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Online Only Clinical Record

A case of paraganglioma of the hypoglossal nerve

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Abstract

Introduction: Paragangliomas are rare tumours arising from the paraganglia of the autonomic nervous system.

Case report: We present a case of a paraganglioma arising from the hypoglossal nerve and producing an unusual clinical picture at presentation.

Discussion: We supply radiological evidence of a paraganglioma originating from the hypoglossal nerve, and thus extend the evidence base for this rare site of origin. Our patient presented as an emergency with long tract neurological symptoms and progressive brainstem involvement. This presentation is not characteristic of paragangliomas in general, which usually have an indolent growth pattern and often demonstrate benign symptoms for a number of years prior to diagnosis. The location of a hypoglossal paraganglioma differs significantly from more common paragangliomas described in the neck and skull base, and this should inform the surgical approach undertaken.

Key words: Paraganglioma; Hypoglossal Nerve; Surgical Procedures

Introduction

Paragangliomas have been described at numerous sites within the head and neck, the most frequent type being carotid paragangliomas ('carotid body tumours'). Jugular, tympanic and vagal paragangliomas are also well recognised,[1,2] albeit less common; other reported sites include the larynx, paranasal sinuses, facial nerve, thyroid and nasopharynx.[3–7]

In 1968, Wilson[8] stated that the hypoglossal nerve was a possible site for paragangliomas, although no evidence was given to support this assertion. Only in the last decade have case reports of hypoglossal paragangliomas been published, although the proximity of these lesions to the carotid artery has rendered pre-operative determination of their origin impossible. Shintani *et al.*[9] and Takayama *et al.*[10] have described tumours with intra-operative findings suggestive of hypoglossal nerve derivation, whilst pre-operative imaging suggested an origination from the vagus nerve within the neck. Maselli *et al.*[11] reported a similar case but made no comment as to what origin their pre-operative imaging suggested. Marchesi *et al.*[12] reported a patient whose familial predisposition had led to paragangliomas originating from the carotid bodies on both sides, the left vagus nerve and the left hypoglossal nerve, although these authors provided no imaging.

In this paper, we present a case of a paraganglioma derived from the hypoglossal nerve, clearly demonstrated by pre-operative imaging.

Case report

A 38-year-old man presented as an emergency to another hospital, with a seven-month history of progressive, left-sided weakness, numbness of the left upper limb, diplopia, choking, slurred speech, and urinary incontinence. Neurological examination revealed asymmetrical long tract signs with reduced limb power, increased tone and reduced sensation. Examination of the cranial nerves revealed left and right gaze invoked nystagmus and diplopia, whilst the left side of the tongue exhibited wasting and fasciculation.

An urgent computed tomography (CT) scan revealed a large lesion centred on the left hypoglossal canal and extending down into the neck, and superiorly into the posterior cranial fossa (Figure 1). The jugular foramen appeared free of disease. Magnetic resonance imaging (MRI) of the brain and cervical spine showed a mass within the lower half of the left posterior cranial fossa, extending down into the foramen magnum and compressing the medulla (Figure 2).

Following these investigations the patient was transferred to our care. Over the course of two weeks the patient developed increasingly severe symptoms of brainstem compromise, with bulbar incompetence and respiratory distress. An elective tracheostomy was performed, and the patient consented to surgical excision of the lesion with pre-operative embolisation.

Initial surgery addressed the cervical and skull base extension of the lesion with a combined left transjugular and far lateral approach. The jugular bulb was exposed and found to be free of tumour, whilst the hypoglossal canal was grossly expanded and diseased. The lesion extended anteriorly into the neck, lying between the internal carotid artery and the internal jugular vein.

Neurosurgical resection took place two days following the neck and skull base surgery. At this time, near-total removal of the lesion was achieved, with only a thin remnant left along the surface of the brainstem. All bulbar nerves except the hypoglossal were maintained in anatomical continuity.

Histopathological examination of the tumour showed nests of small, homogenous, epitheloid cells within a rich

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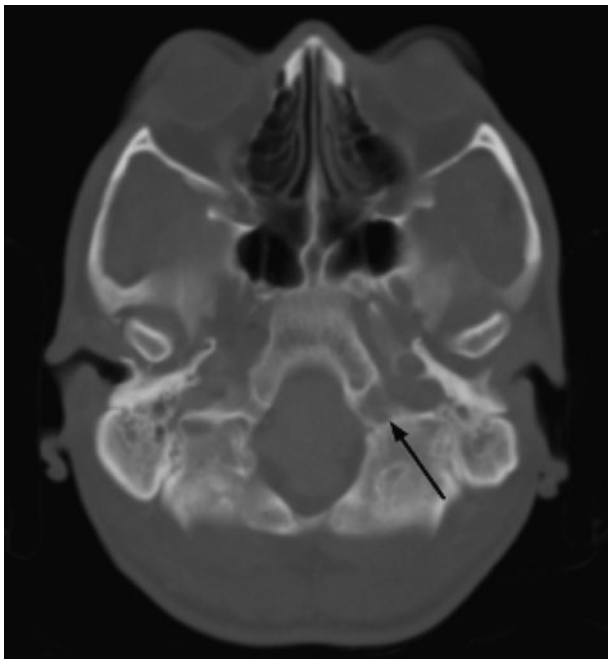

FIG. 1
Composite of axial computed tomography scans showing an expanded left hypoglossal canal (arrow).

vascular network that was largely sclerosed, with both cells positive for synaptophysin and S100-positive sustentacular cells (Figure 3).

Based on the above, a definitive diagnosis of a paraganglioma originating from the hypoglossal nerve was made.

Post-operative recovery was complicated by the development of a pseudo-meningocoele in the neck, requiring repair. The tracheostomy tube was removed five weeks after insertion.

Review at eight months following surgery found the patient dependent upon walking sticks for mobility, but otherwise making progress in terms of global rehabilitation. He had persistent Vth, IXth, Xth, XIth and XIIth palsies, but the VIIth and VIIIth nerves were normal. Despite these palsies, he was judged to have a safe swallow and was able to obtain adequate nutrition from oral intake. With the help of intensive physiotherapy, his shoulder strength was improving. He was also receiving ongoing speech therapy.

Discussion

The radiological imaging presented here, combined with the intra-operative findings, extend the evidence base for paragangliomas of the hypoglossal nerve. Whilst hypoglossal nerve palsies associated with paragangliomas of the jugular bulb are not uncommon,[13] there are few published reports describing paragangliomas derived from the hypoglossal nerve. Of these reports, none have presented radiological evidence supporting this origin.

Of particular interest is the manner in which our patient's tumour presented. Limb weakness and other long tract symptoms are not usually associated with head and neck paragangliomas,[14] although they have been described in patients with hypoglossal schwannomas.[15] More commonly, paragangliomas show a rather indolent pattern of growth, often producing symptoms of pulsatile tinnitus for many years before a definitive diagnosis is made. Clearly, our patient's tumour consisted in large part of a significant intra-cranial component, accounting for the severity of

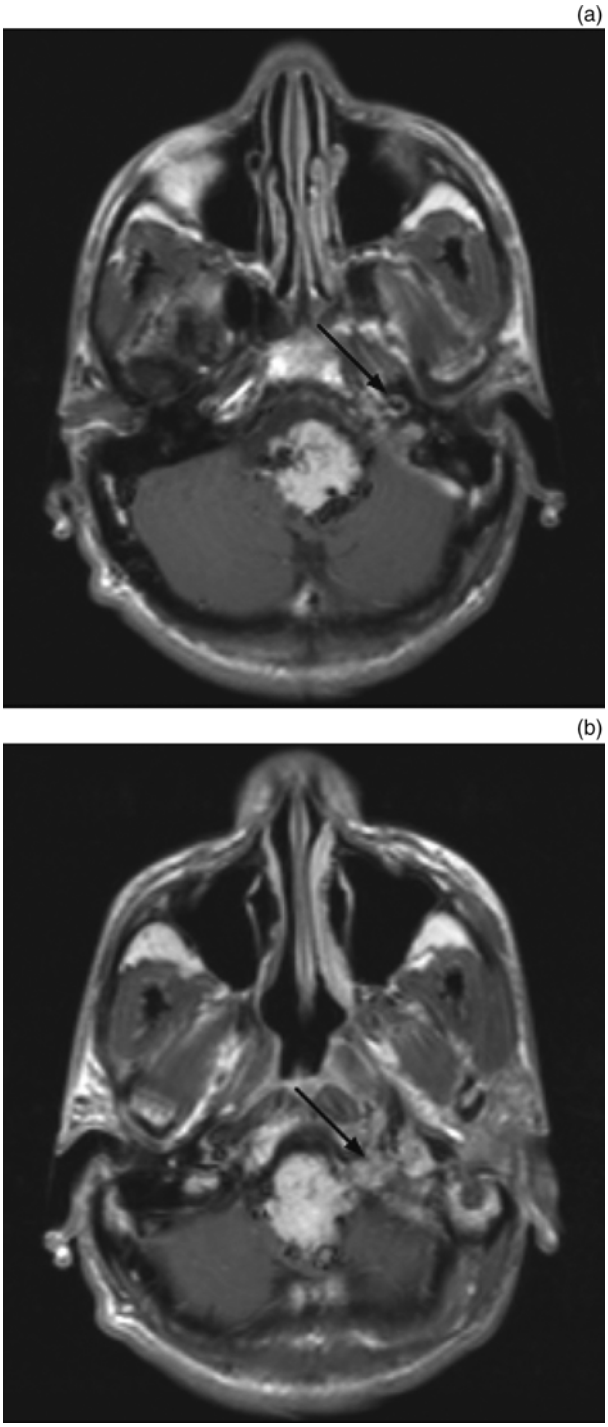

FIG. 2
Axial, T1-weighted, gadolinium enhanced magnetic resonance imaging scans showing (a) tumour within the posterior cranial fossa and a normal jugular foramen (arrow) and (b) tumour within the posterior cranial fossa and left hypoglossal canal (arrow).

symptoms. These lesions may originate in the intra-canalicular component of the nerve, leading to a tendency to grow in a dumbbell configuration. Anatomical studies of the hypoglossal nerve which search for normal paraganglial tissue at this site may help to substantiate this theory.

Perhaps of more clinical relevance is the observation that pre-operative diagnosis of a hypoglossal paraganglioma is of value in guiding the surgical approach to the tumour.

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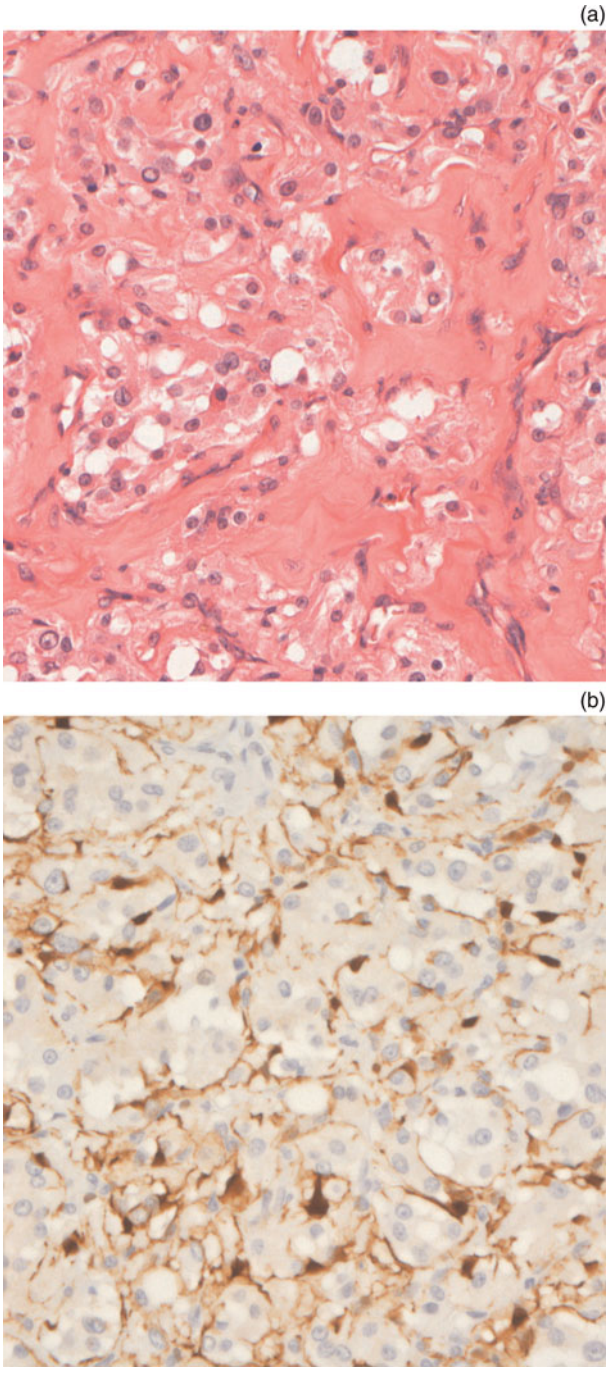

FIG. 3

Photomicrographs of (a) homogeneous, epithelioid cells within a sclerosed vascular network (H&E; ×200), and (b) brown-staining, S100-positive sustentacular cells (×400).

Calzada *et al.*[16] have recommended transjugular craniotomy to address dumbbell-shaped tumours involving the hypoglossal canal, such as paragangliomas of the jugular bulb. However, a hypoglossal paraganglioma such as the one presented here leaves the jugular bulb free of tumour, and therefore access to this region is not required, allowing the risk of damaging cranial nerves IX, X and XI to be minimised. We argue that the most appropriate approach would be a far lateral approach, which avoids risk to the structures of the jugular foramen; however, in our particular patient the value of this approach was eclipsed by pre-operative bulbar compromise. The far lateral approach[11] offers access to the hypoglossal canal, and can be extended anteriorly to address disease in the neck. Significantly, the brainstem is approached antero-laterally, allowing careful dissection of tumour from the emerging lower cranial nerve roots.

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Mr M Farr takes responsibility for the integrity of the content of the paper.
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