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.
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

| 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 |

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

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 pseudomeningocele without any hydrocephalus. A thcoperitoneal 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 multicompartiment 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, transtotic approach help in total excision of the glomus jugulare. These approaches involve considerable bone work and can be extremely time-consuming (as
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 pseudomeningocele.

- 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.” 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. Rosenwasser was the first to document the description of the surgical excision of a glomus jugulare. Although total excision was the goal, a subtotal excision followed by Radiotherapy (RT) was generally performed. 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. House, Glasscoock, 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
Lessons learnt from glomus jugulare surgery

Radiosurgery of glomus jugulare

Asian Journal of Neurosurgery | Volume 14 | Issue 1 | January-March 2019

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Jugular and vagal paragangliomas: Systematic review of the literature and recommendations for management. (1,2,4) RT as the primary modality of treatment is considered in patients with advanced age, those with comorbidities constituting high an anesthesia and surgical risk, large unresectable tumors, patients with multiple and bilateral tumors, patient’s preference, and those without lower cranial nerve palsies. (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.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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