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Original Article

A Middle Path in the Surgical Management of Glomus Jugulare: Lessons Learnt from a Short Series

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Abstract

Introduction: Glomus jugulare are rare vascular tumors arising from the jugular bulb with intra- and extra-cranial extensions. Although considered benign, these tumors are locally invasive with involvement of critical neurovascular structures causing significant morbidity. Due to their highly vascular nature, they pose a challenge for surgeons. **Settings and Design:** This is a retrospective analysis of a short series of glomus jugulare managed in a tertiary state-run referral center. **Subjects and Methods:** Three patients (two females, one male) of ages ranging from 32 to 55 years were evaluated for glomus jugulare and surgically managed. In the first case, total petrosectomy after transposing the facial nerve, cerebellopontine angle exploration, neck dissection and infratemporal fossa approach including removal of the internal jugular vein were done. Case 2 and Case 3 were managed with subtotal resection. Radiotherapy (RT) for residual tumor was given in Case 2. **Results:** Case 1 had secondary hemorrhage on 8th postoperative day to which she succumbed on postoperative day 18. Case 2 and Case 3 have been comfortable with no fresh deficits after 36- and 6-month follow-up, respectively. Apart from this, the authors also enumerate the various "lessons" learnt from this series. **Conclusion:** Planned subtotal resection followed by RT or stereotactic radiosurgery for the residual tumor yields a better outcome with lower morbidity and mortality.

Keywords: *Glomus jugulare, planned subtotal resection, radiosurgery*

Introduction

Glomus jugulare are rare tumors which are locally aggressive with cranial, extracranial, and intracranial involvement.[1] Intracranial involvement is noted in one-third of these lesions.[1] Radical surgical excision is the only therapeutic modality which offers immediate and complete tumor elimination and cure.[1] Although technological development and refinement has yielded significant improvement in surgical results, morbidity rates continue to be high due to lower cranial nerve deficits, cerebrospinal fluid (CSF) leak, infection, prolonged anesthesia and operative time, postoperative ventilation, and tracheostomy complications.[1-4] Radiosurgery offers an attractive noninvasive therapeutic option which has not only been used as an adjuvant therapy for residual and recurrent lesions but also as a primary modality.[5] The authors present a short series of three cases of glomus jugulare treated surgically and the lessons learnt from this experience.

Subjects and Methods

The authors managed three cases of glomus jugulare which are discussed below in detail and also in Table 1.

Case 1

A 55-year-old female was admitted with diminished hearing, left-sided headache, and double vision on looking to the left of 2-year duration. For 6 months, she complained of change in voice and nasal regurgitation at times. On examination, she had left VI nerve palsy, left VII, IX, X, XII nerve paresis, and left VIII nerve deafness. Her imaging revealed a large highly contrast-enhancing lesion involving the jugular foramen and petrous apex with a large intracranial extension into the left cerebellopontine angle (CPA) and extending through the left sigmoid sinus into the left internal jugular vein (IJV) occluding both on the left side [Figure 1]. After a detailed discussion and meticulous planning which included a rehearsal of the surgery on a cadaver, the patient was taken up for preoperative embolization of the left ascending pharyngeal artery (which was the primary feeder) using polyvinyl

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Table 1: Clinical profile of patients in the pre- and post-operative periods

Patient	Side of the tumor	Preoperative deficits	Postoperative deficits	Deficits in the follow-up
Patient 1 (55 years, female)	Left	Left VI, VII, VIII, IX, X, and XII cranial nerve paresis	Left VI, VII, VIII, IX, X, and XII cranial nerve palsies	Not applicable (patient expired on 18th postoperative day)
Patient 2 (33 years, male)	Right	Horizontal gaze-evoked nystagmus; right IX, X, and XII cranial nerve palsies	Preoperative deficits remain, no fresh postoperative deficits	Significant improvement in lower cranial function at 3-year follow-up
Patient 3 (52 years, female)	Left	Left VII palsy, VIII deafness, IX, X, and XI cranial nerve paresis	Left VII, VIII, IX, X, and XI cranial nerve palsies	Improvement noted in VII, IX, X, XI cranial nerve function at 6-month follow-up

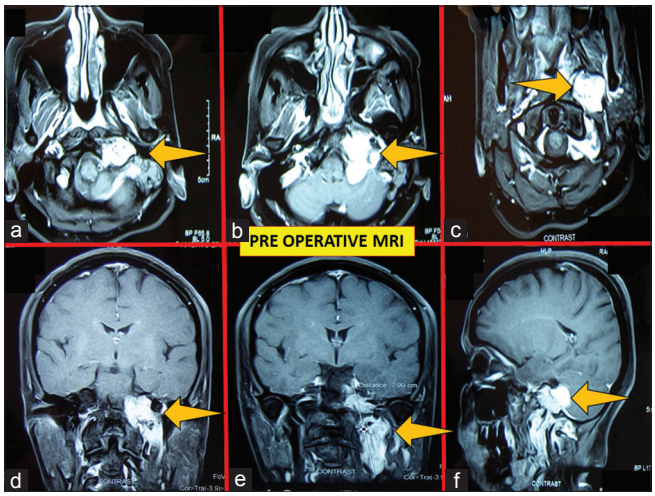

Figure 1: Preoperative contrast magnetic resonance imaging of patient 1: (a-c) are axial images, (d) and (e) are coronal, and (f) sagittal sections showing left glomus jugulare tumor (arrows)

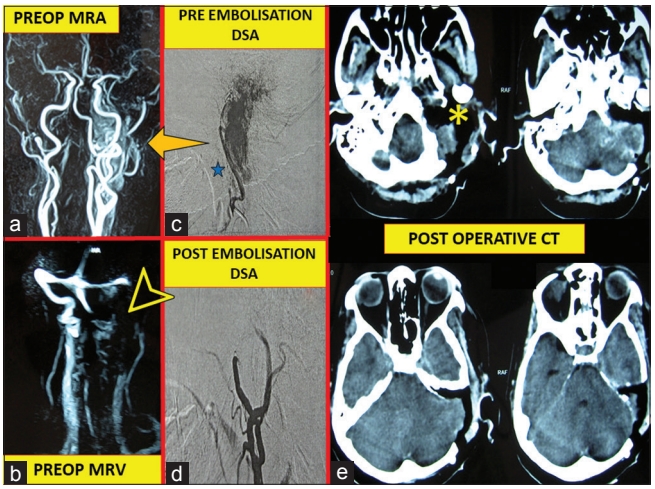

Figure 2: Preoperative and postoperative images of patient 1: (a) Preoperative magnetic resonance angiography showing tumor blush (arrow). (b) Magnetic resonance venography showing total obliteration of sigmoid sinus and proximal half of internal jugular vein (arrow head). (c) Preembolization digital subtraction angiography showing tumor being fed from ascending pharyngeal artery (star). (d) Postembolization digital subtraction angiography showing no tumor blush. (e) Postoperative computed tomography showing near-total petrosectomy (asterisk) with total excision of the lesion

alcohol (PVA) particles. Excellent embolization was achieved [Figure 2a-d]. The patient underwent a total resection of the lesion through a petrosectomy after transposing the facial nerve, CPA exploration and neck dissection and infratemporal fossa approach including removal of the IJV. The surgery lasted 20 hrs spread over 2 days, and the surgical team included neurosurgeons and head and neck surgeons. Postoperatively, the patient was conscious and alert and was extubated the next day. However, she was reintubated the same day. She had left VI, VII, VIII, IX, X, and XII nerve complete palsies. The lower cranial nerve injury occurred at the jugular foramen during excision. Tracheostomy was done, and she was weaned off the ventilator.

Case 2

A 33-year-old male presented with vomiting, hoarseness of voice, and difficulty in swallowing of 6-month duration. On examination, he had dysarthria, impaired convergence, horizontal gaze-provoked nystagmus, and right IX, X, and XII nerve palsies. Imaging revealed a contrast-enhancing lesion in the jugular foramen area with a large CPA extension and extension along the ipsilateral carotid sheath [Figure 3]. Preoperative embolization of the tumor was done using PVA particles. A right postauricular curvilinear myocutaneous flap was raised. A retromastoid craniectomy and mastoidectomy were done, and the sigmoid sinus was skeletonized. After opening the dura on either side of the sigmoid sinus, the latter was clipped and cut [Figure 4]. The large intradural portion of the tumor was excised. The VII–VIII nerve complex could be preserved [Figure 5]. However, the lower cranial nerves were injured at the jugular foramen. The entire IJV along with the tumor was excised up to the infratemporal fossa. Lumbar intrathecal drainage of CSF was done postoperatively along with tracheostomy and Ryle's tube feeding.

Case 3

A 52-year-old female presented with deviation of the angle of the mouth to the right and inability to close the left eye completely for 1 year and headache and diminished hearing in the left ear for 1 month. On examination, she had a left VII nerve palsy, left VIII nerve deafness, and left lower cranial nerve paresis. Imaging revealed a left jugular

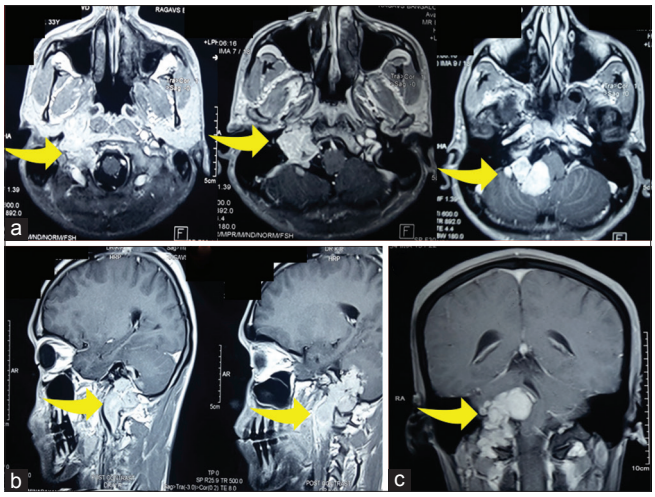

Figure 3: Preoperative images of patient 2: (a-c) Contrast-enhanced magnetic resonance imaging axial, sagittal, and coronal images, respectively, showing tumor in the right cerebellopontine angle with extracranial extension (arrows)

foraminal lesion with extension into the CPA and the middle ear [Figure 6]. After raising a myocutaneous flap, the left external carotid artery (ECA) was isolated and temporarily clamped. A left mastoidectomy was performed along with a retromastoid suboccipital craniectomy. The sigmoid sinus was ligated and cut. The CPA portion of the tumor was then excised along with the portion in the middle ear.

Results

Case 1

Postoperative CT scan revealed near-total petrosectomy with total excision of the glomus jugulare with no intracranial hemorrhage [Figure 2e]. On the 8th postoperative day, the patient developed bleeding from the wound site with sudden onset unconsciousness. She was immediately operated on and the wound reopened. There was a large extradural hematoma which was evacuated. The culprit was the left vertebral artery or one of its muscular branches. The bleeder was clipped and transfixed. No intradural extension of the hemorrhage was noted. She was continued on ventilation and steroids but failed to make any neurological improvement. She expired on the 18th day.

Case 2

Postoperative imaging confirmed major resection of the lesion with a residue in the petrous portion of the temporal bone [Figure 7a]. After 6 weeks, both the tracheostomy and Ryle's tube could be removed, and the patient was ambulant and comfortable, although the lower cranial nerve palsies persisted. The residual tumor was treated with Gamma knife (GK). On 3-year follow up, the patient is ambulant, comfortable with significant improvement in lower cranial nerve function. The recent MRI also shows decrease in size of the residual component [Figure 7b-d].

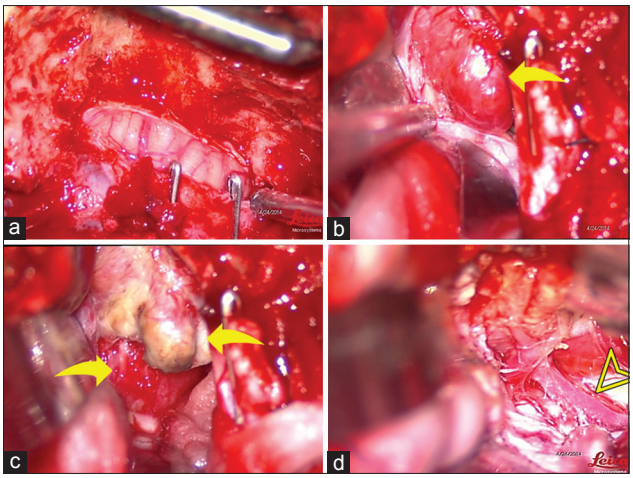

Figure 4: Intraoperative image of patient 2: (a) Dural opening and liga clip application of sigmoid sinus before cutting the sinus. (b) Vascular tumor (arrow) after dural opening in the cerebellopontine angle. (c) Tumor (arrows) being coagulated and separated from adjacent structures. (d) Vascular structures (posterior inferior cerebellar artery) (arrow head) separated from the tumor

Case 3

Postoperatively, the patient had lower cranial nerve palsies and left VII nerve palsy. However, she did not require a tracheostomy. Postoperative scan confirmed near-total excision of the glomus jugulare [Figure 6c]. On follow-up, she developed a persistently large pseudomeningocoele without any hydrocephalus. A thecoperitoneal shunt solved the issue. On 6-month follow-up, she was comfortable with no evidence of residual or recurrent lesion.

Lessons learnt from this series

- Glomus jugulare are rare and locally aggressive tumors
- Surgical excision of these lesions is challenging due to their high vascularity and multicompartment invasion.
- Surgical excision should be considered preferably in patients who already have lower cranial nerve paresis. These patients generally have already compensated from their normal side and hence are able to tolerate intraoperative injury to these nerves. In comparison, patients who develop lower cranial nerve palsy immediately after surgery have a stormy postoperative course
- Surgery should be preceded by meticulous planning and effective communication between all the team members
- Preoperative embolization of the glomus jugulare helps in surgery by reducing intraoperative bleeding and time significantly. If this facility is unavailable, then temporary clamping of the ECA after exposure and before excision also helps in reducing intraoperative blood loss
- Radical approaches in the form of total petrosectomy, transotic approach help in total excision of the glomus jugulare. These approaches involve considerable bone work and can be extremely time-consuming (as

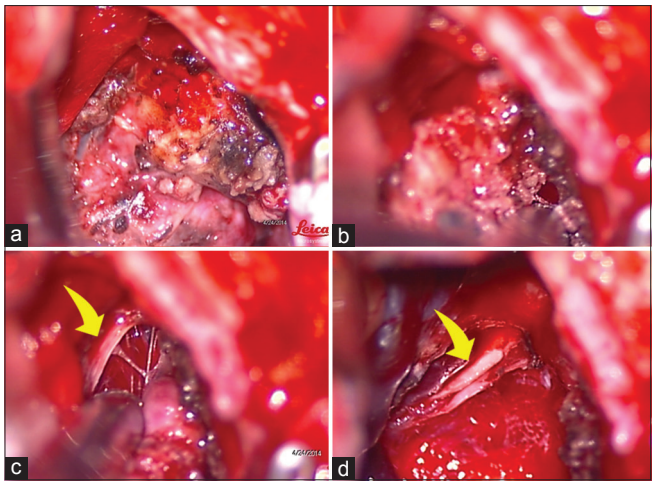

Figure 5: Intraoperative image of patient 2: (a and b) Tumor being coagulated and decompressed, (b) tumor being cut after coagulation at the jugular foramen laterally; (c) tumor being separated from VII–VIII cranial nerves (arrow), (d) VII–VIII cranial nerve complex (arrow) seen after complete excision of intracranial tumor

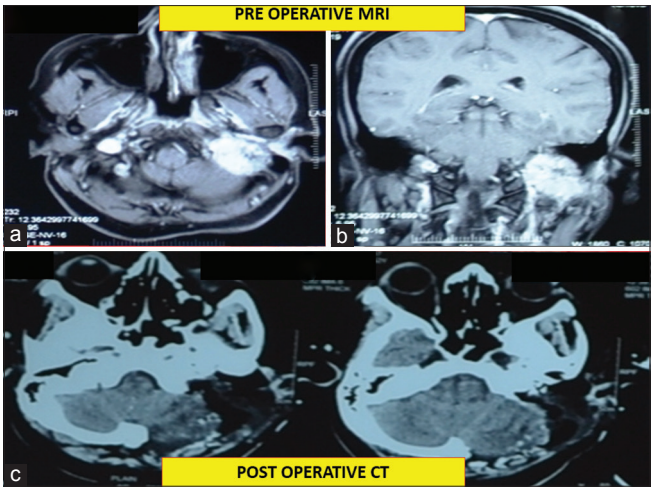

Figure 6: Preoperative and postoperative images of patient 3: (a and b) preoperative contrast-enhanced axial and coronal magnetic resonance imaging images. (c) Postoperative computed tomography shows near-total excision

experienced by us in the first case). These are also associated with higher morbidity rates in the form of intra- and post-operative hemorrhage, CSF leak, and infection

- Authors prefer to excise the intracranial (CPA) component and the extracranial portions of the lesion, leaving the petrous portion for radiosurgery. Excision of the CPA portion of the glomus jugulare helps decompress the brain stem and cranial nerves and thus eliminates any threat to life from the lesion. The extracranial portion is removed using the infratemporal fossa approach up to the distal limit of the lesion. This "middle path" helps reducing intraoperative bleeding and operative time. Moreover, postoperative morbidity is also reduced. Sometimes, the petrous portion is small and can be excised without extensive bone work
- A mastoidectomy is essential to reach the presigmoid dura. This is necessary for sacrifice of the sigmoid sinus whenever required
- It is important to close all the routes of CSF leak effectively. Authors use bone wax and fat graft with glue for this purpose. The authors feel that placing a lumbar intrathecal drain with regular aspiration of CSF postoperatively reduces the incidence of CSF leak and pseudomeningocoele
- Early tracheostomy if required, vigorous chest and limb physiotherapy, and early ambulation play important roles in ensuring a good result from this technically difficult surgery
- Timing of radiosurgery for the residual lesion and its execution is also important and should be discussed and planned with the radiation oncologist
- Proper preoperative counseling and regular follow-up are essential to help the patient, and the caregivers tide over these difficult times. Education of the caregivers for tracheostomy care, Ryle's tube feeds, etc., is important to ensure that the patient gets proper care at home. The authors take this seriously and have made a video to help educate caregivers which is also provided to them at discharge.

Discussion

Glomus jugulare tumors were first noted by Valentin in 1810 and then by Krause in 1878 but received attention only after Guild described it in 1941 and named it "glomus jugularis."[6] Some authors have used the term jugular Paraganglioma synonymous to glomus jugulare. The inaccessibility of the jugular foramen due to its deep location, high vascularity of these lesions, and proximity to vital neurovascular structures has made surgical excision of these tumors extremely challenging.[6] Rosenwasser was the first to document the description of the surgical excision of a glomus jugulare.[7] Although total excision was the goal, a subtotal excision followed by Radiotherapy (RT) was generally performed.[6] Transposition of the facial nerve which was an obstacle was first described by Capps in 1952. However, surgical outcome was poor. In the 1960s and 1970s, advances in technology resulted in better surgical outcomes. The main reasons for this were introduction of the operating microscope, bipolar cautery, microdissection techniques, various skull base approaches, better imaging (computed tomography CT and magnetic resonance imaging), superselective angiography, and preoperative embolization.[6] House, Glasscock, McCabe, Fletcher, Fisch, Al-Mefty, Bordi *et al*., Patel *et al*., and Liu *et al*. have all made significant contributions to the surgical management of these challenging lesions. Pareschi *et al*. have demonstrated excellent results with total excision of the lesion with no mortality or relapse, high percentage

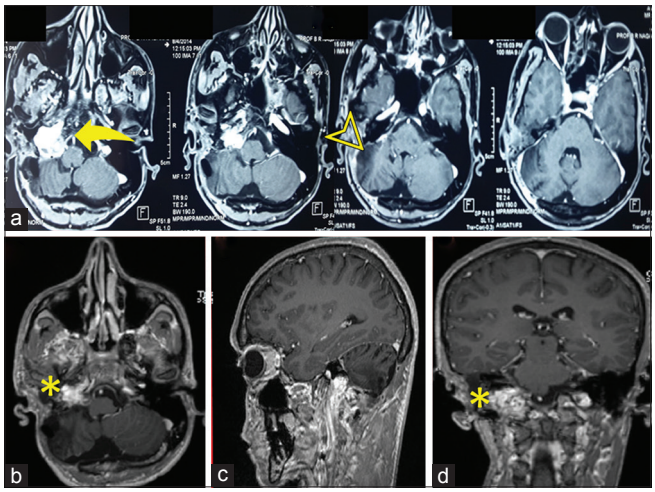

Figure 7: Postoperative images of patient 2: (a) Postoperative magnetic resonance imaging axial images showing residual lesion in the petrous bone (arrow) and complete excision of the intracranial (arrow head) and extracranial components of the lesion. (b-d) (axial, sagittal, and coronal cuts, respectively) showing reduction in tumor size following gamma knife irradiation (asterisk)

of lower cranial nerve preservation in smaller tumors, and a mean hospital stay of 14 days. However, intracranial extension of the tumor was noted in only ten patients in their group of 37 operated patients.[3]

Wanna *et al*. have adopted a policy of subtotal resection of these lesions with functional preservation of the lower cranial nerves. They concluded that when the postoperative residual tumor volume was <20%, no tumor growth was noted up to 45 months.[8] The medial wall of the jugular bulb was kept intact for anatomical and functional preservation of the lower cranial nerves in this study. In our series, Case 2 had <20% residual lesion which was submitted to GK surgery and had shown significant decrease in size in 36 months. The authors advocate consciously leaving behind some of the petrous portion of the tumor to avoid extensive bone resection and also the medial wall of the jugular bulb to avoid injury to the lower cranial nerves. However, sometimes, although anatomically preserved, these lower cranial nerves are functionally affected due to extensive use of bipolar cautery in the vicinity. The advantages of this "middle path" include less operative and anesthesia time, less blood loss, less lower cranial nerve morbidity, and early ambulation. Disadvantages include need for RT for the residual tumors and higher recurrence rates.

RT for these lesions has received considerable attention recently, especially with the introduction of stereotactic radiosurgery (SRS) in the form of GK, linear accelerator (LINAC), etc. It has been used for treatment of residual lesions and regrowths as well as a primary therapeutic modality with considerable success.[1,2,4] RT as the primary modality of treatment is considered in patients with advanced age, those with comorbidities constituting high anesthesia and surgical risk, large unresectable tumors, patients with multiple and bilateral tumors, patient's preference, and those without lower cranial nerve palsy.[1,4,9] Studies have compared outcomes of surgery, RT, and a combination of both (generally seen in the group which received RT for residual tumor). Meta-analysis revealed average local control rates of 78%, 93%, and 85%, respectively.[9] Suárez *et al*. have compared surgery to RT alone and found the latter to have significantly better outcome in tumor control, major complications, and cranial nerve palsies.[1] Tumor control rates have been reported between 93% and 97% using GK and LINAC.[1,2] Studies have compared conventional external beam RT with SRS and have found no statistically significant difference in their outcomes.[1,4] Complications of RT and SRS include hearing loss, osteonecrosis, brain necrosis, radiation-induced tumors, lower cranial nerve palsies, otitis media and externa and sometimes carotid artery stenosis.[1,4,5,9]

Conclusion

There is no consensus on the ideal management of glomus jugulare, and treatment has to be customized to the particular patient. Although complete resection is the ideal goal, poor surgical outcomes in larger glomus jugulare have compelled surgeons to adopt a more conservative philosophy toward these lesions. Planned subtotal resection followed by RT or SRS of the residual tumor yields a better outcome with lower morbidity and mortality.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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