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Recommended Citation

Bari, M. E., Kemeny, A. A., Forster, D. M., Radatz, M. W. (2003). Radiosurgery for the control of glomus jugulare tumours. *Journal of Pakistan Medical Association*, 53(4), 147-151.

Available at: https://ecommons.aku.edu/pakistan_fhs_mc_surg_neurosurg/141

Radiosurgery for the control of Glomus Jugulare Tumours

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Introduction

Glomus jugulare tumours though rare, are the most common tumours of the middle ear and jugulotympanic paragangliomas are the second most common tumours of the temporal bone. The true incidence of this vascular tumour is unknown, but is estimated around one per million.¹⁻³

Glomus tumour was first described by Valentin but its definitive description is credited to Stacey Guild of John Hopkins who coined the term "Glomus Jugulare" in 1941.⁴ Glomus jugulare tumours arise from the paraganglion cells, which are closely associated with the vasculature and cranial nerves derived from the branchial arches (branchiomeric paraganglia). The paraganglia takes origin from the neural crest cells and belongs to the diffuse neuroendocrine system. All these cells can demonstrate neuropeptide content or secretion. Tumours arising from these cells are histologically similar to tumours arising in other chemoreceptors. Although every paraganglioma may potentially secrete catecholamines, clinically functioning paragangliomas are only 1-3% as a four to fivefold elevation of serum norepinephrine is necessary to produce detectable symptoms.^{5,6} Paraganglia of the temporal bones are ovoid bodies measuring .1mm to 1.5mm in diameter.⁶ There are usually three bodies in each ear. They accompany Jacobson's nerve (tympanic branch of glossopharyngeal nerve) or Arnold's nerve (auricular branch of vagus nerve). 50% of the paraganglia are located in the jugular fossa, 10% in the mucosa of the promontory and approximately 20% are found in inferior tympanic calculus. Microscopy demonstrates clusters of the epithelioid (chief) cells and sustentacular cells enmeshed in a web of small blood vessels.

Glomus jugulare are slow growing, locally invasive tumours, showing female preponderance and average age of presentation is between 50-60 years.⁷⁻¹⁰ Although there is no clear racial predilection, the glomus tumour seems to be more common in whites. Ten percent of the tumours are multicentric in origin in the non-familial cases. However this frequency for multicentricity is exaggerated in the familial glomus tumours 25-50%. Glomus jugulare tumours are generally benign but 1-3% metastasise. The common sites for metastasis in order of decreasing frequency are lung, adjacent lymph nodes, liver, spine, ribs and spleen.^{6,11-13}

The interval from the first symptom to diagnosis is usually between 3-6 years. Symptoms are caused by the invasion of adjacent lower cranial nerves, mass effect, and high blood flow through the lesion (pulsatile tinnitus) or because of the neuroendocrine effects. The treatment of these tumours is controversial and consists of surgery, radiotherapy and

embolization. Each has its proponents claiming one is better than the other.

Methods

Since 1985 in the national centre for stereotactic radiosurgery in Sheffield we have treated 14 cases of glomus jugulare tumour with the Leksell gamma knife. All except one, are being followed up at the local referring hospitals. Eight of these patients have been followed up for more than four years. Three out of these eight patients had a minimum follow-up of more than seven years. Four patients had follow-up between 5-6 years, one patient for 4.4 years. Our knowledge about these patients is based on the clinical assessment of the referring neurosurgeons in their follow-up clinics. All patients apart from one were contacted on telephone to obtain an update on their condition. Repeat scans were done at yearly intervals at the referring hospital. Each patient had a minimum of 3 and a maximum of 5 MRIscans in the follow-up period. Two patients had check angiography and one had digital subtraction angiography. These scans were sent to our department to be compared with the treatment scans and measured objectively.

Eligibility Criteria Tumours with maximum diameter less than 40mm in any dimension. No sign of raised intracranial pressure or mass effect. No neuroendocrine function (non functioning tumours only). The parameters studied included: radiological follow up and improvement, tumour size and volume changes, symptomatic improvement and development

Results

Patient Follow up

The minimum radiological follow-up was 39 months, maximum 80 months, mean 44 months. The minimum clinical follow-up was 52 months and maximum was 97 months, mean 74 months.

Patient Demography and Symptoms

There were 3 males and five female patients. All of them were white (Caucasians). The age of the patients ranged between 32-64 years. The symptom duration before the radiosurgery was 1-8 years. Clinical symptoms are shown in Table 1.

Two patients had previous surgery; one had radiotherapy along with surgery. One patient had had unsuccessful embolization prior to radiosurgery. Four patients had stereotactic radiosurgery as primary treatment.

Tumour size, volume, localization and control

In our study one patient's tumour was localised with a combination of CT scan and angiography with a combination of MRI and CT. Two patients had the combination of MRI scan and angiography and three patients' tumours were localised with MRI scan alone.

The target volume of the tumours ranged between 800 cm³ to 6300cm³, mean 4025 cm³. Volume of the actual lesions ranged between 970 -10200 cm³. Target volume is the volume, which received the prescribed volume of radiation at the time of radiosurgery. The tumour volumes were measured on the MRI scans at the time of radiosurgery with the computer to give an accurate reading. The tumours were measured in three directions i.e. height, length, width and the maximum diameter in each direction was recorded. The maximum diameter of the tumour in any plane was between 15-38 mm. According to Fisch classification five patients were class C and three patients were Class D1 tumours. None of the eight tumours in this study increased in size during the follow-up period (Figure 1). Radiological improvement was seen in 5/8 (63%) patients. Reductions in the size of the tumours were slight to moderate.

Dose of radiotherapy

The maximum dose to the periphery of the tumour ranged between 16-25 Gray with 50% isodose curve. The first patient we treated in our centre received 20 Gray after him next five received 25 Gray and two other patients received 16 and 17 Gray respectively. In one patient (IB) the lesion volume was 7800 cm³ while the target volume was only 4300 cm³. This discrepancy is due to the fact that target volume was measured on an angiogram as our neuroradiologist felt more confident to do the planning on angiogram rather than CT where tumour was poorly localised. The other patient (ED) received a sub optimal dose, as tumour coverage was partial. The tumour was extending into the neck and it was not possible to encompass whole of the tumour due technical limitations. Symptomatic improvement was experienced in 6/8 (75%) patients, consisting of decreased intensity of tinnitus, vertigo, headache and patient generally feeling well.

Complications

Two patients showed neurological deficit after radiosurgery. One patient (No.3) presented to us with progressive facial weakness at the time of radiosurgery. His facial weakness continued to progress during the follow-up period, despite radiological control of the tumour. This patient had three operations for his glomus jugulare tumour 20 years ago and had also received 6000 rads of conventional fractionated radiotherapy after the surgery to the residual. It seems probable that his facial weakness is at least in part a late complication of the initial conventional radiation therapy (Figure 2).

One patient (No. 2) showed a grade 2 (House-Brackmann) facial weakness two and half years after radiosurgery but at her last follow-up her facial nerve function was normal. All eight patients are stable after radiosurgery and have not required any further therapeutic intervention for their irradiated tumours, although one patient who had multicentric tumours was operated for her glomus vagale tumour on the opposite side. After surgery she developed problems with her balance and needed gastrostomy for her swallowing difficulties.

Hearing

Subjectively only three patient had useful hearing at the time of radiosurgery. The hearing remained stable in two patients and in one there was a subjective improvement.

Discussion

Glomus jugulare tumours present a formidable management problem because of their location and persistent invasive growth albeit usually slow and indolent. Surgical treatment of these tumours often causes considerable morbidity because of the extensive nature of skull-based procedures. With technical advancement in the surgical field, high speed drills and better microscopes, the focus of glomus jugulare tumour surgery has drifted away from the issue of resectability, virtually all of the glomus jugulare tumours are resectable (86-94%)^{10,13-18}. Anand et al were able to claim complete resection of the tumour in 100% of cases.¹⁹ Nowadays the emphasis is towards the functional outcome.

Most of these patients are able to compensate for their progressive chronic loss of function in the lower cranial nerves but it is the acute onset of lower cranial nerve dysfunction after an operation which gives rise to more trouble. In the published surgical series the risk of post operative lower cranial nerve dysfunction is around 50-83%.^{10,13,15-19} The nerves most commonly effected are glossopharyngeal, vagus, accessory and hypoglossal (31-83%) with the need to repair the paralysed vocal cords in 19% and gastrostomy in 4%. Facial nerve weakness was seen in 16-23% of the cases, while the abducent nerve was affected in 6% of the cases. Hearing impairment or deafness was seen in 4-26%.^{8,10,14,20-22} After the operation a substantial number of the patients are unable to maintain the same kind of life style, as they were able to enjoy prior to surgery. Other surgical complication includes wound infections, meningitis and CSF leaks. Another consideration is the long hospital stay after the operation. In one series of 20 cases the post operative duration of hospital stay was between (8-67days)²³, as most of these (19-42%) needed adjuvant surgery such as vocal cords injection, tarsorrhaphy, cricopharyngeal myotomy, thyroplasty, gastrostomy and facial nerve anastomosis. Even after macroscopically complete resection these patients have to be followed-up for long periods as recurrence can occur after 20 years.^{6,14,22,24} Surgery is not without the risk of mortality. The reported mortality is in the order of 0-6%.^{1,10-12,13,15,18,19}

Glomus Jugulare tumour is not particularly radiosensitive and the presence of vital tumour cells can be seen long after radiotherapy.^{11,22} The changes after the radiotherapy consist of necrosis, oedema, hemosiderin deposition, fibrosis, and vascular changes consisting of myeloid degeneration of the vessel wall, intimal proliferation and thrombosis.²³ Many people believe that the effect of radiotherapy is through the devitalisation caused by the obliteration of the vessels. In the past conventional fractionated radiotherapy was used most for recurrent tumours, medically unfit patients or inoperable cases. The requisite dose is considered to be 45-50 Gray given over 4-5 weeks. The tumour control rates achieved in published series are between 69-100%.^{18,24-29} The goal of radiotherapy treatment is tumour control and treatment is considered a success if there is no tumour progression in the follow-up. Radiotherapy showed subjective improvements in the clinical symptoms of the patients in around 70%

of the cases²². However radiotherapy is not without complications, minor complications 7%, and severe complications like bone and brain necrosis around 2%.^{22,26} Then there is a small risk of secondary malignancy.¹¹

Embolization of the feeding vessels is usually used as preoperative intervention to reduce the blood flow through the tumour. The ascending pharyngeal artery is the main vessel to these tumours but the tumour can receive blood supply from many other vessels namely the stylomastoid artery, middle meningeal branches of external carotid, carotico-tympanic and meningeal branches of internal carotid or from the vertebral artery. Thus the complete obliteration of all the vessels is unlikely. As a preoperative intervention it was seen that it could decrease the amount of blood loss during the surgery by nearly 1500cc. In a published series the average blood loss in embolized patients was around 1122cc and in non-embolized patient it was around 2769cc. The average time for surgery was also consistently less in embolized patients. However contrary to the popular belief embolization didn't help in improved preservation of cranial nerves.^{22,30,31}

Embolization itself carries a significant risk of morbidity (10%)³⁰ the most significant complication associated with embolization are stroke and death. other neurological deficit can occur including cranial nerve palsies. Hitherto embolization as a sole treatment option has to be considered a palliative measure since revascularization of the embolized vessels following endovascular occlusion occurs in 30% of the cases.^{18,30,32} Recently intratumoral embolization has been tried successfully; George et al³² injected a mixture of N-butylcyanoacrylate, lipiodol and tungsten in a 72 year old man with minor swallowing difficulties, sixteen months later the patient is in good condition without any evidence of tumour progression.

Radiosurgery has proven beneficial for arteriovenous malformations so it was natural to try it for other lesions. To a greater extent than radiotherapy this patient friendly, precise single session narrow beam radiation therapy spares the normal brain tissue and effects are targeted to the tumour so the lesion gets a more effective dose. Radiosurgery has no reported mortality or acute morbidity. In the four published series of radiosurgery used for this purpose in English literature, no tumour has increased in size. Thus, hitherto, until now there is 100% tumour control without any mortality or acute morbidity. The largest series, the European multicentre experience, which includes six patients from our centre showed improvement in the neurological deficit in 19 patients out of the 52 patients followed over a long period of time. Permanent neurological complications were only seen in two patients. In one patient hearing and function of facial nerve deteriorated. The second patient also experienced progression in facial weakness.

Our early results are comparable with the other Centres, as tumour control has been achieved without significant complications. In our experience radiosurgery has proven to be a cost effective procedure as the patient is only hospitalised for one or two nights, unlike radiotherapy and surgery. As no acute morbidity is seen, the patients normal lifestyle is not affected, so there is no economic loss because of inability to work.

Conclusion

Radiosurgery has proven to be a safe treatment for glomus jugulare tumour, however five

year follow-up is not sufficient as glomus jugulare tumours are slow growing and recurrence after surgery has occurred even after 20 years of apparently "complete" cure.^{6,14,24} More time is necessary to establish the long-term effectiveness of stereotactic radiosurgery for the treatment of glomus jugulare tumour.

At first surgery seems to be the only treatment which appears to offer a cure to the patient as usually it is possible to excise the tumour, but cure at what expense? Radiosurgery has proven to be an effective treatment and can control the growth of tumour without significant complication. We believe it should be used not only in the patients who are medically unfit or with recurrent tumours, but also should be considered as a primary treatment in suitable cases.

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