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Glomus Jugulare Tumours: a Review of 61 Cases

L. D. Watkins[1], **N. Mendoza**[1], **A. D. Cheesman**[2], and **L. Symon**[1]

[1]Gough-Cooper Department of Neurological Surgery, Institute of Neurology, Queen Square, London and [2]E.N.T. Department, Royal National Throat, Nose and Ear Hospital, London, U.K.

Summary

A retrospective study of 61 patients with glomus jugulare tumours treated at the National Hospital for Neurology and Neurosurgery, and at the Royal National Throat, Nose and Ear Hospital, London. The average age at presentation was 41.7 years. The patients were mainly treated by a posterolateral combined otoneurosurgical approach. 42/61 of the patients had total or subtotal excision of their tumours, 7/61 had partial removal and the remaining 11/61 had no operation. Only one case required a 2-staged procedure. There were two deaths in the postoperative period, one from intracerebral haemorrhage and the other from the left hemisphere infarction.

Postoperative radiotherapy was given to 5/7 of the patients who had partial removal. 3/40 of the patients with total removal had postoperative radiotherapy, and a further 3/40 had received radiotherapy pre-operatively.

Of the 11 patients who did not undergo surgery, 7/11 were treated with radiotherapy and 4/11 had embolisation only.

Keywords: Glomus jugulare tumour; skull base; paraganglioma.

Introduction

Winship and Louzan[25] first proposed the term glomus jugulare tumour, for neoplasms arising from paraganglionic cells in the region of the jugular bulb. The original descriptions of paraganglionic tissue in the petrous temporal bone were by Valentin[23] and Krause[14] in the nineteenth century, and Guild[9] first recognised the histological similarity between the non-chromaffin staining cells in the jugular bulb and that of the carotid body.

The glomus jugulare is thought to be a part of the parasympathetic paraganglion system, which mediates chemosensory reflexes. The other components of this system include the carotid and aortic bodies, along with the glomus tympanicum and ganglion nodosum (respectively along Jacobson's and Arnold's nerves).

Tumours of the glomus jugulare are rare neoplasms, although they are the commonest benign tumour of the middle ear cleft. Due to this fact, and their marked tendency to local erosion of the temporal bone, including intracranial spread, they are of interest both to otologists and neurosurgeons, and usually a combined otoneurosurgical approach is appropriate.

Glomus jugulare tumours typically present in middle age and are more common in females. Histologically these tumours are composed of nests of epithelioid or chief cells in a highly vascular stroma consisting of capillary and pre-capillary calibre blood vessels surrounded by a thin fibrous capsular layer[10]. They have been classified into typical, adenomatous and angiomatous types depending upon the predominance of epithelioid or stromal elements[15].

Although generally considered to be histologically benign they have a tendency to recur locally. The occurrence of malignant glomus tumours is extremely rare: we have previously reported a case of malignant paraganglioma of the glomus jugulare[13], but there are only 16 other such cases in the literature.

The current series is one of the largest reported from personal experience, although Glasscock and colleagues have reported follow-up of 98 glomus jugulare patients[22].

Clinical Material and Methods

We have previously reported a series of 31 patients with glomus jugulare tumours, and included a description of the single-staged posterolateral combined otoneurosurgical approach[2]. The current paper further expands our series to almost double the original number, and includes further follow up. The current study was based on retrospective review of case notes at both clinical centres involved.

Results

Presentation

The presenting features were: unilateral deafness (42/61), tinnitus (34/61), hoarseness (10/61), dysphagia (10/61), headache (9/61), facial pain or numbness (6/61), facial palsy (5/61), ataxia (5/61), diplopia (4/61), papilloedema (2/61) and obscuration of vision (1/61). The average duration of onset of first symptom until presentation was 3 years.

Treatment

The patients were mainly treated by a posterolateral combined otoneurosurgical approach. 42/61 of the patients had total or subtotal excision of their tumours, 7/61 had partial removal and the remaining 11/61 had no operation. Only one case required a 2-staged procedure.

Postoperative radiotherapy was given to 5/7 of the patients who had partial removal. 3/42 of the patients with total removal had postoperative radiotherapy, and a further 3/42 had received radiotherapy pre-operatively.

Of the 11 patients who did not undergo surgery, 7/11 were treated with radiotherapy and 4/11 had embolisation only.

Clinical Outcome

1. Complications

There were two deaths in the postoperative period, one from intracerebral haemorrhage due to paroxysmal hypertension (see discussion of investigations below) and the other from left hemisphere infarction in a patient who had an intracarotid tumour extension which invaded the cavernous sinus and led to forced sacrifice of the carotid.

Some wound complication such as discharge or pseudomeningocele was noted in 8/49 of the patients and in one case this led to meningitis but without long-term sequelae. Other complications were chest infection (1/49), myocardial infarction (1/49), deep venous thrombosis (1/49).

2. Neurological Function

Increases in neurological deficits noted in the immediate postoperative period were: lower cranial nerve (IX, X, XI, or XII) palsy (15/49), facial palsy (8/49), VI palsy (3/49), increased deafness (2/49). If the tumour extended into the posterior fossa, few postoperative lesions of the lower cranial nerves ever recover; only if the tumour were confined to the jugular foramen can the nerves later regain function.

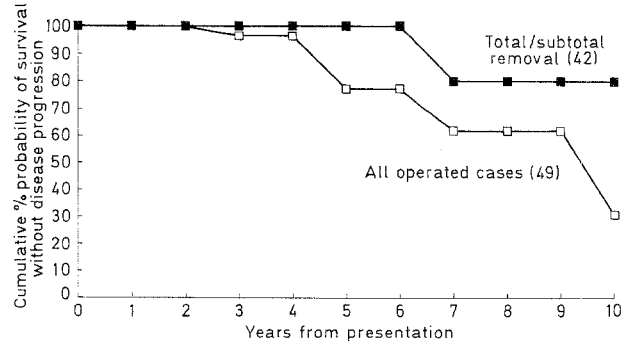

Fig. 1. Kaplan-Meir plot showing cumulative probability of survival without disease progression. Upper curve based on patients who had total or subtotal removal. Lower curve based on all operated patients. Starting number for each curve in parenthesis. Mean follow-up = 3.4 years

3. Survival

Apart from the postoperative mortalities noted above, only 2 of the patients who underwent surgery are known to have subsequently died; at 3 years and 10 years after their operations. It must therefore be considered that the prognosis for postoperative survival is good. It is probably more helpful to analyse the period free of disease recurrence or progression, and this has been expressed as a probability plot of progression-free survival (Fig. 1). A Kaplan-Meir survival probability curve[18] is a widely established method of presenting survival data in oncology: in this case we have taken the end point as diagnosis of progression or recurrence, rather than death. Only those patients who had operations (49) are included, since the others were followed up under the care of radiotherapy or oncology departments at other hospitals.

Discussion

Investigation

The definitive diagnosis is based on high resolution radiographic evaluation, usually computed tomography in the first instance, although the literature variously mentions the use of other neurodiagnostic techniques including hypocycloidal polytomography, posterior fossa cisternography, arterial and venous phase four-vessel angiography and retrograde jugular venography. At the Royal National Throat, Nose and Ear Hospital excellent results have been obtained with sub-

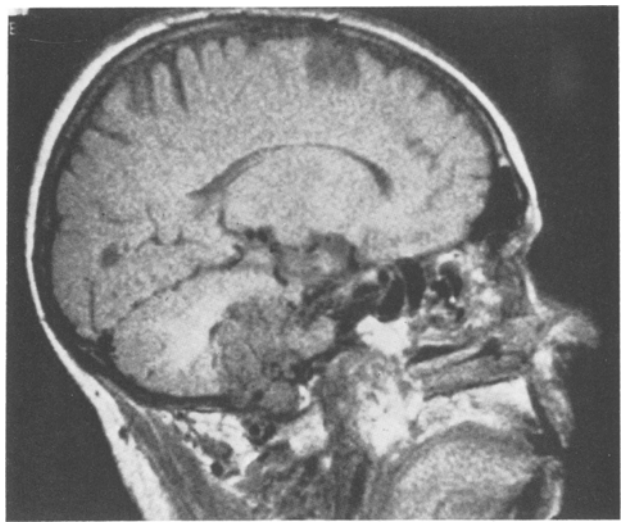

Fig. 2. Sagittal MR scan showing extension into posterior fossa

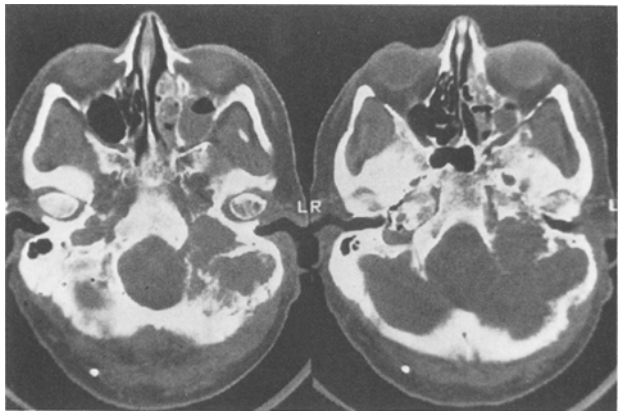

Fig. 3. Axial CT scan showing bone erosion around left jugular foramen

traction gadolinium enhanced magnetic resonance[16]. Current practice in most centres would include magnetic resonance imaging, which can produce incomparable detail of the relationship of the intracranial portion to neurological structures (Fig. 2). However, high resolution computed tomography is still used to evaluate the extent of jugular bulb bony erosion, middle ear and labyrinthine involvement (Fig. 3). Four-vessel angiography is recommended for all patients (Figs. 4 and 5), not only to evaluate vascular supply of the tumour, but also because about 10% of cases have been reported to have bilateral or multiple paragangliomas (carotid, vagal or aortic)[11].

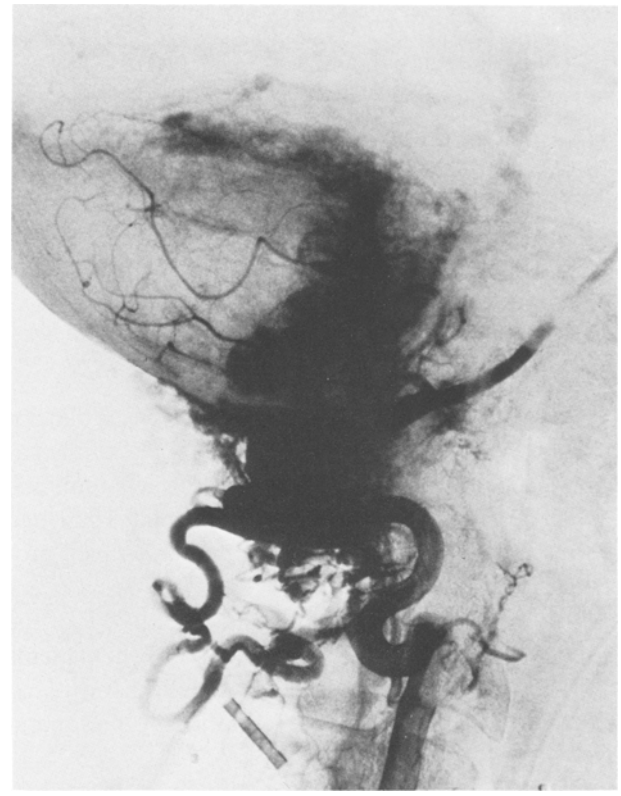

Fig. 4. Angiogram showing vascular supply of a tumour which extends into the posterior fossa

At the time of angiography, usually 4–7 days preoperatively, most patients also undergo embolisation. This has been a considerable contribution to improving haemostasis at operation, and has also been used to treat patients considered too unwell to undergo surgery[1]. In our series, 4 of the cases had embolisation alone.

Rarely, it has been reported that glomus tumours may secrete vasoactive substances similar to pheochromocytoma[3], and that any patient with a history of paroxysmal hypertension should undergo pre-operative assessment of urinary vanillylmandelic acid (VMA, the breakdown product of noradrenaline) and 5-hydroxyindole acetic acid (5-HIAA, the breakdown product of serotonin)[20]. We now recommend that all patients undergo screening, since 2 patients in our series had functioning tumours. One patient had headaches, anxiety attacks, nocturnal diaphoresis and an episode of haematospermia. He was found to be grossly hypertensive. Urinary studies and selective venous sampling revealed an active catecholamine-secreting glomus tumour occupying the left jugular bulb. However, the other case was the patient who died from intra-

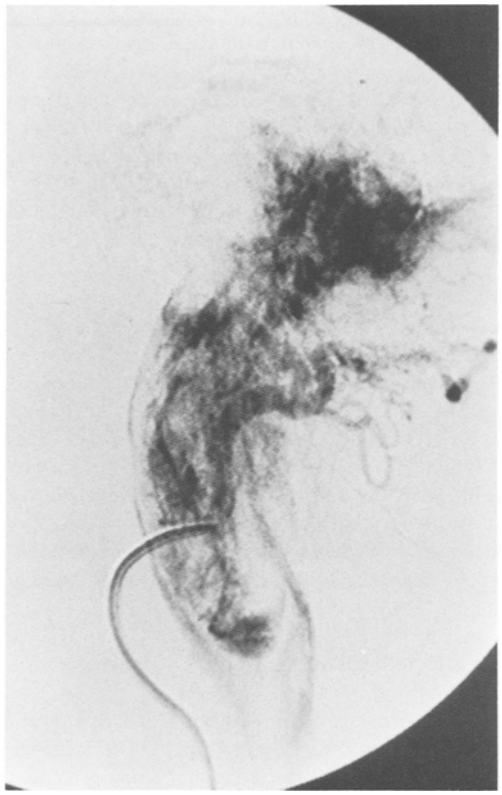

Fig. 5. Angiogram showing tumour in lumen of jugular bulb

cerebral haemorrhage, who had no pre-operative suggestion of paroxysmal hypertension but was found to have unrecognised multiple apudoma, one of which was functional.

Conclusion

The close relationship of these tumours to the neural and vascular structures of the base of the skull has made them difficult to manage operatively, and prone to disastrous postoperative complications. Their relative radio-resistance means that surgery remains the mainstay of management, and improvements in technique are available for the preoperative, operative and postoperative phases of management which make it possible to remove all but the largest tumours with little morbidity or mortality. In 1992 a review of 175 cases from the literature and experience of 26 cases of jugulare foramen paragangliomas, showed an operative mortality rate between 0 and 5%. The rate of incomplete removal of C and D tumours (according to the Fisch classification)[12] was about 15%[7]. This is comparable with our current series, and contrasts with other reports in which the rate of partial removal was 59% for glomus tumours of the skull base and in which the authors recommended conservative treatment[24].

One of the most significant advances in management has been preoperative, transvascular embolisation of these tumours. The procedure should be performed by an experienced radiologist (even in such hands there is a 1% risk accidental internal carotid embolisation).

The infratemporal approach by Fisch[4–6], modified by Glasscock[8], pioneered total removal of lesions which required the exposure of the infratemporal portion of the internal carotid artery. The essential features of this approach are the permanent anterior re-routing of the facial nerve to gain access to the jugular bulb and the resection of the jugular bulb itself. The surgeon benefits from a wide exposure with this approach, good control of the internal carotid artery and access to the intracranial compartment, but the management of intracranial extension can still provide problems, and the manipulation of the facial nerve leads to potential morbidity.

The postero-lateral approach is a modification of the infratemporal or lateral approach. We feel the requirements of this approach are best fulfilled by a neurosurgeon and an ENT surgeon working together as a team, approaching all type D tumours by combining a posterior fossa craniectomy with an infratemporal fossa approach in a one-stage operation. With this approach the facial nerve is not re-routed unless absolutely necessary and the wide postero-lateral exposure allows preservation of the posterior meatal wall. It also enables the compression displacement of the intracranial portion of the tumour into the established bone defect, thus facilitating the intracranial removal. These lesions are seldom adherent to the cerebellum, and in only one of our cases has there been vascular supply from intracranial vessels – a minor branch of the anterior inferior cerebellar artery.

The operative aim is eradication of the tumour, but the potential risk of increasing the patient's deficit must always be considered, and the operative approach chosen to minimise this risk. Access to the base of the skull is the key, while allowing removal of bone from the surface of the tumour, preservation of the facial nerve, and control of the sigmoid sinus. There is no doubt that in those cases where the intracranial extension is considerable, the nerves in the jugular foramen are more at risk. Dissection of the rootlets of IX and X, particularly from a substantial intracranial mass, is generally impossible. We feel that when a more conservative resection is used, postoperative irradiation or the use of an interstitial radioactive implant[21] is re-

commended. This is supported by the finding of significantly improved disease control in patients receiving both radiotherapy and surgery[19].

Often cranial nerve palsies are inevitable, but may be transient and otherwise most patients adapt to the unilateral paresis. Where complete paralysis of the jugular foramen nerves has been present preoperatively, gradual adaptation to their destruction will have ameliorated the swallowing problems, and these are unlikely to be substantially worsened by operation. Even patients with multiple lower cranial nerve deficits can regain normal oral intake, after vocal cord medialisation and palatal adhesion where appropriate, although it is reported that results are less certain with elderly patients[17]. It is in the first few postoperative days that damaging pulmonary complications are most likely to occur, and the cooperation of experienced physiotherapists and the judicious early use of tracheostomy if necessary will prevent disastrous pulmonary complications.

Most glomus tumours are operable in spite of intracranial extension and early surgery avoids the development of further cranial nerve palsies. Careful preoperative investigation and preparation are important. The postero-lateral approach enables the resection of most lesions, including the very large which have already caused brainstem compression and hydrocephalus.

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Correspondence: Lindsay Symon, CBE, TD, FRCS, Gough-Cooper Department of Neurological Surgery, Institute of Neurology, Queen Square, London WC1N 3BG, U.K.