

Glomus Jugulare Tumors: Historical Overview of the Management of This Disease

L. Madison Michael II, M.D.; Jon H. Robertson, M.D.

Neurosurg Focus 17(2), 2004. © 2004 American Association of Neurological Surgeons

Posted 09/17/2004

Abstract and Introduction

Abstract

The treatment of glomus jugulare tumors presents the surgeon with a significant management problem. Because the neoplasm originates in the region of the jugular bulb, it frequently involves the lower cranial nerves, with occasional extension into the posterior fossa. Despite extensive work on the development of surgical and radiation treatment strategies, considerable controversy still exists regarding the optimal management of these lesions. A historical review of the development of management options for glomus jugulare tumors is presented in an effort to offer a foundation for understanding their contemporary treatment.

Introduction

The optimum treatment of glomus jugulare tumors remains controversial. Although they appear to be histologically benign, these tumors are characterized by unpredictable biological behavior. Typically arising from glomus bodies located in the dome of the jugular bulb, this neoplasm is intimately associated with critical neurovascular structures at the skull base. Successful treatment of glomus jugulare tumors requires a clear understanding of the anatomy of the jugular foramen and the natural history of these tumors.

Since the first reported exploration of the jugular bulb by Seiffert in 1934 (as reported in Lundgren^[31]), surgeons have been striving to find the optimal approach and technique to facilitate complete extirpation of glomus jugulare tumors. Earlier treatments focused on limited resection combined with radiation therapy or radiation therapy alone. The relative inaccessibility of the jugular bulb region, the potential for harming the surrounding neurovasculature, and the extremely vascular nature of these tumors dictated a treatment that was, for the most part, consistent with palliation.

As experience was gained, innovative surgical procedures were developed, most notably the combined lateral skull base approaches and the infratemporal approach. In addition to the advancements in surgical approaches, the implementation of adjunctive therapies, such as superselective embolization and gamma knife radiation therapy, and the aggressive treatment of postoperative neurological deficits have resulted in dramatic improvements in long-term outcomes.

In this paper we present a historical review of the development of management options for glomus jugulare tumors. The storied surgical history, especially in regard to the evolution of operative approaches, is reported. Re views of the use of selective embolization as an adjunctive therapy and radiation therapy as an alternative treatment modality are included.

Historical Overview

Surgical Treatment

The inaugural work on the glomus jugularis, or jugular body, was reported by Guild^[21] at the American Association of Anatomists meeting in Chicago in 1941. He described nests of "blood vessels of capillary or of pre-capillary caliber with numerous epithelioid cells between the vessels" located near or in the wall of the jugular bulb. In 1945, Rosenwasser^[36] was the first person to identify the possible relationship between the glomus jugularis, as named by Guild, and "carotid body like" tumors occurring in the temporal bone.

In 1953, Guild^[20] significantly enhanced our understanding of this entity when he published an anatomical study of 88 serially sectioned human temporal bones. He found the presence of glomus formations located only along two nerves: the nerve of Jacobson (tympanic branch of the glossopharyngeal nerve) and the nerve of Arnold (auricular branch of the vagus nerve). The distribution of the observed glomerula along the courses of the respective cranial nerves revealed a distinct proclivity for the adventitia of the jugular bulb, with more than half of the glomus formations located in this region. In light of Guild's discoveries, glomus tumors of the middle ear (glomus tympanicum) and those originating in the jugular bulb region (glomus jugulare) could be differentiated and explicated.

During the 1950s, several authors communicated their efforts to treat glomus jugulare tumors, with mostly discouraging results.^[3,31] The intricate anatomy of the jugular bulb region and the risk of hemorrhage from the tumor, in combination with the lack of high-definition imaging studies to elucidate tumor margins, were significant limitations. In 1951, Weille and Lane^[48] recommended the removal of bone surrounding the tumor to avoid bleeding. Their approach did not call for removal of the bulb because they thought the risk of hemorrhage

was too high. In the same year, Semmes^[39] treated a patient with a glomus jugulare tumor via a suboccipital approach, which he reported in 1953. Although he removed the entire tumor with in the posterior fossa, he made no attempt to remove the lesion from the mastoid or middle ear. The year after that operation Capps^[8] reported on five patients with glomus jugulare tumors. In one case the patient underwent extensive surgery, which involved mobilizing the facial nerve (not previously described), obtaining proximal and distal control of the sigmoid sinus and jugular vein, followed by an unsuccessful attempt to remove the jugular bulb. The eventful postoperative course of this patient led Capps to treat the remaining four patients with radiation therapy alone. In 1953, Albemaz and Bucy^[3] reported on a patient who presented to them with findings of jugular foramen compression and hearing loss. On opening the dura mater no tumor was seen, but the lower cranial nerves appeared to be abnormal. The patient suddenly suffered cardiac arrest on closing, and the autopsy revealed a 1 × 2–cm jugular foramen tumor. At the end of the decade, Meacham and Capps^[32] and Thoms, et al.,^[45] reported their experience with removing glomus jugulare tumors via a suboccipital route. This approach was believed to be sufficient for many glomus jugulare tumors; however, if there was extensive bone involvement, then radiation therapy alone was used. Despite a greater comfort level with the anatomy of this region, it was not until the introduction of more sophisticated imaging techniques that significant progress, with reduced morbidity and mortality rates, was made.^[49]

The early 1960s brought advances in diagnostic technology that enabled practitioners to observe in detail the surgical anatomy, which led to a higher degree of surgical accuracy. Arteriography,^[35] polytomography,^[46] and retrograde jugular venography^[17] allowed the surgeon to view the tumor and select the most judicious approach or treatment (computerized tomography scanning was not introduced until 1972 and magnetic resonance imaging was not available until 1977). In 1964, Shapiro and Neues^[41] reported their experience with a patient in whom a recurrent glomus jugulare tumor arose. They accomplished complete tumor resection, with removal of the jugular bulb and translocation of the facial nerve. Unlike earlier reports, there was minimal blood loss and the patient did well neurologically. Gejrot^[18] described a similar procedure performed in 1965 in a series of four patients.

These reports were important in that they established the foundation for current surgical techniques. They proved that tumor extirpation, along with removal of the jugular bulb and preservation of neural function, was possible. One critical contribution in Gejrot's approach, which persists as a crucial component of modern surgical treatment of glomus jugulare tumors, was that he stressed the importance of maintaining the medial wall of the sigmoid sinus at the level of the jugular bulb in an effort to protect the lower cranial nerves.

Selective approaches to preserve hearing were introduced, most notably by House and Farris, during the latter part of the 1960s. House^[25] described removal of glomus jugulare tumors with preservation of the osseous portion of the ear canal. Facial nerve translocation was not performed, which allowed the use of the facial recess and exposure of the hypotympanum for tumor extirpation. In 1967, Farris^[11] modified the endaural hypotympanotomy of Shambaugh,^[40] stressing a postauricular hypotympanotomy. This approach was very effective in small glomus tumors with medial extension, but it was not effective in tumors that involved the anterior and medial surfaces of the CA.

Further interest in and refinement of surgical techniques continued throughout the 1970s. In 1971, Kempe, et al.,^[29] published a report on the use of a suboccipital craniectomy along with the standard mastoidectomy in removal of a tumor involving both the temporal bone and the posterior fossa. Hilding and Greenberg^[24] reported a similar case in the same year, but included exposure of the internal CA through the glenoid fossa. In 1974, Glasscock, et al.,^[19] published their approach, which used a combination of Shapiro's wide exposure of the skull base and House's technique of using the extended facial recess. In 1977, Gardner, et al.,^[16] detailed a surgical technique in which a combined lateral skull base approach was used by a multidisciplinary team. Their approach consisted of the following three phases: 1) exposure of the skull base through the neck; 2) bone removal within the temporal bone and jugular fossa; and 3) tumor removal, with wound reconstruction to follow. In 1977, Fisch^[12] introduced the infra temporal approach to gain complete access to the internal CA within the temporal bone, the lack of which was a major limitation of the earlier approaches. This method modified the earlier extended hypotympanic approach popularized by Farris in the 1960s. The exposure and control of the CA allowed larger glomus jugulare tumors to be treated with increased safety. Furthermore, Fisch^[13] added a classification scheme for glomus tumors ([Table 1](#)), which he and Jenkins revised in 1981.^[27]

Over the next two decades, continued modifications of established approaches were introduced. In 1982, Jackson, et al.,^[26] stressed the importance of a team approach, with each specialist supervising his or her area of expertise. Similar to Gardner's conclusions, these authors believed that a "divide and conquer" approach offered the best chance for surgical success. In the same report, Jackson and colleagues introduced a new classification scheme for glomus jugulare tumors, recognizing the problem of intracranial extension ([Table 2](#)).

In 1987, Al-Mefty, et al.,^[1] described a combined infratemporal and posterior fossa approach for the removal of giant glomus jugulare tumors that have a large intracranial component. This approach provided access to tumors previously thought to be inoperable and obviated the need for multiple stages of surgery. In 1989, Bordi, et al.,^[4] described a similar modification of the infratemporal and lateral approach, which they called the "single-staged posterolateral combined otoneurosurgical approach." In 1994, Patel, et al.,^[34] published a report on a series of 12 patients with extensive glomus jugulare tumors in which the subtemporal–infratemporal, retrosigmoid, and/or extremelateral transcondylar approaches were used in combination with the usual transtemporal–infratemporal approaches described previously.

Al-Mefty and Teixeira^[2] reported in 2002 on the management of tumors of the glomus jugulare that they termed "complex." To be classified as complex, one or more of the following criteria had to be met: giant size, multiple paragangliomas, malignancy, catecholamine secretion, association with other lesions, previous treatment with adverse outcome, radiation therapy, or adverse effects from embolization. Similarly to Patel and colleagues, they stressed the modification of existing approaches to achieve adequate exposure of the lesion.

A major advancement in the surgical treatment of glomus jugulare tumors occurred with the development of preoperative superselective embolization. After it was introduced by Hilal and Michelsen^[23] and Brismar and Cronqvist^[7] in separate publications, in 1979 Simpson, et al.,^[43] reported the use of preoperative embolization in glomus jugulare tumors in an effort to reduce intraoperative blood loss. Murphy and Brackmann^[33] substantiated the use of preoperative embolization in a 1989 report in which 35 patients were analyzed. They concluded that there was a significant reduction in both intraoperative blood loss and operating time. In addition, embolization led to a higher rate of complete resection. Nevertheless, there did not appear to be a reduction in the risk of injury to the lower cranial nerves. As for the morbidity

associated with the embolization procedure, the current state of technology and expertise in interventional radiology has significantly reduced the incidence of stroke and cranial nerve injury experienced during the early years of its application.

Radiation Therapy

The treatment of glomus jugulare tumors with radiation therapy remains controversial. To date, there is no conclusive data establishing radiation as the optimal primary treatment for all glomus jugulare neoplasms. Spector, et al.,^[44] revealed in 1973 that radiation therapy had relatively little effect on the tumor cells, with the most dramatic changes consisting of a marked increase in the stromal fibrous connective tissue. Several other authors have reported that the primary effect of radiation therapy is a radiation-induced vascular injury.^[6,22] Furthermore, it has been shown that catecholamine secretion is not affected by the application of radiation.^[38] Nonetheless, glomus tumors treated primarily with radiation have been reported in multiple clinical studies describing excellent tumor control with only rare cases of tumor progression.^[5,30,42,47]

Two points must be made concerning the conclusions reached in these studies. First, an overwhelming number of patients were followed up for less than 5 years, and it is a well-known fact that recurrent tumors can arise up to 25 years after the initial treatment.^[9] The second point is that a majority of patients treated with radiation had no change in the size of their tumor. Regardless of the shortcomings of radiation therapy, surgical treatment involves the risks associated with induction of general anesthesia and carries the potential for cranial nerve injury. These problems are not encountered when fractionated radiation is used. For patients who are medically unfit for surgery, of an advanced age, or at significant risk for cranial nerve injury, radiation therapy may be of significant benefit.

In several recent studies the use of gamma knife surgery in the treatment of glomus tumors has been investigated. In 1997, Foote, et al.,^[14] published the first report as a preliminary study. The goal of their study was to evaluate the immediate, acute, and chronic toxicity and the efficacy of stereotactic radiosurgery in patients with unresectable or subtotally resected glomus tumors. No acute or chronic toxicity was demonstrated, and eight of nine tumors remained stable in size at a median clinical follow-up duration of 20 months. In 1999, Eustacchio, et al.,^[10] reported on 10 patients, with radiosurgery being used as the primary treatment in seven of them. The median follow-up duration was 37.6 months, with 60% of patients exhibiting no change in tumor size, and with the remainder showing decreased tumor volumes. In a series published by Jordan, et al.,^[28] in 2000, eight patients who were deemed unsuitable for surgery were treated with stereotactic radiosurgery. One patient experienced intractable vertigo requiring hospitalization. The mean follow-up duration was 27 months, and no patient had an increase in the size of the tumor.

Saringer, et al.,^[37] reported on 13 patients in 2001, with none showing evidence of tumor growth. Two patients experienced radiation-induced cranial neuropathies, both of which were transient. In a follow-up study to their 1996 series, in 2002 Foote, et al.,^[15] reported on 25 patients, 16 of whom were added after the first study. There was one episode of vertigo in their series, similar to the case reported by Jordan, et al.^[28] The median follow-up duration was 35 months, and no tumors exhibited growth during that time. As with most of the series involving fractionated radiation therapy, the follow-up period for these studies is short. We are awaiting future reports with longer follow-up times to demonstrate the efficacy of stereotactic radiation therapy in the management of glomus jugulare tumors. Nonetheless, the results are promising.

Conclusions

Glomus jugulare tumors pose a formidable challenge to the treating surgeon. Despite significant work on the development of surgical and nonsurgical treatment modalities, the optimum management of all glomus jugulare tumors remains controversial. We hope that this historical review of the management of glomus jugulare tumors will offer a foundation for understanding the contemporary treatment of this neoplasm.

Tables

Table 1. Fisch Classification of Glomus Tumors of the Temporal Region

Medscape® www.medscape.com	
Class	Description
A	tumors limited to middle ear space
B	tumors limited to middle ear or mastoid w/o involvement of the infralabyrinthine space of the temporal bone
C	tumors involving infralabyrinthine & apical spaces of temporal bone, w/ extension into the apex
D1	tumors w/ intracranial extension <2 cm in diameter
D2	tumors w/ intracranial extension >2 cm in diameter

Source: Neurosurg Focus © 2004 American Association of Neurological Surgeons

Table 2. Glasscock–Jackson Classification of Glomus Jugulare Tumors

Medscape® www.medscape.com	
Grade	Description
I	small tumor involving jugular bulb, middle ear, & mastoid
II	tumor extending under internal auditory canal; may have intracranial extension
III	tumor extending into petrous apex; may have intracranial extension
IV	tumor extending beyond petrous apex into clivus or infratemporal fossa; may have intracranial extension

Source: Neurosurg Focus © 2004 American Association of Neurological Surgeons

References

- Al-Mefty O, Fox JL, Rifai A, et al: A combined infratemporal and posterior fossa approach for the removal of giant glomus tumors and chondrosarcomas. *Surg Neurol* 28:423–431, 1987
- Al-Mefty O, Teixeira A: Complex tumors of the glomus jugulare: criteria, treatment, and outcome. *J Neurosurg* 97: 1356–1366, 2002
- Albernaz JG, Bucy PC: Nonchromaffin paraganglioma of the jugular foramen. *J Neurosurg* 10:663–671, 1953
- Bordi LT, Cheesman AD, Symon L: The surgical management of glomus jugulare tumours—description of a single-staged posterolateral combined otoneurosurgical approach. *Br J Neurosurg* 3:21–30, 1989
- Boyle JO, Shimm DS, Coulthard SW: Radiation therapy for paragangliomas of the temporal bone. *Laryngoscope* 100: 896–901, 1990
- Brackmann DE, House WF, Terry R, et al: Glomus jugulare tumors: effect of irradiation. *Trans Am Acad Ophthalmol Otolaryngol* 76:1423–1431, 1972
- Brismar J, Cronqvist S: Therapeutic embolization in the external carotid artery region. *Acta Radiol Diagn* 19:715–731, 1978
- Capps FC: Glomus jugulare tumours of the middle ear. *J Laryngol Otol* 66:302–314, 1952
- Cece JA, Lawson W, Biller HF, et al: Complications in the management of large glomus jugulare tumors. *Laryngoscope* 97:152–157, 1987
- Eustacchio S, Leber K, Trummer M, et al: Gamma knife radiosurgery for glomus jugulare tumours. *Acta Neurochir* 141: 811–818, 1999
- Farrion JB: Glomus tumors. Postauricular hypotympanotomy and hypotympanoplasty. *Arch Otolaryngol* 86:367–373, 1967
- Fisch U: Infratemporal fossa approach for extensive tumors of the temporal bone and base of the skull, in Silverstein H, Norrell H (eds): *Neurological Surgery of the Ear*. Birmingham, AL: Aesculapius, 1977, pp 34–53
- Fisch U: Infratemporal fossa approach to tumours of the temporal bone and base of the skull. *J Laryngol Otol* 92:949–967, 1978
- Foote RL, Coffey RJ, Gorman DA, et al: Stereotactic radiosurgery for glomus jugulare tumors: a preliminary report. *Int J Radiation Oncol Biol Phys* 38:491–495, 1997
- Foote RL, Pollock BE, Gorman DA, et al: Glomus jugulare tumor: tumor control and complications after stereotactic radiosurgery. *Head Neck* 24:332–339, 2002
- Gardner G, Cocke EW Jr, Robertson JT, et al: Combined approach surgery for removal of glomus jugulare tumors. *Laryngoscope* 87:665–725, 1977
- Gejrot T: Retrograde jugularography in the diagnosis of abnormalities of the superior bulb of the internal jugular vein. *Acta Otolaryngol* 57:177–180, 1964
- Gejrot T: Surgical treatment of glomus jugulare tumours with special reference to the diagnostic value of retrograde jugularography. *Acta Otolaryngol* 60:150–168, 1965
- Glasscock ME III, Harris PF, Newsome G: Glomus tumors: diagnosis and treatment. *Laryngoscope* 84:2006–2032, 1974
- Guild SR: The glomus jugulare, a nonchromaffin paraganglion, in man. *Ann Otol Rhinol Laryngol* 62:1045–1071, 1953
- Guild SR: A hitherto unrecognized structure, the glomus jugularis, in man. *Anat Rec* 79 (Suppl 1):28, 1941 (Abstract)
- Hawthorne MR, Makek MS, Harris JP, et al: The histopathological and clinical features of irradiated and nonirradiated temporal paragangliomas. *Laryngoscope* 98:325–331, 1988
- Hilal SK, Michelsen JW: Therapeutic percutaneous embolization for extra-axial vascular lesions of the head, neck, and spine. *J Neurosurg* 43:275–287, 1975
- Hilding DA, Greenberg A: Surgery for large glomus jugulare tumor. The combined suboccipital, transtemporal approach. *Arch Otolaryngol* 93:227–231, 1971
- House W: Discussion in McCabe BF, Rosenwasser H, House W, et al: Management of glomus tumors. *Arch Otolaryngol* 89:170–178, 1969
- Jackson CG, Glasscock ME III, Nissen AJ, et al: Glomus tumor surgery: the approach, results, and problems. *Otolaryngol Clin North Am* 15:897–916, 1982
- Jenkins HA, Fisch U: Glomus tumors of the temporal region. Technique of surgical resection. *Arch Otolaryngol* 107: 209–214, 1981
- Jordan JA, Roland PS, McManus C, et al: Stereotactic radiosurgery for glomus jugulare tumors. *Laryngoscope* 110:35–38, 2000

29. Kempe LG, VanderArk GD, Smith DR: The neurosurgical treatment of glomus jugulare tumors. *J Neurosurg* 35:59–64, 1971
30. Larner JM, Hahn SS, Spaulding CA, et al: Glomus jugulare tumors. Long-term control by radiation therapy. *Cancer* 69: 1813–1817, 1992
31. Lundgren N: Tympanic body tumours of the middle ear. Tumours of the carotid body type. *Acta Otolaryngol* 37:367–379, 1949
32. Meacham WF, Capps JM: Intracranial glomus-jugulare tumor with successful surgical removal. *J Neurosurg* 17:157–160, 1960
33. Murphy TP, Brackmann DE: Effects of preoperative embolization on glomus jugulare tumors. *Laryngoscope* 99:1244–1247, 1989
34. Patel SJ, Sekhar LN, Cass SP, et al: Combined approaches for resection of extensive glomus jugulare tumors. A review of 12 cases. *J Neurosurg* 80:1026–1038, 1994
35. Riemenschneider PA, Hoople GD, Brewer D, et al: Roentgenographic diagnosis of tumors of the glomus jugularis. *AJR Radium Ther Nucl Med* 69:59–65, 1953
36. Rosenwasser H: Carotid body tumor of the middle ear and mastoid. *Arch Otolaryngol* 41:64–68, 1945
37. Saringer W, Khayal H, Ertl A, et al: Efficiency of gamma knife radiosurgery in the treatment of glomus jugulare tumors. *Minim Invasive Neurosurg* 44:141–146, 2001
38. Schwaber MK, Gussack GS, Kirkpatrick W: The role of radiation therapy in the management of catecholamine-secreting glomus tumors. *Otolaryngol Head Neck Surg* 98:150–154, 1988
39. Semmes RE: Discussion of Alexander E Jr, Beamer PR, Williams JO: Tumor of the glomus jugulare with extension into the middle ear. *J Neurosurg* 8:515–523, 1951
40. Shambaugh GE Jr: Surgical approach for so-called glomus jugulare tumors of the middle ear. *Laryngoscope* 65:185–198, 1955
41. Shapiro MJ, Neues DK: Technique for removal of glomus jugulare tumors. *Arch Otolaryngol* 79:219–224, 1964
42. Sharma PD, Johnson AP, Whitton AC: Radiotherapy of jugulo-tympanic paragangliomas (glomus jugulare tumours). *Laryngol Otol* 98:621–629, 1984
43. Simpson GT II, Konrad HR, Takahashi M, et al: Immediate postembolization excision of glomus jugulare tumors: advantages of new combined techniques. *Arch Otolaryngol* 105: 639–643, 1979
44. Spector GJ, Maisel RH, Ogura JH: Glomus tumors in the middle ear. I. An analysis of 46 patients. *Laryngoscope* 83: 1652–1672, 1973
45. Thoms OJ, Shaw DT, Trowbridge WV: Glomus jugulare tumor: report of a case with surgical removal. *J Neurosurg* 17: 500–504, 1960
46. Valvassori GE: Laminography of the ear. Normal roentgenographic anatomy. *Am J Roentgenol Radium Ther Nucl Med* 89:1155–1167, 1963
47. Wang ML, Hussey DH, Doornbos JF, et al: Chemodectoma of the temporal bone: a comparison of surgical and radiotherapeutic results. *Int J Radiat Oncol Biol Phys* 14:643–648, 1988
48. Weille FL, Lane CS Jr: Surgical problems involved in the removal of glomus-jugulare tumors. *Laryngoscope* 61:448–459, 1951
49. Williams HL, Childs DS Jr, Parkhill EM, et al: Chemodectomas of the glomus jugulare (nonchromaffin paragangliomas) with especial reference to their response to roentgen therapy. *Ann Otol Rhinol Laryngol* 64:546–566, 1955

Reprint Address

Address reprint requests to: L. Madison Michael II, M.D., Department of Neurosurgery, University of Tennessee College of Medicine, 847 Monroe Avenue, Suite 427, Memphis, Tennessee 38163. email: lmichael2@pol.net

L. Madison Michael II, M.D., and **Jon H. Robertson, M.D.**, Department of Neurosurgery, The University of Tennessee at Memphis; and Semmes-Murphey Neurologic and Spine Institute, Memphis, Tennessee
