

Glomus Jugulare Tumors: Certain Clinical and Radiological Aspects Observed Following Gamma Knife Radiosurgery

*Wael A. Reda, *Khaled MF. Saoud, *Amr MN. Al-Shahaby,
**Khaled Abdel Karim

Departments of *Neurosurgery, **Clinical Oncology, Ain Shams University

ABSTRACT

Introduction: Glomus Jugulare tumors represent a great therapeutic challenge. Previous papers have documented good results with these tumors. However, the relationship between clinical improvement and tumor shrinkage has never been assessed. **Material and Methods:** There were 14 patients, 9 females and 5 males. The mean follow up period was 28 months (range 6 to 60 months). All the tumors except one were Fisch Type D and the mean volume was 14.2 cm³ (range 3.7 cm³ to 28.4 cm³). The mean prescription dose was 13.6 Gy (range 12 to 16 Gy). **Results:** No tumor has continued to grow. Eight are smaller and 6 unchanged in volume. Two patients with bruit have no improvement in symptoms. All the other 12 patients have symptomatic improvement of dysphagia in 5, dysphonia in 4, facial numbness in 3, ataxia in 3 and tinnitus in 2. Single patients have experienced improvement of vomiting, vertigo, tongue fasciculation, hearing, headache, facial palsy and an accessory paresis. One patient developed a transient facial palsy. Symptomatic improvement began commonly before any reduction in tumor volume could be detected. The mean time to clinical improvement was 6.5 months whereas the mean time to shrinkage was 13.5 months. **Conclusions:** Gamma Knife treatment of glomus jugulare tumors is associated with a high incidence of clinical improvement with few complications, using the dosimetry recorded here. Clinical improvement would seem to be a more sensitive early indicator of therapeutic success than radiological volume reduction. Further follow up will be needed.

Key words: Glomus jugulare tumor, paraganglioma, radiosurgery, gamma knife, tumor volume

INTRODUCTION

Glomus tumors are rare^(5,6,10,15) slow growing tumors. Surgical resection can be difficult and associated with significant morbidity as a result of the their location and nature. Thus non-operative treatment methods are an attractive alternative. Radiosurgical treatment has been reported on numerous occasions^(1,5,9,12,14,16,17,19) and the effectiveness of Gamma Knife surgery in the first few years after treatment has been well documented^(1,5,9,11,14,17,19). One aspect of management which has been recorded before is clinical improvement in the presence of unchanged tumour volume^(2,3,4,9). To date, this phenomenon has been recorded but never assessed. The

purpose of the current series is to record a series of patients and comment on the relationship between the clinical and radiological response after treatment and the relationship between these two responses.

MATERIAL & METHODS

There were 14 patients, 9 females and 5 males; selected from a total of 27 referred patients. Grounds for refusing a patient were as follows. In 8 patients the tumour was too large. In 2 patients the tumor was largely extracranial and inaccessible to the Gamma Knife. In two patients investigations were requested but the patients never returned. In one case metal clips placed at the time of surgery produced artifacts which made geometrically

accurate imaging impossible. The mean follow up period was 28 months (range 6 to 60 months). All the tumors except one were Fisch Type D and the mean volume was 14.2 cm³ (range 3.7 cm³ to 28.4 cm³). The mean prescription dose was 13.6 Gy (range 12 to 16 Gy). In three patients previous surgery had confirmed the diagnosis. In the remainder the diagnosis was based on MR findings and a typical angiogram with supply mainly by the ascending pharyngeal artery.

RESULTS

No tumor has continued to grow. Eight are smaller and 6 unchanged in volume. Two patients with bruit have no improvement in symptoms. All the other 12 patients have symptomatic improvement of dysphagia in 5, dysphonia in 4, facial numbness in 3, ataxia in 3 and tinnitus in 2. Single patients have experienced improvement of vomiting, vertigo, tongue fasciculation, hearing, headache, facial palsy and an accessory paresis. One patient developed a transient facial palsy. Symptomatic improvement began commonly before any reduction in tumor volume could be detected. The mean time to clinical improvement was 6.5 months whereas the mean time to shrinkage was 13.5 months.

Illustrative Case

This is an unusual case in view of the age of the patient who was only 16. He presented in April 2003 with epistaxis. He was packed and

cauterization was performed. After this he has developed continuous nodding of the head. He also had more or less continuous blinking. In May 2003 he had some sort of attack with unconsciousness without incontinence. He also complained of right sided retromastoid pain. On examination there was slight rotation and tilting of the chin to the left. There was a minimal right hypoglossal weakness with wasting and deviation but no fasciculation. There was a clearly visible cherry red tumor behind the eardrum on the right side. He was deaf in that ear. There was no vagus deficit. He was treated with 12 Gy to the 35% isodose with 91% cover and a conformity index of 1.22. An MRI showed the tumor and an angiogram showed that it was vascular and mainly supplied by the ascending pharyngeal artery with a contribution from the middle meningeal artery. The patient was treated on 9th July 2003 and the tumor appearance is shown in figure 1. The extension into the neck is the reason for a low prescription isodose because otherwise the tumor would be outside the reach of the Gamma Knife. The patient came to follow up at 6 months and at that time no visible change was visible on the MR images. Moreover the extent of the tumor in the middle ear seemed unchanged. However there was a clear clinical improvement. He reported that the blinking had ceased. His tongue was now normal and the tilting and rotation of the head had resolved.

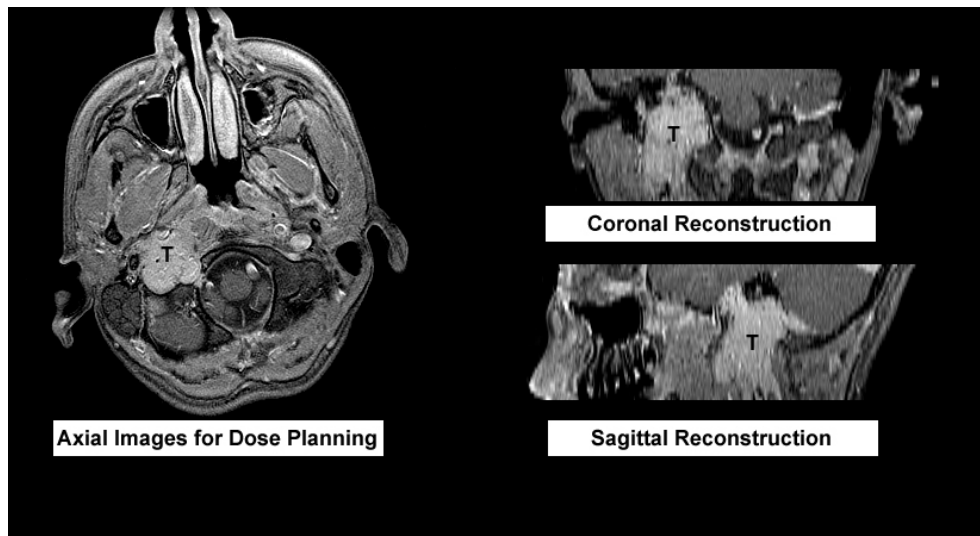


Figure 1 An axial image of the tumor with coronal and sagittal reconstructions. The head is rotated in the frame to bring the target closer to the center of the frame. The extension of tumor below the cranium requires the use of a lower isodose to treat it and in this case the 35% isodose was used.

DISCUSSION

This series can be criticised on two grounds. In the majority of patients there was no histological diagnosis and the series is selected. The absence of biopsy is unavoidable in many cases since these vascular tumors do not lend themselves to stereotactic procedures and open surgery even for biopsy could be a major undertaking and it was considered inappropriate to expose patients to such a procedure which was not aimed at therapy. The case selection is also unavoidable since at presentation not all glomus tumors are suitable for radiosurgery, mostly because of their size. Thus, it is emphasised that radiosurgery cannot be the only primary treatment for these tumors but that in cases where the tumor is of a suitable volume it represents an attractive alternative to surgery.

Glomus tumors are rare^(5,6,10,15,20) and mostly benign^(5,6,12,17). They produce lower cranial nerve symptoms which have a marked effect on quality of life^(5,10,12,14,17,20). The tumor entity

was first described by Valentin in 1840⁽¹⁸⁾. However, Guild⁽⁸⁾ proposed the term glomus tumor at a meeting of the anatomists in Chicago in 1941. The tumors are thought to arise from either the glossopharyngeal or vagus nerves and the cells contain chromaffin. The classical treatment has been surgery with or without radiotherapy^(5-7,10,12-14,17,19). Because of the location, local anatomy and vascularity of these tumors, post-operative complications in the form of new cranial neuropathy are not uncommon^(6,7,15,20). In addition, total removal is not always possible^(6,7,15,20). Moreover, these complications are compounded by the more usual complications of surgery such as hematomas, CSF leaks and infections^(6,7,15,20). Radiotherapy has been difficult to assess because of the slowness of the response of the tumors. However, there is evidence that low dose treatment is associated with recurrence^(7,13) and that high dose treatment may be associated with radiation induced complications^(5,7,10,19).

In view of the various difficulties associated with conventional

treatments it was natural to consider radiosurgery as a treatment modality and there are now a number of series recording good short term results^(1-5,9-14,16,17,19). The first paper recording the results of treating glomus tumors with radiosurgery was published in 1973. Because of the recent introduction of this technique for glomus tumor management it is unavoidable that late follow up results will only begin to appear at a future date. Nonetheless, the early results make this treatment modality attractive, providing it is born in mind that in common with the experience recorded in this paper, radiosurgery will always be limited to a selected group of patients and will never be appropriate for all patients.

Only passage of time will permit the registration of late follow up. However the tumor shrinkage recorded in most series of gamma knife treatment^(1,2,4,5,11,14,16,17,19) is in contrast to the lack of such shrinkage following radiotherapy¹². This suggests radiosurgery is a more useful technique for these tumors.

The current series is consistent with earlier reports in that eight tumors shrank while 6 remained unchanged^(2,4,5,9). On the other hand 10 patients experienced clinical improvement of a wide variety of symptoms. In addition the clinical improvement antedated radiological shrinkage in all the patients whose tumors showed shrinkage during the observation period of this study. This mismatch between clinical improvement and volume reduction has been reported previously in a number of papers^(2,3,4,9) but has never been analysed or made the subject of comment. If clinical improvement occurs before recordable changes in volume this could be a more reliable measure of early therapeutic success. In no patient in this series with clinical improvement has suffered a subsequent

deterioration. If this is confirmed in other studies it will be a useful assessment parameter in the management of these tumors.

The mechanism of clinical improvement in the absence of radiological improvement is not known and may be difficult to determine. Among the more likely explanations are inaccurate volume determination, changes in blood supply or changes in chemicals produced by the tumor. It is tempting to suggest that the most likely cause is imperfections in the measurement of changes in tumor volume. Precise tumor volume determination on MRI is in practice difficult or too time consuming to be undertaken routinely unless a given MR machine is equipped with an automatic segmentation protocol. This protocol is currently not included as a standard part of MR setup. This leaves the radiologist with the alternative of measuring distances across tumors and this is at best an approximation.

This series confirms that gamma knife surgery seems to be a useful treatment technique for selected glomus jugulare tumors. It also suggests that early clinical improvement may be a useful measure of long term therapeutic success, but longer follow up will be needed to confirm this notion.

Conclusion

Clinical improvement following gamma knife surgery for glomus jugulare tumors may, on further observation prove to be a reliable index of early therapeutic success, preceding shrinkage

REFERENCES

1. **Bari ME, Kemeny AA, Forster DM, Radatz MW.** Radiosurgery for the control of glomus jugulare tumours. *J Pak Med Assoc* 53(4): 147 – 151, 2003

2. **Eustacchio S, Trummer M, Unger F, Schrottnner O, Sutter B, Pendl G.** The role of Gamma Knife radiosurgery in the management of glomus jugular tumours. *Acta Neurochir Suppl* 84: 91 – 97, 2002
 3. **Foote RL, Coffey RJ, Gorman DA, Earle JD, Schomberg PJ, Kline RW, Schild SE.** Stereotactic radiosurgery for glomus jugulare tumors: a preliminary report. *Int J Radiat Oncol Biol Phys* 38(3): 491 – 495, 1997
 4. **Foote RL, Pollock BE, Gorman DA, Schomberg PJ, Stafford SL, Link MJ, Kline RW, Strome SE, Kasperbauer JL, Olsen KD.** Glomus jugulare tumor: tumor control and complications after stereotactic radiosurgery. *Head Neck* 24(4): 332 – 338, 2002
 5. **Gerosa M, Visca A, Rizzo P, Foroni R, Nicolato A, Bricolo A.** Glomus jugulare tumors: the option of gamma knife radiosurgery. *Neurosurgery* 59(3): 561 – 569, 2006
 6. **Gjuric M, Seidinger L, Wigand ME.** Long-Term Results of Surgery for Temporal Bone Paraganglioma. *Skull Base Surgery* 6(3): 147- 152, 1996
 7. **Gottfreid OB, Couldwell WT.** Comparison of radiosurgery and conventional surgery for the treatment of glomus jugulare tumors *Neurosurg Focus* 17(2): 22-30, 2004
 8. **Leber KA, Eustacchio S, Pendl G.** Radiosurgery of glomus tumors: midterm results. *Stereotact Funct Neurosurg* 72 Suppl 1: 53 – 59, 1999
 9. **Lim M, Gibbs IC, Adler JR, Chang SD.** Efficacy and safety of stereotactic radiosurgery for glomus jugulare tumors *Neurosurg Focus* 17(2): 68-72, 2004
 10. **Liscak R, Vladyka V, Wowra B, Kemeny A, Forster D, Burzaco JA, Martinez R, Eustacchio S, Pendl G, Regis J, Pellet W.** Gamma Knife radiosurgery of the glomus jugulare tumour - early multicentre experience. *Acta Neurochir (Wien)* 141(11): 1141 – 1146, 1999
 11. **Michael LM, Robertson JH.** Glomus jugulare tumors: historical overview of the management of this disease *Neurosurg Focus* 17(2): 1-5, 2004
 12. **Maarouf M, Voges J, Landwehr P, Bramer R, Treuer H, Kocher M, Muller R-P, Sturm V.** Stereotactic Linear Accelerator-Based Radiosurgery for the Treatment of Patients with Glomus Jugulare Tumors. *Cancer* 97(4): 1093-1098, 2003
 13. **Pollock BE.** Stereotactic radiosurgery in patients with glomus jugulare tumors *Neurosurg Focus* 17(2): 63-67, 2004
 14. **Ramina R, Maniglia JJ, Fernandes YB, Paschoal, JR, Pfeilsticker LN, Neto MC, Borges G.** Jugular foramen tumors: diagnosis and treatment *Neurosurg Focus* 17(2): 31-40, 2004
 15. **Saringer W, Khayal H, Ertl A, Schoeggl A, Kitz K.** Efficiency of gamma knife radiosurgery in the treatment of glomus jugulare tumors. *Minim Invasive Neurosurg* 44(3): 141 – 146, 2001
 16. **Sheehan J, Kondziolka D, Flickinger J, Lunsford LD.** Gamma knife surgery for glomus jugulare tumors: an intermediate report on efficacy and *J Neurosurg* 102 Suppl: 241 – 246, 2005
 17. **Varma A, Nathoo N, Neyman G, Suh JH, Ros SJ, Par KJ, Barnett GH.** Gamma knife radiosurgery for glomus jugulare tumors: volumetric analysis in 17 patients.
-

- Neurosurgery 59(5): 1030 – 1036, 2006
- 18. Watkins LD, Mendoza N, Cheesman AD, Symon L.** Glomus Jugulare Tumours: a Review of 61 Cases Acta Neurochir 130: 66-70, 1994
- 19. Guild SR.** The glomus jugulare, a nonchromaffin paraganglion in man. Ann Otol Rhinol Laryngol 62: 1045- 1071, 1993
- 20. Valentin G.** Uber eine gangliose Anschwellung in der Jacobsonchen Anastomose des Menschen. Arch Anat Physiolog Lpz 16: 287-290, 1840.
-