Glomus Tumors: When does Controversy become Convention?

Alan G. Micco, MD, FACS
Department of Otolaryngology, Assistant Professor
Department of Neurological Surgery, Northwestern University Feinberg School of Medicine
Chicago, Illinois, USA

Introduction

Glomus tumors are rare benign neoplasms in the head and neck. Glomus tympanicum and glomus jugulare lesions are associated with the temporal bone. The usual presenting symptom is pulsatile tinnitus followed by hearing loss. There is no controversy with the management of glomus tympanicums as they are mainly treated with surgical removal. Controversy occurs in the management of glomus jugulare tumors because of the lower cranial morbidity associated with the surgical removal.

Materials and Methods

Glomus tympanicum and jugulare cases will be discussed. Work up and therapeutic management will be reviewed. In glomus jugulare cases, conventional surgery versus gamma knife stereotactic radiation will be discussed. Outcomes and complications will be reviewed.

Results

Glomus tympanicums respond well to surgical excision. Glomus jugulare surgery is commonly associated with dysphonia and dysphagia. Patients commonly require a short term feeding tube because of the risk of aspiration. While stereotactic radiation does prevent further growth of the lesion, it does not correct the pulsatile tinnitus or conductive hearing loss as well as surgical excision.

Conclusion

There appears to be significantly higher morbidity with surgery versus stereotactic radiation. Patients should be counseled appropriately, especially about the cranial nerve deficits. Also, they should express realistic expectations of the results particularly the resolution of the tinnitus and hearing loss.

Introduction

Rosenwasser initially described glomus tumors in 1945. He described them as a parangangioma centered around the jugular foramen. Glomus tumors affect one in 1.3 million people. The female to male ratio is 3:6:1. Glomus tumors can be bilateral and secrete metanephines in 2% of the cases.

Glomus tumors are classified by their position.

Middle ear presentation alone is classified as a glomus tympanicum. In the neck, along the great vessels, they are classified as glomus jugulares. If they arise at the jugular foramen, they are classified as glomus jugulares.

Classical surgery of glomus jugulare requires an extensive temporal bone dissection, as well as identification of the great vessels in the neck, and possible intracranial surgery if there is extension into the poste-
rior fossa. These operations can last hours, increasing ones anesthesia risks. Preoperative embolization has helped tremendously to control blood loss and to decrease the incidence of blood transfusion\(^1\). It has also been observed however that the embolization itself can lead to cranial nerve injuries due to devascularization.

The glomus jugulare tumors present the surgeon with the biggest dilemma, in that classical surgical treatment of this benign lesion often leads to co-morbidities of lower cranial nerve neuropathies. There have been multiple studies that have shown that the incidence of facial nerve paralysis is 25\%, and cranial nerves IX-XI up to 50\%. Also there have been reports of hypoglossal nerve injury. These complications can lead to the need for tracheotomy and feeding tubes in order to protect the airway.

Other morbidities include CSF leaks, up to 28\%. Also there can be significant blood loss associated with removal. This also means subjecting patients to the possible complications associated with blood transfusions.

It is sometimes a difficult decision to put patients through this operation when the cranial nerve morbidity is so high for benign disease. Several non-surgical therapies have been tried to treat glomus jugulars. These include embolization alone and external beam radiation. The results of these therapies alone never showed consistent control. The advent of gamma knife has provided the practitioner with a new instrument to treat these tumors. Multiple studies have shown the benefit of this treatment especially in slowing the growth of these tumors and preventing complications.

Gamma Knife is a form of stereotactic radiation. This is the sole stereotactic modality that will be discussed in this study. Lars Leksell first described GK in 1967. The concept is to deliver a highly concentrated dose of gamma radiation directly to the tumor without affecting the surrounding tissues. The radiation is delivered with the assistance of a stereotactic frame which is placed prior to the treatment. This treatment modality is well described in the literature.

Gamma knife was initially used by the neurosurgeons. In fact when gamma knife was first started for lesions of the temporal bone, it was met with some skepticism in the Neurology community. In 1996, we presented a case of severe complications from gamma knife treatment of an acoustic neuroma\(^2\). The patient had presented with severe lower cranial nerve injuries. At the time, the treatment protocol involved radiation levels that are now twice what is currently used. This was the likely cause of such severe complications.

As the protocols were refined, more and more patients with benign lesions such as acoustic neuromas began receiving gamma knife instead of having traditional open surgery. At this time the neurologists became more involved in the use of gamma knife. In 2004 Wackym et al. presented their long-term data of results of gamma knife performed by a neurologist\(^8\). The results were in fact quite good, and this lead to man more neurologists obtaining training and privileges for gamma knife. Certification requires that a physician be a neurosurgeon, neurologist, or radiation oncologist. They must take a course given by experiences professional, with approval of the manufacturer of the gamma knife unit. The individual then has to meet the credentialing of their gamma knife center.

The attractive aspect of gamma knife for treatment of glomus tumors is an effective way to stop its growth without causing any further cranial nerve neuropathies. This review will discuss the current literature as well as present our results with treatment of glomus tumors with gamma knife.

The literature is replete with articles extolling the virtues of this treatment. In the last few years there have been articles by Martin et al, Gerosa, Krych and Lim\(^6\)–\(^8\). One of the largest studies evaluating the safety of gamma knife around the jugular foramen was Martin et al. from the University of Pittsburgh Group\(^10\). They had follow-up up to 85 months on 35 patients with jugular foramen schwanommas. There was a variation
in treatment doses with the doses ranging from 14 to 30 Gy at the 50% isodose. All but two of the patients had tumors that either stabilized or regressed. However, the important aspect of this large study was that not only did some of the pre-existing cranial nerve neuropathies resolve, but not a single patient had a new neuropathy develop. This showed the safety of gamma knife around the jugular foramen. One would expect that the results for glomus tumors would be similar.

**Methods**

Gamma knife is a well-orchestrated therapeutic procedure that involves multiple specialists. Our center utilizes a neurosurgeon, neurologist, radiation oncologist and a physicist in the planning for each patient. All of the participants have certified training. A Northwestern, our center utilizes a Leksell C unit. We have no experience with any other stereotactic unit.

Since 1999 we have treated 12 patients with glomus jugulare tumors. There were 9 females and three males. The age range was 28 to 71 with six of the patients over 50 years old. Most tumors were treated up to 14 Gy although one of the early patients was treated to 30 Gy. All patient’s plan had the 50 % isodose line at the edge of the tumor. All patients received one treatment except for one patient who had two non-contiguous recurrences after traditional surgery.

**Results**

Of our patients, there was a range of tumors size from 1.5 to 2 cm up to 10 cm. We also had one patient that had two rounds of gamma knife for separate recurrences after resection of a large Fisch Class D2 intracranial glomus tumor. Like the other published studies, all of our patients showed no evidence of further growth and more importantly, no development of new cranial nerve neuropathies.

A typical gamma knife planning protocol is shown in figure 1. This shows serial sections through the tumor in an axial plane. The yellow line represents the 50 % isodose line.

Large tumors are amenable to gamma knife therapy. A 71 year-old patient presented with slurred speech

![Fig 1: Typical gamma knife treatment plan for glomus tumor](image-url)
and weakness in the right shoulder. She also complained of some minor difficulty swallowing. The presenting MRI is shown in figure 2. The patient opted for gamma knife verses open surgery. A large field was irradiated. The treatment plan is shown in figure 3. After the treatment the patient had no further cranial nerve neuropathies. There were no new cranial nerve neuropathies. This patient was treated in 2006 and has not shown any signs of regrowth. This case demonstrates that unlike acoustic neuromas, large glomus tumors can be treated primarily with gamma knife.

Gamma knife is also useful in cases of recurrent glomus tumors after open surgery. A 28 year-old male underwent a trans-mastoid resection of a large Fisch D2 glomus tumor. Pt did well with no cranial nerve neuropathies. One year later the patient experienced a recurrence of the pulsatile tinnitus. The MRI in figure 4 shows the first recurrence. The patient had his first recurrence appear low along the remaining portion of the jugular foramen. The treatment plan is shown in figure 5. He tolerated his first treatment well. One year later he noted more pulsatile ringing. A new MRI showed recurrence proceeding anteriorly, medial to the carotid. The patient had a second round of gamma knife session (figure 6) since it was in a different location and has done well since. There has been no further growth or any recurrence since that time.

All nine of our patients have shown no further growth of the tumor since their therapy. More importantly there had been no development of any new cranial nerve neuropathies. Compared to the open cases we have performed, the complication rate is much less. In the last five years, we have performed three open
cases for glomus jugulare. All of the patients had transient dysphagia. None required airway protection. Two of the three had a permanently paretic cord, but had good compensation by the other cord and did not require tracheotomy or prolonged feeding tube placement.

Discussion

Glomus tumors are a locally aggressive vascular paraganglioma. Patients usually present with pulsatile tinnitus and conductive hearing loss. Traditionally these tumors have been treated with preoperative embolization and open surgery. Unfortunately removal of these tumors has a high morbidity rate, particularly in respect to the lower cranial nerves.

Gamma knife radiation was rapidly adopted by the neurosurgeons. Initially neurotologists did not embrace this technology because there were several early reports of cranial nerve complications with gamma knife treatment of acoustic neuromas. Over time, the protocols were refined and we now see outstanding results with minimal complications.

Our data, along with that of many other groups, show that gamma knife radiation is a now an acceptable primary treatment for glomus tumors. We have shown good results with treatment protocols using up to 14 Gy to the 50% isodose line. This is not surprising since gamma knife is also a primary treatment for other intracranial vascular lesions including cavernomas and arteriovenous malformations. Because of the results with gamma knife treatment of glomus jugulare, and the lack of nerve cranial nerve neuropathies with the treatment, GK should be offered as an option other than surgery. We have to remember
that despite the potential for locally aggressive disease, this is still a benign tumor. So now, the controversy has become the convention.

References

1. Wiet RJ, Harvey SA, O’Connor CA. Recent advances in surgery of the temporal bone and skull base. South Med J. Jan 1993; 86 (1); 5-12.


