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Glomus jugulare tumour with metastases to cervical lymph nodes

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Abstract
Glomus jugulare tumours are classically described as benign tumours with a long time course often measured in decades. Although these tumours may be locally invasive, most cases are histologically benign and metastases are rare. The case of a malignant glomus jugulare tumour with a particularly aggressive pattern of spread is presented. At the time of surgery, which was within 12 months of the development of symptoms, intracranial spread and metastasis to cervical lymph nodes had already occurred, demonstrating that glomus jugulare tumours are not always benign.

Key words: Glomus jugulare tumour; Neoplasm metastasis

Introduction

Glomus jugulare tumours are paragangliomas arising from paraganglionic tissue in the jugular bulb. They are the commonest benign tumours of the temporal bone and present with a variety of symptoms according to the local structures involved.[1] They have been classified as types A to D, as shown in Table I.[2] The growth of glomus jugulare tumours is typically slow, with one case reported as surviving untreated for 60 years.[3] Local invasiveness is common but malignancy is rare.[4]

Metastasis from glomus jugulare tumours, which defines the presence of malignancy, is rare with a reported incidence of between one and four per cent.[1,5,6] This is likely to be an overestimate due to preferential reporting of cases with metastasis and due to the occurrence of multicentric glomus tumours.[7] We present the case of a particularly aggressive malignant Fisch type D glomus jugulare tumour with cervical lymph node metastases detected at the time of surgery.

Case history

A 34-year-old man was referred to the Department of Otoneurosurgery, Addenbrooke's Hospital, Cambridge with a 12-month history of right-sided pulsatile tinnitus,

TABLE I
CLASSIFICATION OF GLOMUS JUGULARE TUMOURS[2]

Type A	Tumours localized to the middle ear cleft (glomus tympanicum)
Type B	Tumours limited to the tympanomastoid area without destruction of bone in the infralabyrinthine compartment of the temporal bone
Type C	Tumours extending and destroying bone of the infralabyrinthine and apical compartment of the temporal bone
Type D	Tumours with intracranial extension

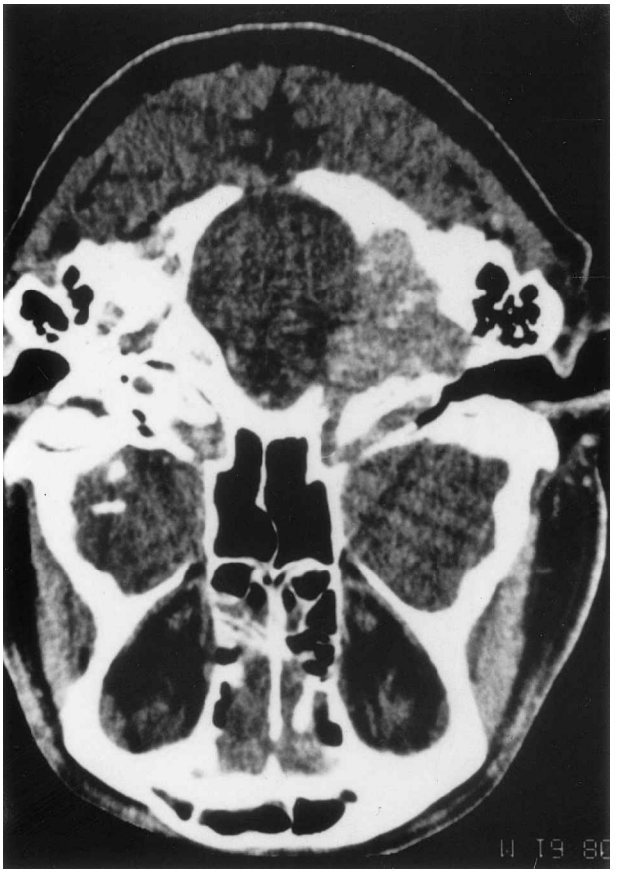

Fig. 1
Axial CT scan of the head showing the large glomus jugulare tumour in the right petrous temporal bone extending into the posterior cranial fossa.

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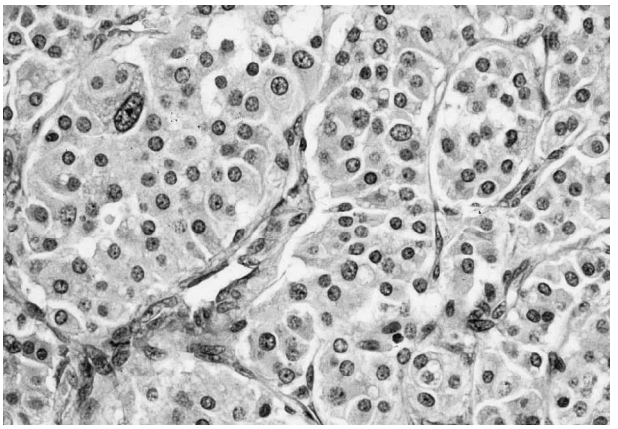
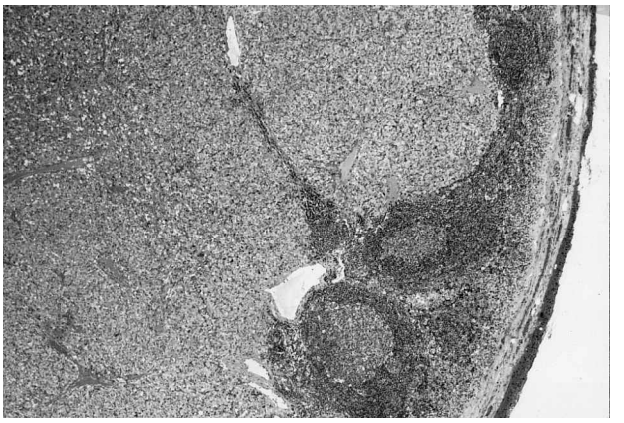


(a)



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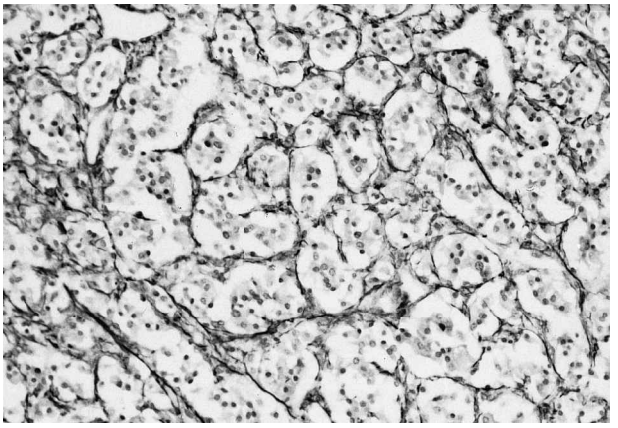
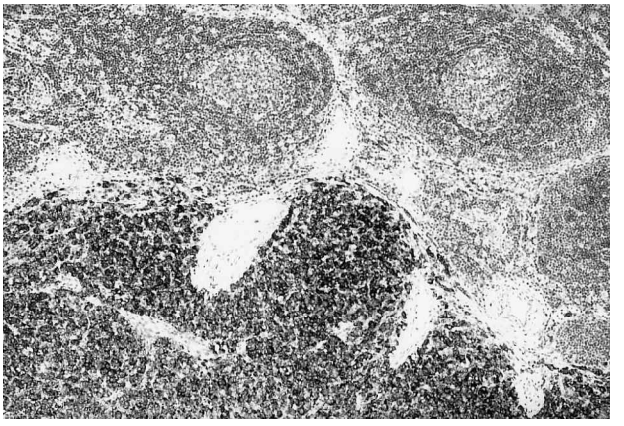


(b)



(d)

FIG. 2

a. Higher power view of tumour showing a packeted architecture. The tumour cells have moderately pleomorphic nuclei with granular chromatin and small nucleoli. No mitoses are seen in this field (H & E; × 370).
b. Medium power view of a reticulin preparation demonstrates the packeted architecture of the paraganglioma. (Reticulin; × 180).
c. Low power view of metastatic deposit within a lymph node. Residual lymphoid follicles are seen around the edge of the pale mass of tumour tissue (H & E; × 18).
d. Low power view of lymph node showing deposit of metastatic tumour, darkly stained by immunohistochemistry for synaptophysin. Surrounding lymph node tissue, in which several lymphoid follicles are seen, is negative for synaptophysin, and is palely stained with the counterstain (synaptophysin immunohistochemistry, H & E; × 14).

hearing loss and headaches. There were no other symptoms and there was no past medical history of note. Otoscopy revealed a red, pulsatile mass behind the right tympanic membrane. Examination, including examination of cranial nerves and balance, was otherwise unremarkable. A pure tone audiogram showed a conductive hearing loss of 30 dB in the right ear.

A high-resolution computed tomographic (CT) scan and a magnetic resonance imaging (MRI) scan suggested a very large right-sided glomus jugulare tumour with intracranial involvement (Fisch type D), as shown in Figure 1. A carotid arteriogram suggested a very large tumour extending from the skull base to below the level of the carotid bifurcation with blood supply from the external carotid, internal carotid and vertebral arteries. Chest radiograph and abdominal CT scan were normal.

In view of the size of the tumour and the patient's relative youth it was decided to treat the tumour with surgery rather than radiotherapy. The tumour was embolized two days pre-operatively. Excision of the tumour was performed via an infra- and trans-temporal route.[8] At operation the tumour was found to be extending below the level of the carotid bifurcation, and to be extensively invading the skull base, lateral venous sinus, cerebellopontine angle, posterior fossa dura and cochlea. The facial nerve was transposed anteriorly. Three cervical lymph nodes inferior to the tumour were suspected to be involved and were excised. A lumbar drain was inserted.

Post-operative palsies of cranial nerves VII, IX, XI and XII were noted. The patient was slow to recover consciousness and a repeat CT scan two days post-operatively showed swelling of the right cerebellar hemisphere with compression and displacement of the fourth ventricle and hydrocephalus. A frontal external ventricular drain was inserted with good effect.

Histopathological examination of the specimen, as shown in Figure 2, revealed fibrofatty tissue extensively infiltrated by tumour composed of cells with round-to-ovoid, moderately pleomorphic nuclei, granular chromatin and small nucleoli. Mitoses were not conspicuous. The cells had abundant eosinophilic cytoplasm and formed a packeted architecture with well-defined groups of cells (Zellballen) separated by fibrovascular stroma, including vascular channels. Focally the stroma appeared hyalinized. Tumour necrosis was not a feature. There was an infiltrative pattern of spread with tumour invading into, but not through, the dura mater. In one region, poorly circumscribed nodules of tumour encased a segment of muscular artery. The artery showed an acute arteritis with infiltration of its walls by polymorphonuclear neutrophils

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and with surrounding organizing haemorrhage, indicating vascular damage, but direct tumour invasion was not seen. Metastatic deposits of tumour were identified in several of the local lymph nodes. Immunohistochemical studies showed the tumour cells to be positive for synaptophysin, neurone specific enolase, PGP9.5, S100 and focally for chromogranin. Immunohistochemistry to cytokeratins with an antibody to a pan-cytokeratin and with CAM 5.2 was negative. The appearances were those of a paraganglioma (glomus tumour) with an agressive pattern of spread and metastases to regional lymph nodes.

In view of the histology, post-operative radiotherapy was given. Four months post-operatively his facial nerve function had recovered to House grade III, he had compensated well for the palsies of cranial nerves IX, XI and XII and his balance was good.

Discussion

Most glomus jugulare tumours are histologically benign, although they may be locally invasive. Prediction of malignant behaviour on histological appearances alone is difficult, although features such as increased mitoses, necrosis, vascular invasion and decreased expression of neuropeptides have been suggested as markers of potential aggression.[9] The only absolute criteria of malignancy remain local and distant metastases. In this case, increased mitotic activity and necrosis were not features, but the tumour did show evidence of local infiltration and vascular damage. Malignancy was established by the presence of metastases in several local lymph nodes.

A review on the subject of glomus jugulare tumours with metastases is presented elsewhere in this issue.[10] This review suggests that these tumours are a more aggressive subset of glomus jugulare tumours with a shorter duration of symptoms before diagnosis and higher rates of persistent or recurrent local disease and death. The case reported here was a particularly aggressive tumour with metastases diagnosed at the time of surgery and within a year of the development of symptoms. This report demonstrates that glomus jugulare tumours are not always benign.

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