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Case Report
Clinical Pathology

Jugular foramen tumour resulting in hypoglossal denervation pseudohypertrophy: a rare and significant cause for tongue asymmetry

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Abstract. Paragangliomas of the jugular foramen are rare. They may present with symptoms of compression of the glossopharyngeal or vagus nerves, or due to secretion of catecholamines from chromaffin cells within the tumour. This case describes a rare presentation of glomus tumour. A 67-year-old patient presented with a 2-month history of right-sided tongue swelling. She was found to have an obvious swelling on the right side of the tongue but no obvious weakness or fasciculation on initial examination. Ultrasound confirmed diffuse muscle swelling, but no lesion within the tongue. Magnetic resonance imaging of the neck revealed an ipsilateral glomus jugulare tumour that extended to the hypoglossal canal, and had resulted in ipsilateral denervation pseudohypertrophy of the lingual muscles. This paper reviews presentation of glomus jugulare tumours and contributes a novel presentation of a rare entity.

Key words: glomus jugulare; glomus tumour; paragangliomas; schwannoma; hypoglossal; myositis; lingual; pseudohypertrophy.

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Jugular foramen tumours are rare lesions that may represent paragangliomas, schwannomas or meningiomas[1]. Paragangliomas are rare neuroendocrine tumours. These lesions most commonly affect the abdomen, but can rarely occur in the craniofacial structures. When they involve the cranial base, they are known as glomus tumours. Although malignant paragangliomas can occur, they are usually benign tumours that grow slowly.

Paragangliomas originate from neural crest cells, and therefore may occur at the carotid bifurcation, or follow the distribution of the autonomic nervous system. A paraganglioma at the jugular foramen of the skull base is also known

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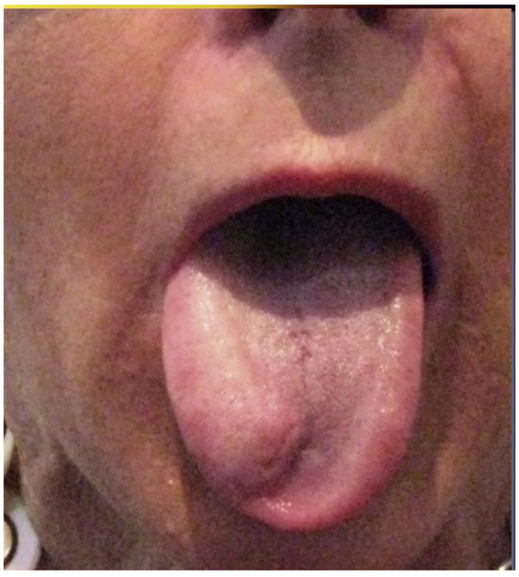

Fig. 1. Clinical photograph of the tongue: right-sided swelling of the tongue muscles, presenting as asymmetry. Note preservation of the tongue tip.

as a glomus jugulare[2]. Here, it often arises within paraganglionic tissue around the glossopharyngeal or vagus nerves[3].

Some glomus jugulare tumours may contain secretory chromaffin cells, resulting in release of catecholamines. This may present as labile blood pressure or tachyarrhythmias[3].

Glomus jugulare may present with direct pressure on the contents of the jugular foramen, the middle ear, or the tympanic membrane[1]. Pressure onto the middle ear or vestibulocochlear nerve can result in vertigo or tinnitus. Direct extension to the tympanic membrane can result in a bulging red tympanum on direct visualization. Pressure on the spinal accessory nerve can result in weakness of the ipsilateral sternocleidomastoid and trapezius. Pressure on the adjacent hypoglossal canal can cause impingement of the hypoglossal nerve. The resulting pseudohypertrophy of the tongue leading to diagnosis of the

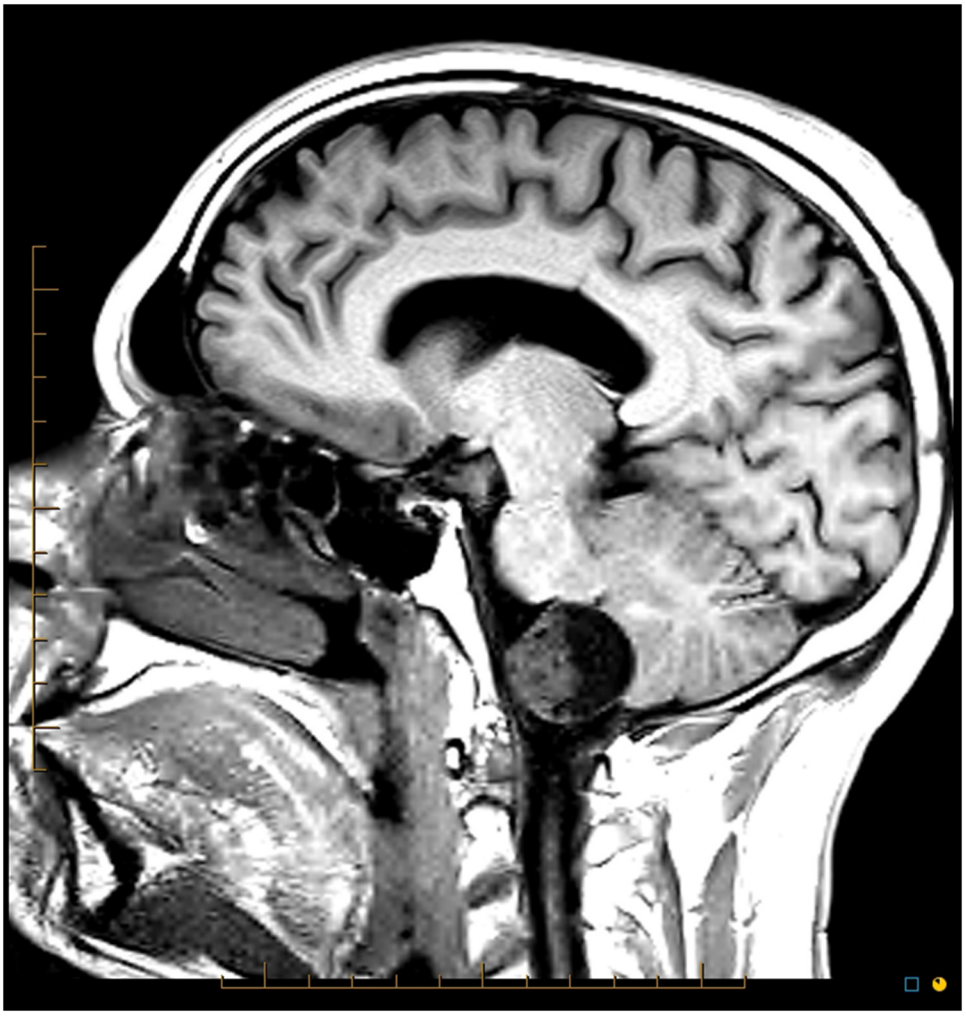

Fig. 2. Magnetic resonance image of lesion measuring 17 × 22 × 10 mm centred on the right jugular foramen. The lesion is of mixed signal and has ill-defined edges. The lesion also abuts the adjacent hypoglossal canal. T1 image in sagittal plane demonstrating salt and pepper appearance of glomus tumour.

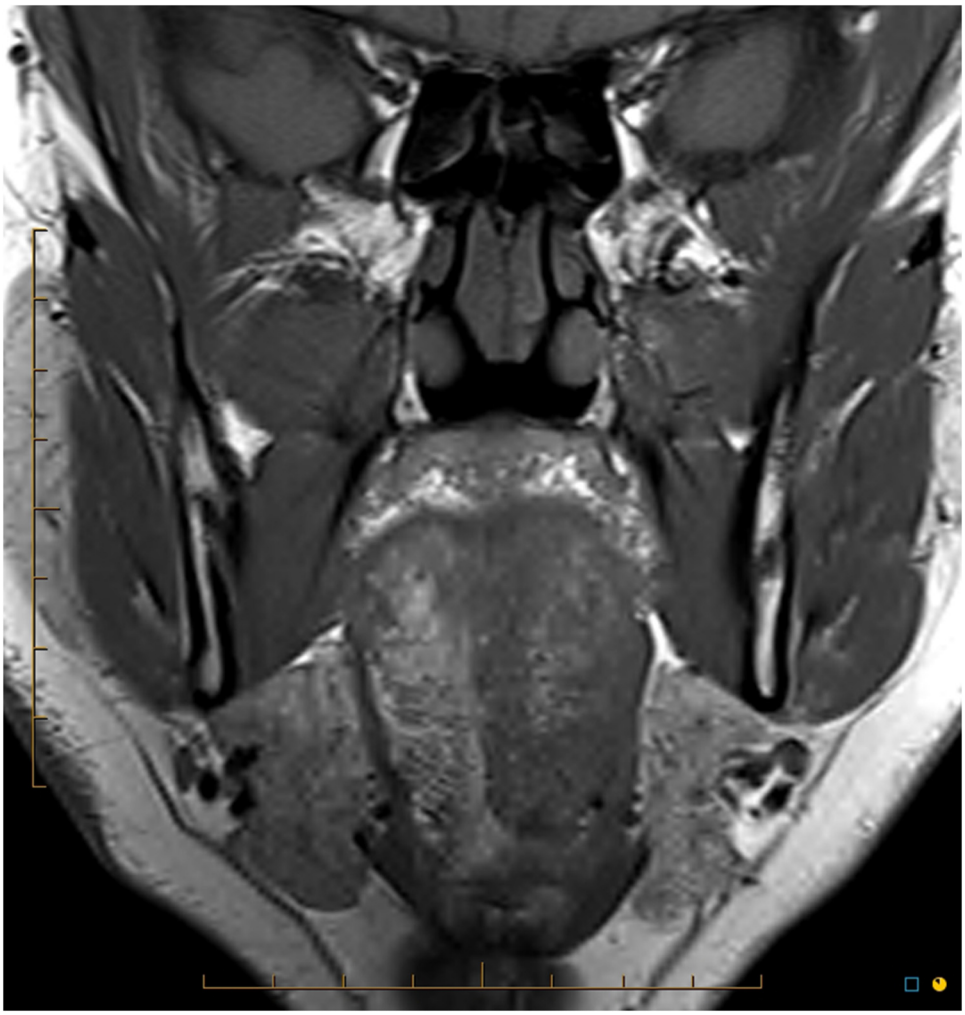

Fig. 3. Coronal T1 weighted magnetic resonance image of the patient's tongue showing increased fat infiltration (bright signal) of the right tongue. The left tongue maintains its normal darker signal representing muscle.

glomus jugulare has not previously been described.

Case

A 67-year-old female was referred to her Oral and Maxillofacial Surgeon with a 2-month history of right tongue swelling (Fig. 1). She was otherwise well, apart from atrial fibrillation, for which she took a beta blocker and an anti-platelet agent. On examination, the patient was noted to have diffuse swelling of the right side of the tongue, with no obvious weakness or fasciculation of the tongue. It was noted that she had a hoarse voice which had been present for a few months.

She was referred to an Otolaryngologist 4 weeks later for laryngoscopy by which time atrophy and fasciculation of the right side of the tongue was noted, she was also noted to have pulsatile tinnitus, and a red discolouration of the inferior half of the right tympanic membrane. Vocal cord function and pharyngeal sensation was intact on upper airway endoscopy, although the patient was found to have an intermittent hoarse voice.

An ultrasound of the tongue was suggestive of non-specific and diffuse muscular inflammation affecting the right tongue. Magnetic resonance imaging (MRI) revealed a lesion of $17 \times 22 \times 10$ mm in dimension, centred over the right jugular foramen of the skull base (Fig. 2). There was evidence of impingement of the hypoglossal and vagus cranial nerves. MRI of the tongue confirmed diffuse oedema of the right tongue consistent with denervation myositis. There was also fatty replacement of muscle tissue, resulting in pseudohypertrophy (Fig. 3). The base of skull lesion was consistent with a glomus jugulare, or a paraganglioma situated at the jugular foramen. Compression of the vagus nerve was hypothesized to have resulted in the patient's hoarse voice, and involvement of the hypoglossal nerve resulted in an ipsilateral denervation pseudohypertrophy.

The lesion was confirmed to be a stage 2 right glomus jugulare tumour with extension to the adjacent hypoglossal canal. This is in accordance with the Glasscock–Jackson classification, which defines a stage 2 glomus jugulare tumour as extending under the internal auditory canal, and which may extend into the intracranial vault[4]. Features that supported this diagnosis on imaging included the characteristic 'salt' (hyperintensity from blood products representing microhaemorrhages) and 'pepper' (signal voids representing small vessels) within the tumour mass on T1 pre-contrast MRI imaging.

However, computed tomography of a glomus tumour classically shows a permeative-destructive appearance at the bony margins, whereas in this case the bony

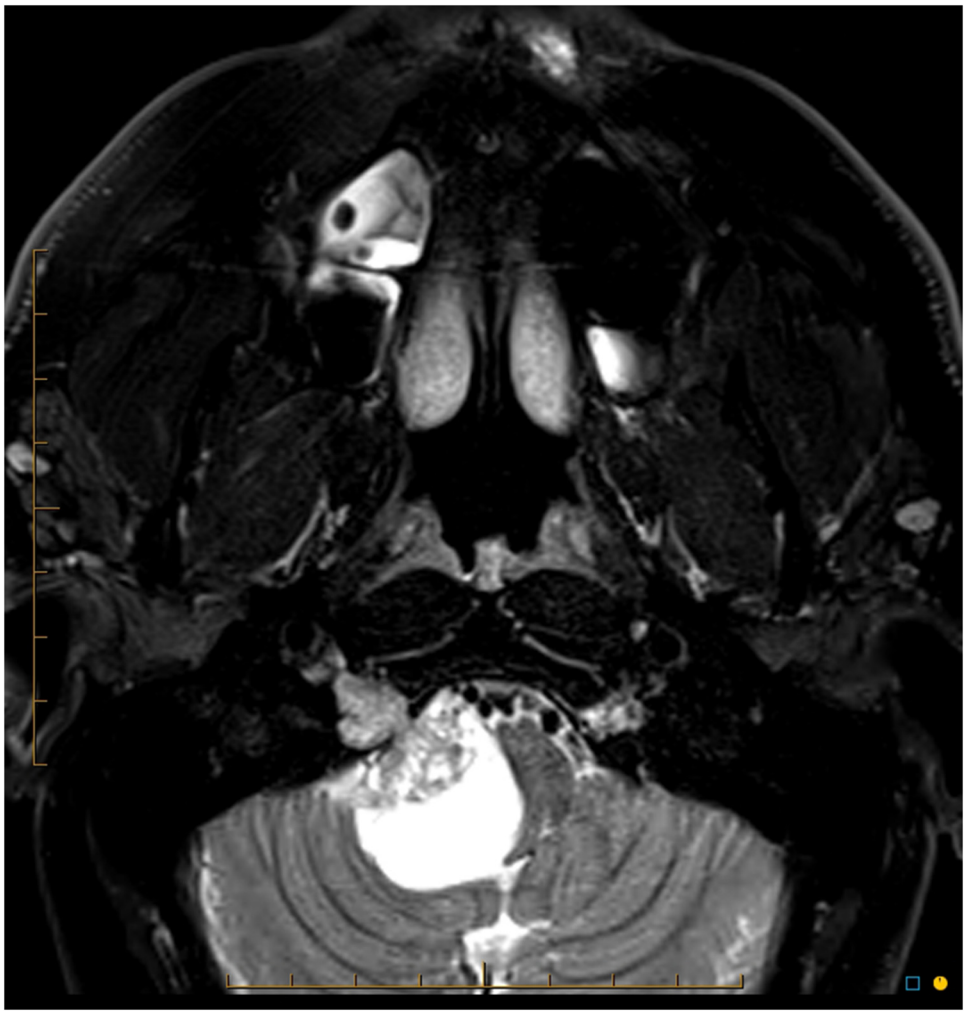

Fig. 4. Axial T2 fat-saturated magnetic resonance image showing cystic component of tumour, and tumour extension through the hypoglossal canal.

margin was smooth and scalloped, more suggestive of a nerve sheath tumour. Lastly, the lesion had a sizeable cystic component, which is also usually more typical of nerve sheath tumours (Fig. 4).

The patient was referred for primary hypofractionated stereotactic radiotherapy. This resulted in improvement of her myositis and weakness of her tongue, as well as resolution of the ipsilateral tinnitus.

The patient was followed up for 3 years with regular MRI showing no further growth of the glomus jugulare.

Discussion

Paragangliomas affecting the jugular foramen, also known as glomus jugulare, can present with symptoms of compression of the glossopharyngeal or vagus nerves. Less commonly, they can present with symptoms of increased circulating catecholamines. Although glomus jugulare is known to cause compression of the adjacent hypoglossal nerve, it's resulting denervation pseudohypertrophy, causing ipsilateral tongue enlargement, has not been previously described.

Pseudohypertrophy as a phenomenon has been described in cases of diabetic neuropathy, where instead of muscle atrophy, a diffuse fatty replacement of calf muscle presented as a pseudohypertrophy[5]. This denervation pseudohypertrophy has also been described in the tongue, as a result of idiopathic hypoglossal nerve palsy, but not previously because of a glomus tumour[6]. In this case, the ipsilateral tongue swelling appeared localized to the lateral third of the tongue (Fig. 1). This may be explained by some cross innervation of the midline and tongue tip.

The mechanism by which extension of the glomus tumour to the hypoglossal nerve causes denervation pseudohypertrophy is not known. Idiopathic hypoglossal nerve palsy is shown to cause a denervation pseudohypertrophy of the ipsilateral tongue that is characterized histologically by densely nucleated muscle fibres surrounded by increased amounts of perifascicular fatty infiltrate[6].

Glomus jugulare are low- grade neoplasms affecting the temporal bone and skull base. Several classification systems exist, two of which are the Fisch classification[7] (Table 1) and the Glasscock–Jackson classification[4] (Table 2).

The management of glomus jugulare can be either surgical excision, radiotherapy or observation.

Surgical excision is associated with significant morbidity due to damage to the adjacent cranial nerves[8]. This may be deemed unacceptable for a benign tumour.

Radiotherapy may be delivered either via external beam, or stereotactic. External beam radiotherapy (EBRT) was found to be as effective as surgery in tumour

Table 1. The revised Fisch classification of glomus tumours.

Stage	Description
A	Tumour limited to jugular foramen and middle ear
B	Tumour limited to middle ear, mastoid, but does NOT extend to infralabyrinthine space of temporal bone
C	Tumour extends into the petrous apex of the temporal bone
D	Tumour extends intracranially
	D1: <2 cm extension
	D2: >2 cm extension

Table 2. The Glasscock–Jackson classification of glomus jugulare tumours.

Stage	Description
I	Tumour limited to jugular foramen, middle ear and mastoid air cells
II	Tumour extends under the internal acoustic meatus
III	Tumour extends into the petrous apex of the temporal bone
IV	Tumour extends into clivus or infratemporal fossa

control, with a reduced morbidity[9], especially for larger lesions. Some studies even describe radiotherapy as superior to surgery, or a combination of surgery and radiotherapy, in achieving local control of glomus tumours[10].

Radiosurgery has been described for cranial base paragangliomas[11], where patients (n = 6) underwent a surgical resection, followed by radiosurgery, with a mean of 15.6 Gy. Benefits of radiosurgery are that it is relatively non-invasive and can be provided in an outpatient setting[3].

A meta-analysis of radiosurgery of glomus jugulare tumours found a 97% rate of success, defined by tumour volume stability or reduction on imaging, at a follow-up mean of 36 months[3]. Atypical clinical presentations require the treating surgeon to maintain an open mind. Swelling and weakness of one side of the tongue may represent pathology of the ipsilateral hypoglossal nerve, such as from compression secondary to a glomus jugulare.

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Competing interests

None.

Ethical approval

Not applicable.

Patient consent

Informed consent was gained for inclusion of a de-identified photograph.

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