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Glomus jugulare tumours

A review of twelve cases treated with radiotherapy

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Introduction

GLOMUS tumours belong to a group of tumours called non-chromaffin paragangliomas and were first described by Rosenwasser in 1945. They are believed to be neoplasms of the glomus structures which lie along the course of the tympanic branch of the glossopharyngeal and along the auricular branch of the vagus nerves. They are also found in the adventitia of the dome of the jugular bulb and in the region of the vagal ganglia.

Glomus tumours are relatively rare tumours and yet they are the most common benign neoplasms involving the middle-ear. Lack *et al.* (1977) reported the distribution of 73 paragangliomas that occurred in 69 patients. There were 44 carotid body, 13 vagal body, 8 jugulo-tympanic, and 3 nasal paragangliomas.

They are vascular, encapsulated and usually behave in a benign manner (being locally invasive). They may cause extensive destruction within the petrous temporal bone. The overall histological appearance is of nests of polyhedral epithelioid cells separated by vascular septa (Fig. 1).

General features of glomus tumours:

1. There may be a familial tendency (Geokoop, 1932; Bartels, 1949; Ladenheim and Sachs, 1961; Alford and Guilford, 1962; Rosenwasser, 1969; Lack *et al.*, 1977).
2. They are more common in females (Alford and Guilford, 1962; Glasscock *et al.*, 1974; Harrison, 1974; Jackson and Koshiba, 1974; Spector, *et al.*, 1975b; Lack, *et al.*, 1977).
3. They can occur bilaterally (Bickerstaff and Howell, 1953; Alford and Guilford, 1962; Spector *et al.*, 1975b; Ogura *et al.*, 1978; Lack *et al.*, 1977).
4. A small proportion occur in association with carotid body tumours (Alford and Guilford, 1962; Spector *et al.*, 1975b; Lack *et al.*, 1977).
5. Nor-adrenalin secreting glomus tumours have been described which can produce symptoms and signs similar to a phaeochromocytoma (Duke *et al.*, 1964; De Lellis and Roth, 1971).
6. Rarely they become malignant and metastasise (Rosenwasser, 1958; Alford and Guilford, 1962; Taylor *et al.*, 1965; McCabe and

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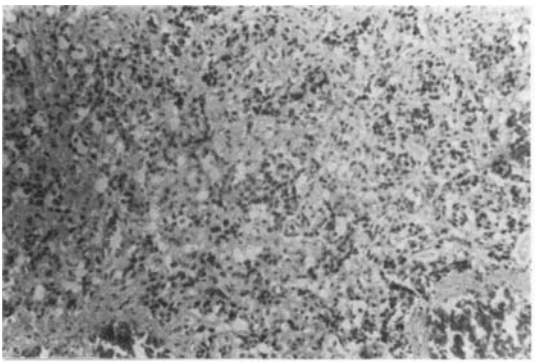


FIG. 1

Glomus jugulare tumour. Clumps of epithelioid cells can be seen separated by a vascular stroma.

Fletcher, 1969; Friedmann, 1974; Harrison, 1974; Spector *et al.*, 1975a; Druck *et al.*, 1976).

7. There may be a higher incidence of other associated malignancies being present simultaneously (Spector *et al.*, 1975b).

Clinical Presentation.

These tumours are classified according to their site of origin, i.e. glomus tympanicum, glomus jugulare and glomus intravagale. The clinical presentation and subsequent sequential development of symptoms vary according to the site of origin. It is often difficult to distinguish the source of the tumour as it is often extensive and involves several regions at the time of presentation.

Glomus tympanicum tumours arising from the middle ear characteristically cause a conductive hearing loss, a sensation of fullness in the ear, pulsatile tinnitus, purulent discharge and bleeding. Whilst the tumour in confined to the middle ear, it gives a 'setting sun' appearance behind an intact tympanic membrane. As it expands, it extends through the tympanic membrane and may present as a cherry red polyp at the external meatus (Fig. 2). It can cause facial nerve palsy as well as sensorineural deafness, with vertigo due to medial growth and labyrinthine erosion.

Tumours arising from the jugular body cause a jugular foramen syndrome, i.e. paralysis of the IX, X and XI cranial nerves. As the tumour expands it erodes bone and may involve the XII nerve in its separate canal (Fig. 3). If it has eroded into the middle ear it will cause symptoms and signs similar to a glomus tympanicum tumour.

Glomus intravagale tumours spread along the base of the skull to produce a pulsatile mass at the angle of the jaw or a parapharyngeal swelling. It will usually involve the last four cranial nerves.

Intracranial spread may involve the V and VI cranial nerves and produce a Horner's syndrome. At this stage the patient may complain of pulsatile headache.

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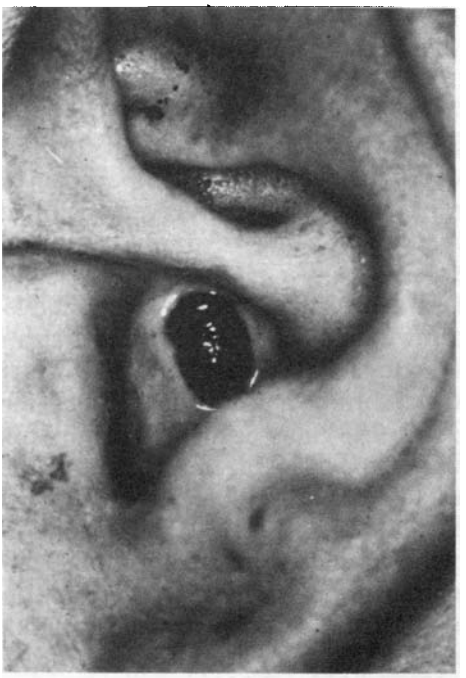


FIG. 2
A glomus tumour presenting at the external meatus.



FIG. 3
Case No. 10 demonstrating palsies of the left VII and XII cranial nerves.

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Pain is uncommon and is considered a sinister symptom as it seems to be indicative of malignancy (McCabe and Fletcher, 1969).

Diagnosis

Provisional diagnosis is made from the history, clinical examination and radiology. A glomus tumour must be suspected in a patient who presents with aural symptoms and involvement of the last four cranial nerves. (Aural symptoms being deafness, pulsatile tinnitus, vertigo, discharge and haemorrhage. Facial palsy may also be considered here as an aural symptom). Haemorrhage may be spontaneous or start after minor trauma such as following coughing, sneezing or straining.

Examination may reveal a typical cherry red aural polyp either behind an intact tympanic membrane or presenting in the external auditory canal. Brown's sign (Brown, 1953) may be elicited using a pneumatic auriscope. The tumour can be seen to pulsate as the pressure in the external meatus is raised. A bruit can sometimes be heard over the mastoid process and a palpable thrill may be felt over a mass at the angle of the jaw.

Radiology provides essential information regarding the site of origin and extent of the tumour. Jugular venography is important in distinguishing a glomus tympanicum from a glomus jugulare tumour and it will also delineate any extension down the jugular vein. This information is vital because a tumour that is still confined to the middle-ear is more amenable to surgical treatment than a glomus jugulare tumour presenting with middle-ear involvement.

Selective angiography and subtraction angiography will demonstrate the vascularity of the tumour and the origin of its blood supply. It is helpful in differentiating a high lying jugular bulb from a glomus tumour as they both have a similar macroscopic appearance. The venous phase will detect blockage of the jugular bulb.

There is some disagreement as to whether biopsy should be performed due to the danger of haemorrhage (Glasscock *et al.*, 1974). The characteristic findings on angiography are probably sufficient criteria for the diagnosis to be made.

CAT and brain scan will further aid the diagnosis by demonstrating any intracranial spread.

If a biopsy is taken to confirm the provisional diagnosis adequate tissue must be removed. Birrell (1955) pointed out that the surface of the vascular mass in the external meatus commonly has the appearance of modified granulation tissue; other areas, particularly those deep in the tumour, may show predominately fibrous tissue. Due to the danger of profuse bleeding at the time of biopsy cross matched blood should be available for transfusion.

Treatment

The management of these tumours varies according to the site of

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origin, its extent, the age and general condition of the patient. Surgery, radiotherapy and cryotherapy used singly or in combination are the methods of treatment.

Ligation of the external carotid prior to surgery will assist in controlling bleeding, as will embolization (Ogura *et al.,* 1978).

Small tumours confined to the middle-ear are usually well controlled by surgery, irradiation being reserved for the treatment of residual tumour. Slightly larger tympanicum tumours may require more extensive surgical procedures, as described by Shambaugh (1955), House and Glasscock (1968). If mastoidectomy is undertaken following radiotherapy there is a danger of subsequent radio-osteonecrosis developing.

There is indecision as to the best line of treatment for larger tumours as they involve inaccessible areas in close proximity to vital structures. Morbidity and mortality associated with surgery should be taken into account when comparing it to the relative safety of radiotherapy. Probably a combination of surgery and radiotherapy offer the best chance of curing these patients (McCabe and Fletcher, 1969; Hatfield *et al.,* 1972; Spector *et al.,* 1975c; Cole, 1977).

Material

The twelve patients in this study with glomus tumours were seen and treated at the Peter MacCallum Hospital, Melbourne, between 1958 and 1980. Table I summarizes their presentation and progress after treatment.

There were 8 females and 4 males. This ratio of 2 : 1 is probably not relevant due to the limited number of cases. Most larger series report a ratio of 4 : 1 (females : males).

The age distribution is from 20 to 80 years old at the time of presentation. Nine patients had radiological evidence of extensive erosion of the petrous bone. The right ear was involved in seven patients, with the left ear being involved in the remaining five. There were no bilateral cases nor were any carotid body tumours found.

Eleven had radiotherapy in air, the total dosage ranging from 2,400 to 5,200 rads. one patient was given 2,000 rads under hyperbaric oxygen. Two patients only had radical mastoidectomies, one before and the other following radiotherapy.

Two patients are alive with tumour still evident, 2 years and 2 months after receiving radiotherapy. Four cases are alive and well more than ten years after treatment. Case No. 6 is the longest surviving patient; she has remained well without visible tumour for 18 years.

Three patients have died in this series. In Case No. 2 the tumour diminished in size after receiving 2,040 rads, but it became evident again four years later. The tumour again shrank following a further 2,400 rads but death occurred four years after the last treatment from intracranial spread. Case No. 4 died from a cerebrovascular accident two years after receiving radiotherapy and 5 years after the onset of her symptoms. Case

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TABLE I

Case No.	Sex	Age	Period of symptoms	Cranial nerves involved	Treatment	Outcome
1	F	55	7 years	VII	2,500 rads	VII recovered. Alive and well 8 yea after radiotherapy.
2	M	65	10 months	—	2,040 rads and 4 years later 2,400	Tumour shrank initially but recurre 4 years later. Given further 2,40(rads. Two years after this treatmei VII palsy developed. Died from intracranial extension 7 years aft first treatment.
3	F	38	5 years	X, XI	5,000 rads	Alive and well 10 years after radiotherapy.
4	F	57	3 years	VII	4,990 rads	Died 2 years after radiotherapy from a cerebrovascular accident.
5	F	48	3 years	VII	Rad. mastoid. 4,000 rads.	Alive and well 15 years after radiotherapy.
6	F	52	2 years	VII, XII	5,200 rads	VII recovered as did deafness. Aliv and well 18 years after radiotherap
7	M	33	3 years	—	5,000 rads Rad. mastoid.	Alive and well 13 years after radiotherapy.
8	F	80	7 years	VII	2,400 rads	Alive with tumour 2 years after radiotherapy.
9	M	60	6 years	VII, XI, X, XI, XII	2,500 rads	VII recovered. Alive 3 years aftei radiotherapy.
10	F	75	3 years	VII, XII	2,500 rads	Died 4 years after radiotherapy. Cause unknown.
11	M	20	4 years	IX, X, XI, XII	2,500 rads	Alive and well 3 years after radiotherapy.
12	F	62	11 years	—	2,000 rads hyperbaric oxygen	Alive 6 months after radiotherapy Tumour still visible.

No. 10 died of unknown causes four years after having radiotherapy and six years after the onset of her symptoms.

Table II lists the presenting symptoms in order of their frequency. Table III demonstrates that the facial nerve was the commonest cranial nerve involved. In three cases facial nerve palsy recovered following radiotherapy. No side-effects from radiotherapy were recorded.

TABLE II

SYMPTOMS ON PRESENTATION

Deafness	11
Facial paralysis	7
Bleeding from ear	4
Tinnitus	4
Vertigo	4
Pain	2
Hoarseness	2

TABLE III

CRANIAL NERVES INVOLVED AT PRESENTATION

VII	7
XII	4
X	3
XI	3
IX	2

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Discussion

Glomus tumours are usually slow-growing and patients can live for long periods harbouring one. Bickerstaff and Howell (1953) reported a case with extensive intracranial involvement who had had symptoms for 42 years before treatment was instituted. It is therefore apparent that the usual index for measuring treatment success (i.e. five-year survival rate) is not applicable to this tumour. Radiotherapy, however, has been shown to be effective here in causing shrinkage and disappearance of tumour as well as recovery of cranial nerve function. In the patients who have survived for longer than ten years it can definitely be concluded that radiotherapy either halted growth of the tumour, preventing further morbidity, or in fact may have completely eradicated it. Radiotherapy has been found to be equally as effective by other authors (Glover and Block, 1972; Hatfield *et al.*, 1972; Silverstone, 1973; Thomsen *et al.*, 1975; Cole, 1977; Simko *et al.*, 1978).

Summary

This series reviews the presentation and results of treatment with radiotherapy in twelve cases of glomus jugulare tumour. The results demonstrate the effectiveness of radiotherapy in treatment.

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