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Glomus jugulare tumour presenting with isolated accessory nerve palsy

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Abstract
Glomus tumours of the skull base are rare, and most frequently present with symptoms of hearing loss and tinnitus. Diagnosis is often delayed due to the slow growth of the tumour. We describe the previously unreported occurrence of a glomus tumour presenting with a unilateral accessory nerve palsy.

Key words: Cranial Nerve Injuries; Glomus Jugulare Tumour; Accessory Nerve

Introduction

Glomus tumours of the skull base are rare, and most frequently present with symptoms of hearing loss and pulsatile tinnitus. Eight per cent of reported case series present with an ipsilateral facial nerve palsy.[1] Other higher cranial nerve palsies may also occur. Diagnosis is often missed until the tumour is very extensive, with the average delay to diagnosis being six years from onset of symptoms.[2] This paper describes the case of a glomus tumour presenting with a unilateral accessory nerve palsy. This had been treated as a simple frozen shoulder for two years prior to the diagnosis of glomus jugulare being made.

Case report

A 41-year-old lady presented to her local ENT department with a six-month history of right otalgia radiating to the neck, unilateral hearing loss, pulsatile tinnitus and hoarse voice, as well as fluctuating dysphagia and loss of appetite. She denied symptoms of aspiration, but had been admitted to hospital the previous year with a chest infection. On closer questioning she also described a two-year history of right shoulder pain throughout all ranges of movement, and weakness of abduction greater than 90° which had preceded the other symptoms by two years. There was reported to have been no obvious muscle wasting, and she had been diagnosed with frozen shoulder and treated with physiotherapy. Her previous medical history included seronegative arthritis, and long-standing visual field problems. She had not noticed any change in her vision recently.

Otoscopic examination revealed the classic appearance of a rising sun mass in the right middle ear. Examination of the cranial nerves revealed right sensory neuropathy of the ophthalmic division of the trigeminal nerve, right lateral rectus palsy, right vocal fold palsy, and right accessory nerve palsy (Figure 1). Facial nerve function was normal. Pure tone audiogram revealed a 15 dB conductive hearing loss on the right, with normal hearing on the left.[3] A gadolinium enhanced magnetic resonance image (MRI) scan was organized and demonstrated a large soft tissue mass arising from the right jugular foramen, and causing localized bony destruction (Figure 2). The patient was then referred to a tertiary centre for further management.

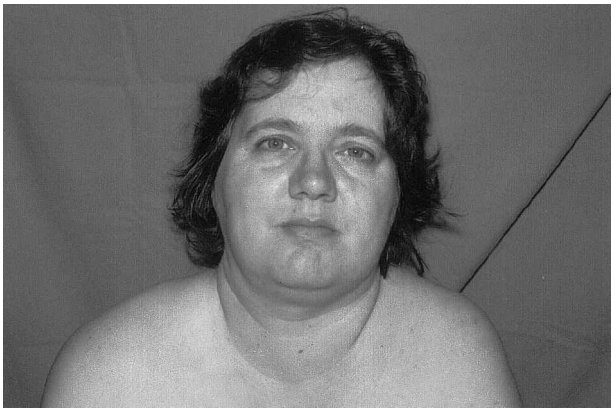

Fig. 1
Patient shrugging shoulders, demonstrating right accessory nerve palsy.

Further investigations were subsequently performed including a computed tomography (CT) scan, digital subtraction angiography, video fluoroscopy and neurophysiological testing (motor studies and electromyography (EMG)). The CT scan (Figure 3) demonstrated a large 5 cm glomus jugulare tumour, with considerable extension along the skull base and erosion of the petrous temporal bone at the jugular foramen. Within the petrous temporal bone the tumour extended medially close to the midline, into the upper neck, and intracranially it was classified as D2 on the Fisch classification (Table I).[4] Video-fluoroscopy showed mild weakness of the tongue base and pharyngeal constrictors, but no evidence of aspiration. Neurophysiological tests confirmed the presence of a moderately severe partial axonotmesis of the right accessory nerve in the proximal segment. In view of the presence of her ipsilateral vocal fold palsy, and concerns that it would not be possible to preserve the vagus nerve

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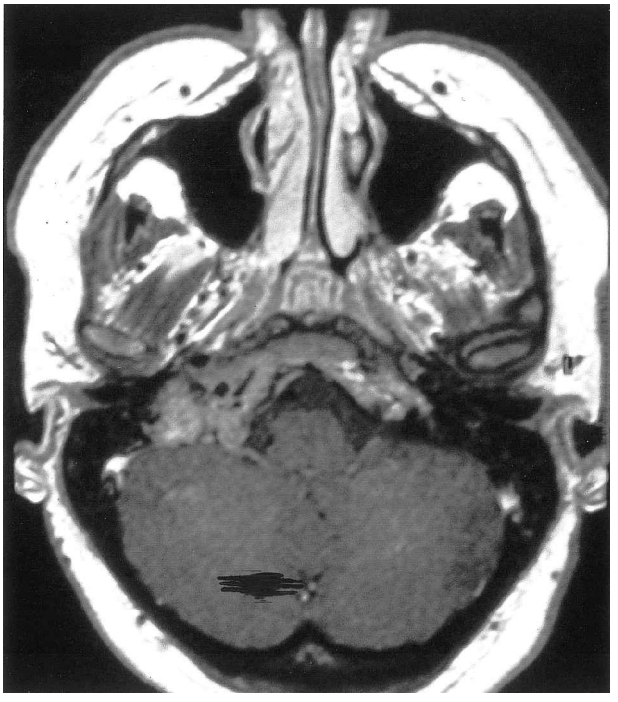

FIG. 2

Axial MRI with gadolinium, showing extensive right glomus tumour crossing midline.

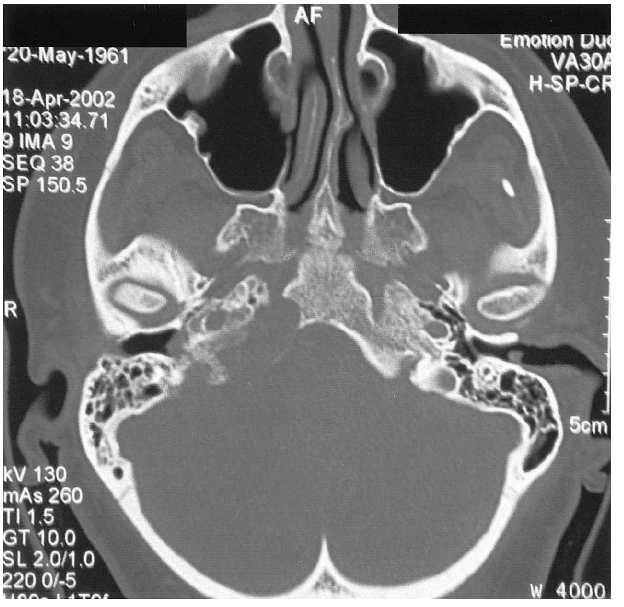

FIG. 3

Axial CT, showing extent of bony destruction from right glomus tumour.

intra-operatively, the patient was felt to be at high risk of aspiration post-operatively. In order to pre-empt this, she had a pre-operative percutaneous endoscopic gastrostomy (PEG) inserted.

Angiography demonstrated that the right ascending pharyngeal artery formed the principle vascular supply to over 90 per cent of the tumour. Three further supplies to the tumour were identified, from the right anteroinferior cerebellar artery, supplying the cerebellopontine cistern component of the tumour, and tiny branches arising from the right occipital artery, and from the right internal carotid artery. The ascending pharyngeal branch was successfully embolized, resulting in a greater than 90 per cent reduction in the vascularity of the tumour.

Two days later the patient underwent surgical resection of the tumour, via a postero-lateral approach. The facial nerve was preserved *in situ,* and the tympanic cavity and external auditory canal were left undisturbed. The tumour was removed in entirety, including the intradural portion. Unfortunately all the nerves of the jugular foramen were encased in tumour and had to be excised in order to achieve total tumour resection. Post-operatively the patient made a slow recovery, complicated by a minor wound infection, and a CSF leak which required surgical obliteration. At her latest review (six months post-operatively), her facial nerve function is normal (House Brackmann Grade 1) and she has only a mild ipsilateral conductive hearing loss She has successfully undergone thyroplasty, and has a reasonable quality of voice.

Discussion

The glomus jugulare is a collection of paraganglionic cells, derived from the neural crest, which are found in close association with the jugular bulb. Paraganglionic cells are found widely distributed within the autonomic system, and are divided into two groups on the basis of their endocrine activity. The paraganglia of the adrenal medulla secrete adrenaline and noradrenaline, and on histological examination stain chromaffin positive. Non-physiologically active paraganglia have a negative chromaffin reaction, and the glomous jugulare falls into this group. It is from this group of cells that glomus tumours arise. Thus, only a very small proportion (two per cent)[5] of glomus tumours have any endocrine activity, which can be diagnosed by measurement of 24-hour urinary vanillylmandelic acid (VMA) levels.

The histological appearance of glomus jugulare tumours is similar to that of normal glomus jugulare tissue, with only rare mitotic figures and a well-defined capsule. Malignant transformation and metastatic spread are rare, but these tumours are locally destructive and invasive of surrounding bone and neural tissue. The majority of glomus tumours arise sporadically, but there are literature reports of familial glomus tumours, with an autosomal dominant pattern of inheritance and variable penetrance.[6]

Classic presenting features of glomus jugulare tumours include hearing loss (69 per cent) and pulsatile tinnitus (55 per cent). Examination may reveal a middle-ear mass, the so-called 'rising sun sign' (75 per cent). Cranial nerve deficits most commonly affect the vagus (16 per cent), glossopharyngeal (16 per cent) and facial nerves (eight per cent).[1] Involvement of the accessory nerve has previously

TABLE I

OLDRING AND FISCH CLASSIFICATION OF GLOMUS JUGULARE TUMOURS[4]

Type A	Tumours localized to the middle-ear cleft (glomus tympanicum tumours)
Type B	Tympanomastoid tumours with no destruction of bone in the infralabyrinthine compartment of the temporal bone
Type C	Tumours invading the infralabyrinthine region and extending towards the petrous apex with destruction of the infralabyrinthine compartment of temporal bone
Type D	Tumours with intracranial extension

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only been described as a late symptom of extensive glomus jugulare tumours, with multiple cranial nerve neuropathies. The case presented here is unusual in that an isolated accessory nerve palsy was the presenting feature, and had been treated as a simple frozen shoulder for two years before any otological symptoms developed.

- **A patient with a glomus jugulare tumour with an accessory nerve palsy at presentation is reported**
- **This association has not been reported previously**

Differentiating between an accessory nerve palsy and a frozen shoulder can be a difficult clinical diagnosis. The classic features of an accessory nerve palsy are shoulder pain (due to joint subluxation), weakness, which is limited to shoulder abduction, mild winging of the scapula and wasting of trapezius muscle. In comparison, the features of a frozen shoulder are of pain and weakness throughout all ranges of movement. There is often a preceding history of trivial trauma which triggered the onset of symptoms. The characteristic course of the disease is of increasing pain and decreasing range of movement, which gradually resolves over a time course of 18 months. The case presented here was further complicated by the patient's preceding history of seronegative arthritis, which contributed to the delay in diagnosis.

There are several different classification systems for glomus jugulare tumours; the most widely used is that by Oldring and Fisch, which is based on tumour site and size (Table I).[4] Although there have been subsequent modifications to this system, the original classification as reproduced here is still the most useful.

The diagnosis of glomus jugulare tumours is frequently delayed as a result of the characteristically slow growth of these skull base tumours. Whilst the classical features of a middle-ear mass, pulsatile tinnitus and loss of hearing occur in the majority of patients, this case illustrates that a unilateral cranial nerve palsy may be the only presenting feature. Therefore, a careful clinical assessment of the ear should be carried out in all unilateral upper cranial neuropathies and may reveal an unsuspected underlying skull base pathology.

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