Glomus jugulare tumours

Synonyms: chemodectomas, nonchromaffin paragangliomas

What are glomus jugulare tumours?[1]

Glomus jugulare tumours are rare, slow-growing, very vascular tumours of a group called paragangliomas. They are derived from glomera jugulare (or glomus bodies) which themselves are derived from neural tissue and arise within the jugular foramen of the temporal bone.

They occur at such sites as the carotid body and vagus nerve. The tumours may also extend to involve the middle ear. Much more rarely, they may be found at other sites, including the periaortic area, trachea, larynx, mandible, nose, ciliary ganglion and Fallopian canal.

These tumours tend to be benign but they can be locally aggressive. This is important because of their proximity to the lower cranial nerves and to some major vascular structures.[2]

Only about 4% metastasise. Metastases have been found in the lung, lymph nodes, liver, vertebrae, ribs and spleen. The base of the skull is eroded with extension to the mastoid and occipital bones.

Between 2% and 4% of tumours produce catecholamines, noradrenaline (norepinephrine) or dopamine, resulting in a clinical picture similar to phaeochromocytoma with hypertension and tachycardia. Tumours may also produce somatostatin, vasoactive intestinal polypeptide (VIP) and calcitonin.

They sometimes run in families in an autosomal dominant fashion with incomplete penetrance.[3] The gene responsible for hereditary paragangliomas is on band 11q23.[4]

Because of the insidious onset of symptoms, these tumours often go unnoticed and delay in diagnosis is frequent. They may therefore be very large at the time of diagnosis.

How common are glomus jugulare tumours?[1]

- The annual incidence of glomus jugulare tumours is about 1 in 1.3 million people per year. However, it is the most common tumour of the middle ear and the second most common tumour of the temporal bone.
- They tend to present between 40 and 70 years of age (range: 6 months to 88 years).
- There is a female preponderance of between 3 and 6:1.
- It more commonly occurs on the left side; multicentric tumours are found in 3-10% of sporadic cases and in 25-50% of familial cases.

Glomus jugulare tumour symptoms (presentation)[5]

Symptoms

- The most common symptoms are deafness and a pulsatile tinnitus.
- There may be associated vertigo.
- Pain in the ear is uncommon.
- If the jugular foramen syndrome develops (paresis of cranial nerves IX to XI), there may also be complaints of hoarseness and symptoms associated with dysphagia.
- Less commonly, these tumours produce facial nerve palsy, hypoglossal nerve palsy or Horner's syndrome.[6]
- Ataxia and brainstem symptoms may also develop.
- Headaches can occur as a result of intracranial extension.

Signs

- The hearing loss may be conductive or sensorineural.

- There may be otorrhoea, haemorrhage, bruit and the presence of a middle-ear mass: otoscopic examination reveals a characteristic, pulsatile, reddish-blue tumour behind the tympanic membrane but this may represent the tip of the iceberg of this tumour.
- If the jugular foramen syndrome develops, look for evidence of paresis of cranial nerves IX to XI. It is pathognomonic for this tumour but it usually follows a year after the initial symptoms of hearing loss and pulsatile tinnitus.
- Look for evidence of intracranial extension, manifested by signs associated with hydrocephalus and elevated intracranial pressure.
- Involvement of the dural sinuses mimics sinus thrombosis.

A small number of patients present primarily with a phaeochromocytoma-like picture - eg, perspiration, pallor, nausea, hypertension and tachycardia.

Associated diseases[7]

Other tumours that have been reported in association with glomus jugulare tumours are:

- Phaeochromocytoma.
- Parathyroid adenoma.
- Thyroid cancer.
- Neurofibromatosis-1.

Investigations[7]

- Audiometry will show a mixture of sensorineural and conductive loss, the former more marked as the tumour expands.
- Plain skull X-rays may show evidence of the lesion with enlargement of the lateral jugular foramen and fossa.
- The best imaging technique is CT scanning combined with MRI with diethylenetriamine pentaacetate (DTPA) enhancement.
- Arteriography may be required before resection of large tumours.
- There should be screening for catecholamines.

Differential diagnosis

The list is long and includes:

- Otitis media.
- Chordoma.
- Histiocytosis X.
- Meningioma.
- Schwannoma.
- Neurofibroma.
- Primary or metastatic carcinoma.
- Cholesteatoma.
- Aneurysm.
- Lymphoma.

Classification

The most commonly used classifications are Glasscock-Jackson and Fisch. The Fisch classification describes four stages of tumour development.[8]

- **A** - tumour limited to the middle-ear cleft (glomus tympanicum).
- **B** - tumour limited to the tympanomastoid area with no infra-labyrinthine compartment involvement.
- **C** - tumour involving the infra-labyrinthine compartment of the temporal bone and extending into the petrous apex:
    - C1 - tumour with limited involvement of the vertical portion of the carotid canal.
    - C2 - tumour invading the vertical portion of the carotid canal.
    - C3 - tumour invasion of the horizontal portion of the carotid canal.

- **D** - tumour with intracranial extension.

    - D1 - tumour with an intracranial extension less than 2 cm in diameter.
    - D2 - tumour with an intracranial extension greater than 2 cm in diameter.

Glomus jugulare tumour treatment and management[5] [7]

There are different therapeutic options but the optimal treatment remains controversial.

- Observation without intervention is an option as 65% of the tumours remain stable and sometimes regress in size. Close follow-up is necessary with serial brain MRI with and without intravenous contrast.
- Traditional treatments involving surgical resection, external beam radiotherapy or both, have a significant risk of morbidity. Stereotactic radiosurgery has become increasingly popular treatment. External beam radiotherapy and stereotactic radiosurgery are comparable to surgical intervention in patients with jugular paragangliomas.
- Alpha- and beta-blockers may be required for some weeks before treatment if catecholamine production causes high and labile blood pressure. Any surgical approach may depend on the Fisch stage of the tumour.
- Large tumours that affect the lower cranial nerves present a high risk of postoperative complications, especially in older patients. Embolisation, radiotherapy, gamma knife radiosurgery and intratumoural injection of cyanoacrylate glue are other options.[9] [10]
- Conservative surgery has been combined with postoperative gamma knife radiosurgery which has enabled tissue diagnosis and improvement of symptoms without some of the serious complications of more extensive surgery.[2]
- Genetic screening is appropriate for individuals with a positive family history.[11]

Complications

Surgery can lead to damage of the cranial nerves. Other complications include bleeding, cerebrospinal fluid (CSF) leak, meningitis, uncontrollable hypotension/hypertension and tumour regrowth.[12] Death can also occur.

Prognosis[5] [13]

- Glomus jugulare tumours tend to grow slowly with only a small proportion metastasising. The associated cranial nerve palsies tend to be more cosmetic than debilitating.
- Even 20 years after treatment, the survival rate is 94% (77% of patients remain symptom-free).
- Depending on the study population, the overall five-year survival for metastatic paragangliomas is 35-60%.

Further reading

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Authored by:	Peer Reviewed by: Dr Hayley Willacy, FRCGP	
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