SURGERY OF JUGULAR FORAMEN LESIONS - AN INSTITUTIONAL EXPERIENCE

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Abstract:
Jugular foramen tumors are rare skull base tumors having complex neurovascular anatomy. Most common being paragangliomas, lower cranial nerve schwannomas and meningiomas. Being the conduit for important neuro-vascular structures, potential complications following surgery are a frequent source of morbidity. Retrospective study done from March 2008 to September 2014. All patients underwent high-resolution computer tomography (HRCT) of temporal bones and magnetic resonance imaging (MRI) with angiography in pre-operative period. Surgical approach and pre-operative endovascular intervention depended on the site and extent of the lesion. Extent of tumor removal was determined at the time of surgery. Patients with incomplete excisions in postoperative imaging were subjected to radiotherapy depending on histo-pathological diagnosis. Total of 22 patients of jugular foramen lesion were operated. Histopathological diagnosis included paraganglioma(n=18), schwannomas(n=2), plasmacytoma(n=1), squamous cell carcinoma(n=1). Eighth cranial nerve was the most common involved nerve preoperatively (100%). Infratemporal Fossa approach, along with its modification, and conservative jugulopetrosectomy approach were used for surgical excision as per the extent of tumor. Complete excision was done in 73% (n=16), and post-operative radiotherapy was given to rest of the patients. The most common postoperative complication was lower cranial nerve deficit in immediate post op period(n=15, 68%) (new onset n= 3, 13% and n=12, 55% had preoperative paresis), on follow up lower cranial nerve palsy improved in 50% in follow up (n=7). Most common tumor of the jugular foramen was paraganglioma followed by lower cranial nerve schwannoma. Surgical management depends on the extent of tumor. Multidisciplinary approach provides better patient outcome. Histopathological diagnosis aids to give targeted radiotherapy to the local site, and hence surgical excision of JFT (Jugular foramen tumor) is recommended.

Introduction:
Jugular foramen has a complex, variable anatomy between the petrous, sphenoid and the occipital bone. The lateral part (pars venosa) is filled by
sigmoid-jugular complex, while the medial part (pars nervosa) contains the inferior petrosal sinus, the cranial nerves 9th, 10th, 11th, the superior ganglion of the glossopharyngeal nerve and the jugular ganglion of the vagus nerve.

The jugular foramen can be involved by several varied pathologies ranging from glomus tumors, schwannomas, meningiomas, chordomas, chondrosarcomas, sarcoma, hemangiopericytoma, plasmacytoma temporal bone carcinoma and metastases. Out of these, paragangliomas are the most common lesion of the jugular foramen. Even among the most common lesions, there is no uniformity of opinion and no unequivocal guideline on the best therapeutic approach. None of the existing therapies like endovascular intervention, surgical approaches, radiotherapy, and chemotherapy can restore functions that have been lost due to pathological process.

The proximity of this foramen to nerves, arteries, veins, meninges, brain tissue and cochlea-vestibular system makes its surgical intervention a great challenge. The effectiveness of surgery compared with radiotherapy as the primary treatment in this area; especially for glomus tumors is a matter of controversy since a long time. Many centers across the world have shifted to radiosurgery as primary mode of treatment without histo-pathological diagnosis, which may vary in few cases. This article evaluates the importance of imaging and the role of surgery, histo-pathological diagnosis in the management of jugular foramen lesions.

**Material and methods:**
This retrospective study included 22 surgically managed patients between March 2008 and September 2014 with jugular foramen lesions in a tertiary care institute. Patients were reviewed according to their clinical presentations, radiological findings, surgical approaches, postoperative histopathology used and outcome following treatment. Minimum follow up was of 3 years and maximum 8 years.

Clinical evaluation included thorough survey of neuro-otological complaints and evaluation of 7th, 8th and lower cranial nerve function including pure tone audiometry. 7th cranial nerve function was graded according to House Brackman classification. Presence of lesions elsewhere was excluded with the help of physical examination and radiological investigation (USG abdomen). Urinary catecholamine levels were determined in all patients.

All patients were subjected to a high-resolution CT scan (HRCT) and MR imaging with contrast (gadolinium). A bilateral carotid angiography was done in all cases. Pre-operative Balloon Test Occlusion (BTO) was performed when the tumor appeared to involve the ICA (wall irregularities and stenosis of the arterial lumen). Diagnostic and therapeutic digital subtraction angiography (DSA) was done in all patients, to evaluate the feeders to tumors and for selective embolization.

**Surgery:**
A team of Neuro-Otologist and Neuro-Surgeon performed surgery.

**Infratemporal Fossa Approach A: (described by Fisch):** A ‘C’ shaped incision was given 5 cm behind the retro auricular groove, which extended in the neck region at the level of hyoid bone (Figure 1).
Figure 1: C shaped retro auricular incision 5 cm post to post aural sulcus with extension into neck, draped neck with ioban covering.

Skin, subcutaneous tissue and musculoperiosteal flap was elevated in two layers. (Figure 2)

Figure 2: Raised skin flap (SF), Musculoperiosteal flap (MPF), Anterior palva flap(P), Exposure of mastoid after raising IBF(inferiorly based flap) Cul-de-sac closure of external auditory canal was done in two layers (Figure 3).

Figure 3: Cul-de-sac closure
Neck control was taken by securing the internal carotid artery (ICA), external carotid artery (ECA) and jugular vein with umbilical tapes (Figure 4).

Figure 4: Exposure of neck done with controls over ECA(external carotid artery) and ICA(internal carotid artery) Sternoceleidomastoid muscle was delineated and separated from mastoid tip. Mastoidectomy was done and mastoid was drilled till sinus plate, dural plate and sino-dural angle were delineated. Further drilling was done flush to facial nerve canal. Tympanic membrane, malleus and incus were removed. Facial nerve was identified and exposed from geniculate ganglion till the stylomastoid foramen. Posterior belly of digastric was cut. Mastoid tip was amputated, and facial nerve was freed from stylomastoid foramen keeping a part of soft tissue surrounding it. Facial nerve was completely dissected from its canal and was transposed anteriorly (Figure 5).
Conservative jugulopetrosectomy:

The cases in which tumor was confined to jugular foramen with minimal extension into middle ear this approach was used. The initial steps of the surgery are same as infratemporal fossa approach, but here, instead of radical mastoidectomy, a wide cortical mastoidectomy was done. An extended facial recess approach was used to enable visualization of tumor. Further the tympanic bone in the antero-inferior region was drilled to expose the lower part of infra-tubal ICA. Jugular bulb if involved was packed with gelfoam and care was taken not to injure its medial wall. Keeping the boundaries of sigmoid sinus, and jugular bulb the tumor was excised from skull base.

Surgical approach depended on the site and extent of the lesion. Extent of tumor removal was determined at time of surgery, followed by a contrast MRI within two months of follow up. We used the following grades to describe the completeness of tumor removal:

Grade 1. Total resection: no residual tumor.
Grade 2. Near total resection: more than 95% of tumor removed
Grade 3. Sub-total resection: more than 50% of tumor removed

All the excised tumors processed for detailed histo-pathological examination and the tumor character analyzed. Only those patients with incomplete excisions with pathologically radio-sensitive tumors were subjected to fractionated radiotherapy. Further follow up included MRI at 6 months, 1 year after surgery and every 2 years thereafter.
Results:
Total no of operated patients was 22. The mean age of the patients was 36.9 years (range 5 to 62) with a female preponderance (M: F=0.6:1). The most frequent otologic symptoms (chart 1)

Chart 1: Symptomatology

were hearing loss (n=22, 100%), followed by tinnitus (n=15, 68%), mucopurulent or hemorrhagic otorrhoea (n=10, 45%) and hoarseness of voice (10, 45%). We observed “red reflex”, a pathognomic sign of glomus jugulare in four patients (18.%) and three patients (14%) had tumor mass protruding into the external auditory canal. (Figure 6)

Figure 6:

Figure 6: Right sided image T1 contrast MRI showing contrast enhancing mass in jugular foramen extending into EAC, left end image shows endoscopic picture of EAC with tumor (external auditory canal), TM-tympanic membrane. Middle image shows CT head with post-operative cavity.

Preoperatively, facial nerve dysfunction of various House-Brackman grades was noted in 9 (41%) patients. Seven (32% N=22) patients presented with symptoms of dysphagia. Six patients (27% N=22) had associated gait disturbances. Three (14%) patients presented with headache and one (5%) patient had concurrent visual deterioration and recurrent vomiting. One (5%) patient had a catecholamine-secreting tumor and presented with severe headache attributable to uncontrolled hypertension.

All patients had preoperative single or multiple cranial nerve deficits. Eighth cranial nerve (n=22, 100%) was the most frequently affected nerve followed by the lower cranial nerves (n=12, 55%), the seventh cranial nerve (n=9, 41%), and the fifth cranial (n=3, 14%) nerve in descending order (Chart 2).

Chart 2: Preoperative cranial nerve deficits associated with 22 jugular foramen tumors (n=22)

One patient had history of previously operated carotid body tumor. One (5%) patient had cavernous sinus involvement, 4 patients (18%) had extension into the clivus while one patient (5%) had extension to foramen magnum on MR imaging (figure 7).

Figure 7: T1 contrast MRI images showing tumor in jugular foramen. Middle cranial fossa, foramen magnum, Right IJV

U.P. Journal of Otorhinolaryngology & Head and Neck Surgery  
Vol. 8, Special Edition, April 2020
Average tumor size was 4.0 cm (7 patients had tumor size more than 5 cm)
Based on angiography, tumors were mainly supplied from ascending pharyngeal artery, followed by posterior auricular artery (Figure 8).

Of the 22 patients who underwent surgery, the facial nerve was preserved in 4 cases, infiltrated by tumor and therefore deliberately transected in 4 cases, accidentally transected after tumor removal in 2, and permanently transposed in 12 cases. The overall rate of anatomical preservation of the facial nerve was 73 % (16 patients). A sural nerve grafting was performed in 2 cases. In 2 patients, with long-standing paralysis the facial nerve was not repaired. Lower cranial nerve schwannoma did not show involvement of facial nerve. In 2 patients, facial nerve was repaired by end-to-end anastomosis after accidental transection. Lower cranial nerve was preserved if not grossly involved by tumor. Manipulation of lower cranial nerve during tumor dissection and hemostasis led to new onset cranial nerve palsy in 3 patients. 12 patients which already had lower cranial nerve palsy had some worsening.

Complete grade 1 excision was done in 16 patients (72%), a Grade 2 excision was done in 5 patients (23%) and in one case (5%) a subtotal grade 2 excision done because of malignant histopathology. In all 6 patients where complete excision was not done underwent radiotherapy in postoperative period (5 glomus jugulare and and 1 squamous cell carcinoma).

The histopathology revealed glomus jugulare tumors in 18(n=22, 82%), schwannomas in two (9%) and metastatic squamous cell carcinoma & plasmacytoma in one each case (5%) (chart 3).

Chart 3: Histopathological distribution of jugular foramen lesion(n=22)
The estimated average blood loss was 600 ml (range 0.5 to 2 L). Five patients (22.72%) developed wound bulge and 5 patients (22.72%) had CSF leak from operative site. All of them
responded to conservative treatment. One patient (4.45%) of CSF leak had developed meningitis, which responded to higher antibiotics as per culture and sensitivity. Eighteen (82%) patients developed facial palsy in immediate postoperative period. The improvement in facial nerve function at 6 months is shown in Table 1.

**Table 1: Post-operative Facial Nerve Functional Outcome in 22 patients at 6 months**

| House Brackmann Grade | Left In Canal (n, %) | Anterior Transposition (n, %) | Sural Nerve Graft (n, %) | End to End Anastomosis (n, %) | Long Standing Paralysis (
| n, %) |
|-----------------------|----------------------|-----------------------------|-------------------------|-------------------------------|-----------------|
| I                     | 4 (100%)             | 6 (50%)                     |                         |                               |                 |
| II                    | -                    | 3 (41.6%)                   | -                       | -                             |                 |
| III                   | -                    | 1 (8.3%)                    | -                       | 1 (50%)                       |                 |
| IV                    | -                    | -                           | 2 (100%)                | 1 (50%)                       |                 |
| V                     | -                    | -                           | -                       | -                             |                 |
| VI                    | -                    | -                           | -                       | -                             | 2 (100%)        |

As mentioned in this table; all patients except only those two with H & B grade IV facial palsy did improve of facial paresis at the end of six months following surgery. Fifteen (68%) patients had fresh or worsened lower cranial nerve function after surgery(Table 2) and needed nasogastric feeding in immediate postoperative period. But seven patients out of these (31.81%) could start taking orally after two months.

**Table 2:**

<table>
<thead>
<tr>
<th>New Cranial nerves deficit</th>
<th>n</th>
<th>Patients (%)</th>
<th>N=22</th>
</tr>
</thead>
<tbody>
<tr>
<td>5th</td>
<td>1</td>
<td>4.5%</td>
<td></td>
</tr>
<tr>
<td>7th</td>
<td>9</td>
<td>40.9%</td>
<td></td>
</tr>
<tr>
<td>8th</td>
<td>0</td>
<td>0 (0)</td>
<td></td>
</tr>
<tr>
<td>Lower Cranial Nerves</td>
<td>3</td>
<td>13.6%</td>
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Tracheostomy was done in 2 patients (9.09%) as they had developed acute respiratory distress and prolonged ventilator support was anticipated (beyond 5-7 days).

On long term follow up only one patient with Glomus Jugulare where complete excision was done had reoccurrence 3 years after surgery and subsequently underwent radiotherapy. The patient with squamous cell carcinoma died due to metastasis 1 year after radiation. Rest (n=20) did not had any recurrence in follow up. One patient who underwent surgery and radiation therapy for residual tumor has new onset lower cranial nerve palsy despite tumor being stable in MRI.

**Discussion:**

Clinical presentation of all jugular foramen tumors constitutes hearing loss, tinnitus along with lower cranial nerve palsies. Larger tumors may also be indicated by nonspecific symptoms (e.g., headache, increased intracranial pressure) or symptoms due to compression of adjacent structures such as the cerebellum and brain stem (e.g., nystagmus, ataxia, disequilibrium, long tract signs). The differentiation of vascular and non-vascular tumors of jugular foramen requires a high degree of suspicion. Proper clinical examination and imaging studies in preoperative period can help us to reach a probable diagnosis.

However, we cannot confirm the histopathology of the tumor unless we perform a biopsy. The jugular foramen tumors include cholesteatoma, paraganglioma, metastasis, squamous cell carcinoma, schwannoma, meningioma, plasmacytoma, chondrosarcoma, chordoma, neurofibroma and hemangiopericytoma and arterio-venous malformations. Careful analysis of imaging features and clinical correlation can help to reach a specific diagnosis; however, these tests are not completely reliable for definitive diagnosis. In our experience, Glomus jugulare tumor was hypointense on T1, isointense on T2, with multiple flow voids within it. On MRI with gadolinium
enhancement, the tumor shows marked homogeneous enhancement. High resolution CT temporal bone revealed erosion of petrous bone, carotid canal, facial canal, external ear, middle ear, inner ear, mastoid bone and jugulocarotid spine. Our findings are comparable to findings of weber et al and Mafee et al.21,22. However in one of our cases with similar characteristics the surgical excision biopsy revealed the diagnosis of schwannoma.

The JF schwannomas appeared as smooth-marginated, round, mass, hypointense on T1-weighted images and hyperintense on T2-weighted sequences with enhancement after gadolinium. On HRCT, JFSs showed regular erosion of the bony margins of the JF without infiltration.

Both plasmacytoma and squamous cell carcinoma are destructive lesion of temporal skull base.

The complex anatomy of the skull base makes the jugular fossa lesions inaccessible clinically and surgically. The spread of tumor along the routes of least resistance namely the carotid and fallopian canal, along with intra-dural extension makes complete surgical excision indispensable.

The most common lesions of JF are glomus tumors. Sanna et al. has reported a series of 32 patients with non-vascular lesions of the jugular foramen namely lower cranial nerve schwannoma and meningioma. Vogl and Bisdas have stated that metastases, usually from primary tumors in the lung, breast, and prostate give rise to destructive lesions that infiltrate the JF.23. The first case report on skull base plasmacytoma was reported by Chappell and Mathers.24. Literature review of temporal bone plasmacytoma by Chiang et al. suggest that plasmacytoma are destructive lesions and can extend from petroclival region to skull base.25.

The surgical approaches in jugulare foramen depend on the extension of tumor. If the tumor is extradural with extension into jugulare foramen, infralabyrinthine and apical compartments of petrous bone, the vertical segment of the internal carotid artery, and the upper jugulo-carotid space, an infratemporal fossa approach type A is recommended. As this approach requires anterior transposition of facial nerve for better exposure, immediate facial palsy in postoperative period is common. However, facial nerve function gradually improves over a period. In our study out of 12 patients, who underwent anterior transposition of facial nerve, 50% had grade 1 facial nerve function at 6 months of postoperative period. This finding is comparable to study of Pareschiet al.26.

If the tumor is extending till cavernous sinus, engulfing internal carotid artery, and till vertebro-basilar system, subtotal resection followed by radiotherapy in postoperative period is recommended. This view is in consistence with findings of Matthew et al.27.

Tran Ba Huy et al. concluded in his comparison study of 88 patients that radiotherapy and surgery achieve similar oncological outcomes, but the former achieves tumor control with less morbidity.28. Radiation therapy acts by vascular endothelial injury to the vessel walls. Hence, the blood flow of the tumor is gradually decreased leading to cell necrosis. The long-term results of radiation therapy are still awaited.29,31. Much controversy remains regarding the management of jugular foramen lesions, especially glomus jugulare, in view of surgery and current advances in radiotherapy. Recent advances in Stereotactic radio surgery (SRS) can achieve similar results compared to surgery for paraganglioma or glomus tumors.32. However, as mentioned earlier it is important to confirm the histology before subjecting the patients for radiotherapy or radiosurgery.

Intermediate term results of SRS shows that new onset cranial nerve palsy are seen in few patients pertaining to involvement of hearing due to exposure of cochlea to radiation and trigeminal neuralgia and lower cranial nerves.33. Spina A, et al concluded that longer follow up and larger cohort study are needed to outline role of SRS in Glomus
Therefore, we recommend surgical debulking followed by radiotherapy if required, in any jugular foramen lesions. Surgical approach can help us diagnose other suspected lesions in this complex area and targeted radiotherapy can be ensured. Also, primary radiotherapy in large JF tumors can cause considerable brain stem injury, further deteriorating patients' clinical condition. Moreover, it is prudent to obtain histopathological confirmation before submitting the patients for radiotherapy because as we observed in not all cases the tumors came out to be radiosensitive glomus tumors, instead some may prove to be a benign schwannoma or a highly malignant squamous cell tumors requiring multimodal treatment. The major limitation of this study is small sample size and lack of long term follow up. The comparison of vascular and non-vascular tumors group is not possible due to small sample size.

Long term surgical results are comparable with SRS as primary modality with therapy and less chances of new onset cranial nerve palsy later on as seen in SRS treatment for glomus jugular lesions9.

Conclusions:

All tumors of jugular foramen are not paragangliomas and should not be send for SRS as primary modality treatment, whereas most common jugular foramen tumor is glomus jugular paraganglioma. The most common clinical presentations in our series were hearing loss and tinnitus. Postoperative morbidity of patients was related to the extent of the tumor, involvement of cranial nerves and brainstem. Multidisciplinary approach and a judicious subtotal excision in complex jugular foramen lesions can prevent postoperative morbidity and the surgery helps to prevent the new onset cranial nerve deficit if complete excision is done. Histopathological diagnosis aids to give targeted radiotherapy to the local site, and hence surgical excision of JFT is recommended.

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**How to Cite this Article:**
https://doi.org/10.36611/upjohns/se/2020/1

**Conflict of Interest:** None

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