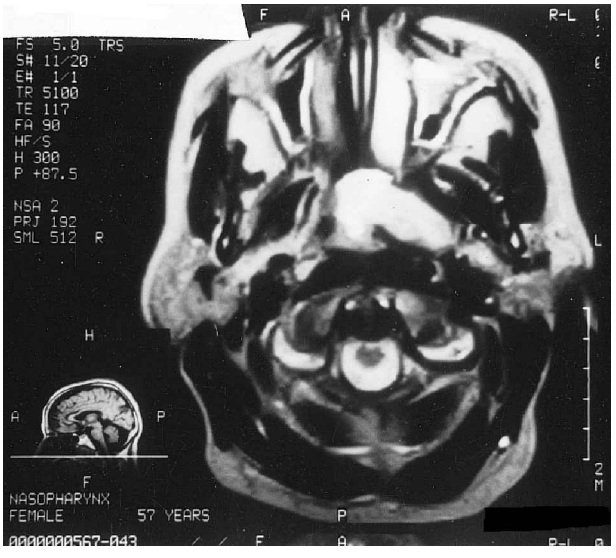

FIG. 3
T2-weighted MRI scan showing salt and pepper appearance of tumour and extension into nasopharynx.

https://doi.org/10.1258/002221504772784658 Published online by Cambridge University Press

It is especially useful for the treatment of elderly symptomatic patients, or patients who are unwilling to undergo the risks, complications and sequelae of large JTP surgery. Recent trends in the therapy of JTPs include stereotactic radiosurgery[8] and the administration of the radiolabelled somatostatin analogue ^{111}indium octreotide.[11]

Conclusion

A high index of suspicion of JTPs is necessary in patients with pulsatile tinnitus. Adequate radiological evaluation of the tumour is facilitated by recent radiographic techniques. We have presented a case of an advanced JTP with an extraordinary symptom. To the best of our knowledge, this is the first case of JTP presenting with recurrent epistaxis.

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Dr K. G. Effat takes responsibility for the integrity of the content of the paper.
Competing interests: None declared

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https://doi.org/10.1258/002221504772784658 Published online by Cambridge University Press