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Jugular Foramen Tumor; Experience in Tertiary Care Center

Dr B Hayagriva Rao[1], Dr P Prahaladu,[2] Dr Kadali Satya Vara Prasad[3], Dr B. V S Raman[4], Dr K Krishna Kishore,[5]

[1]*associate Professor Department Of Neurosurgery Andhra Medical College*
[2]*assistant Professor Department Of Neurosurgery Andhra Medical College*
[3]*professor And Head Of Department Department Of Neurosurgery Andhra Medical College*
[4]*senior Resident Department Of Neurosurgery Andhra Medical College*
[5]*professor And Head Of Department Department Of Otorhinolaryngology Andhra Medical College*
[7]*assistant Professor Department Of Neurosurgery Andhra Medical College*
Corresponding Author:Dr P Prahaladu

Abstract:
Objective
Jugular foramen tumour is a locally invasive, tumor, which grow slowly and causes various symptoms such as pulsatile tinnitus and lower cranial nerve palsy. Complete surgical resection is regarded as the ideal management of these tumors. The goal is to study the clinical characteristics and most effective surgical approaches for jugular foramen tumour.
Methods
Retrospective analysis of 7 jugular foramen tumour ,patients who underwent surgical resection between 2011 and 2017 was performed. Clinical records were reviewed for analysis of initial clinical symptoms and signs, audiological examinations, neurological deficits, radiological features, surgical approaches, extent of resection, treatment outcomes and complications.
Results
Most common initial symptom was hearing loss, followed by pulsatile tinnitus. Seven out of 7 patients had at least one lower cranial nerve palsy. Total of 7 operations took place in 7 patients. Total resection was achieved in 5 cases, when partial resection was done in 2 cases. Two patients with partial resection received gamma knife radiosurgery (GKS). No mortality was encountered and there were few postoperative complications.
Conclusion
Neurologic examination of lower cranial nerve palsy is crucial since most patients had at least one lower cranial nerve palsy. All tumors were detected in advanced stage due to slow growing nature and lack of symptom. Angiography with embolization is crucial for successful tumor removal without massive bleeding. Transmastoid retro auricular approach can be considered as a safe, satisfactory approach for removal of jugular foramen tumours. In tumors with intracranial extension, combined approach is recommended in that it provides better surgical view and can maintain the compliance of the patients.

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I. Introduction

The anatomy of the jugular foramen (JF) is complex.(FIGURE 1) Its intracranial aperture courses antero- laterally and inferiorly to the extracranial opening at the skull base and contains the posterolateral pars venosa and antero- medial pars nervosa, partially separated by the jugular spine of the temporal bone and sometimes completely by a fibrous or bony septum.Traditionally, the pars venosa contains the jugular bulb, from the sigmoid sinus, along with the vagus (X) and spinal accessory nerves (XI) and the posterior meningeal branch of the ascending pharyngeal artery, which runs between cranial nerves X and XI. Conversely, the pars nervosa includes the glossopharyngeal nerve (IX) and the inferior petrosal sinus. Lesions involving the JF exhibit significant diagnostic and surgical challenges. A large variety of lesions may develop or be identified in the JF. These lesions arise from structures within the foramen or from adjacent structures [1]. The most frequent intrinsic lesions are glomus jugulare (GJ) tumors, schwannomas of the lower cranial nerves, and meningiomas [2,3].we retrospectively reviewed 7 patients who had lesions involving the JF. The clinical presentation, diagnoses, management, and outcome (including complications) are discussed.

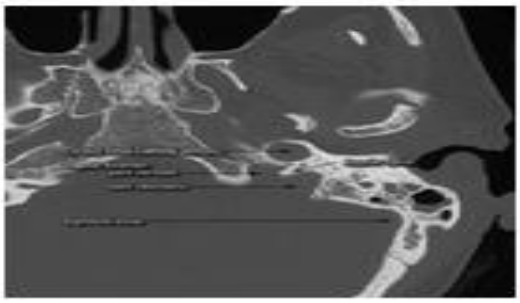


FIGURE 1

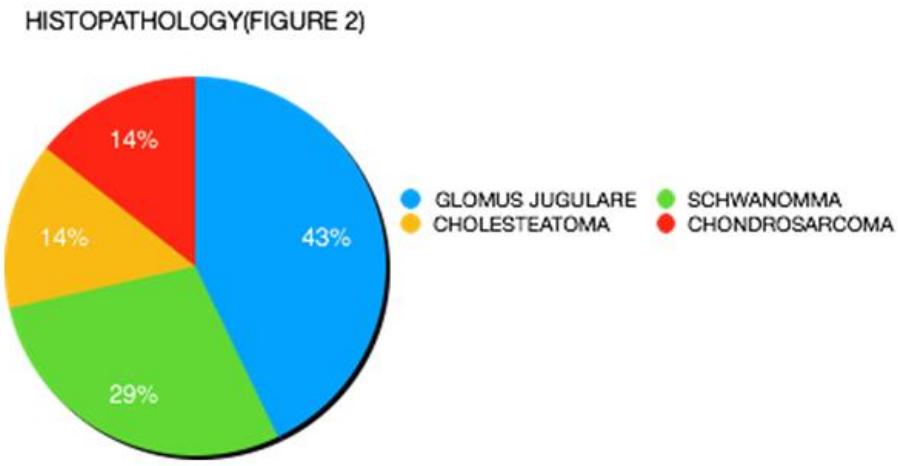
II. Materials and Method

The charts of patients who had lesions involving the JF that were operated on by the senior author from 2011 to December 2017 were reviewed. The data were reviewed specifically for age, sex, presenting symptoms, results of the neurologic examinations, location and extent of the lesions, surgical management, postoperative outcomes, and follow-up findings. The extent of lesion removal was determined by the intraoperative findings and radiologic follow-up. Postoperative complications were recorded. The preoperative clinical assessment included neurologic examinations, hearing tests, facial nerve electroneurography (ENoG), and assessment of vocal cord function. Temporal bone high-resolution computed tomography (HRCT) and magnetic resonance imaging (MRI) were performed for the radiologic diagnosis. Digital subtraction angiography (DSA) was performed for diagnosis and to guide the preoperative embolization in cases of highly vascularized lesions. Facial nerve function was graded according to the House-Brackmann (H-B) and Burres-Fisch scales. Intraoperative facial nerve monitoring was conducted. The follow-up consisted of periodic clinical evaluations with pure-tone audiometry (PTA) and HRCT and/or MRI scans.

III. Results

Demographic characteristics

The sample consisted of 7 patients diagnosed with lesions involving the JF. Among them, 3 patients had GJ tumors, 2 patients had JF schwannomas, and 1 patient had cholesteatoma and 1 patient had chondrosracoma(FIGURE 2). The mean age of the 7 patients was 31.05 years (range 20–60 years). Four cases (55.26%) were left-sided and 3cases (44.74%) were right-sided. There were 4 (55.26%) females and 3 (44.74%) males. The mean follow-up time was 28 months (range 2–98 months).

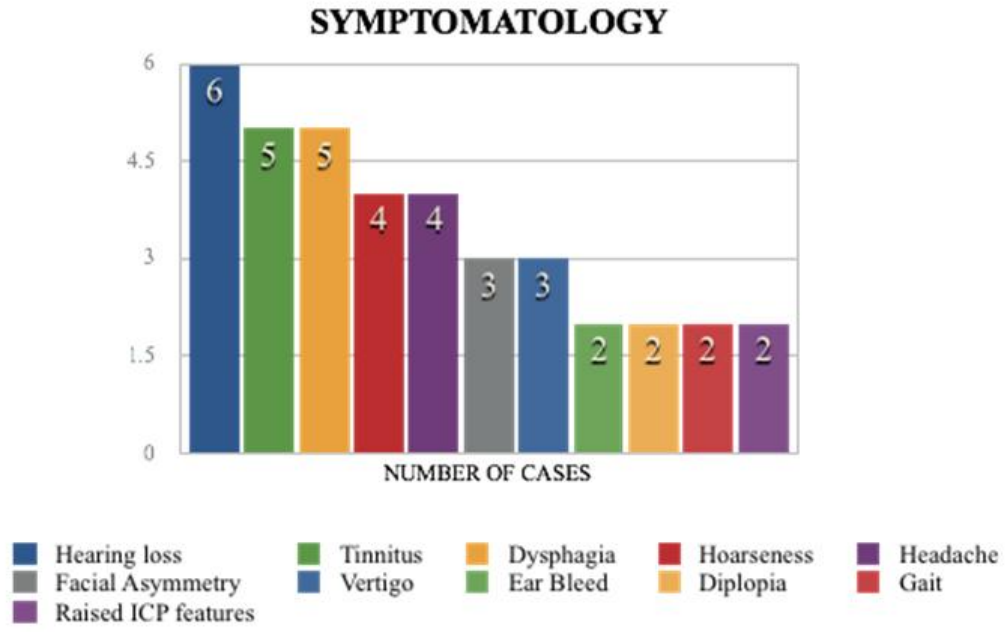


Clinical manifestations

The most frequent symptoms included conductive hearing loss followed by otorrhea, pulsatile tinnitus, otalgia, and facial nerve paresis (FIGURE 3).The preoperative examinations identified seven patients with deficits of the lower cranial nerve.

Treatment

All of the patients in this series were operated on by the senior author. Gross total removal of the lesions was achieved in 5 of the 7 cases and confirmed by postoperative serial radiological evaluation. Subtotal removal of the lesion was accomplished in one GJ tumor due to extensive bleeding during the operation, and one JF schwannoma due to the giant tumor. We applied the POSTAURICULAR TRAS MASTOID WITH CERVICAL approach for proximal control as no pre operative emboolization and with anterior facial nerve rerouting . The tumors were completely excised in 5 of the 7 patients; subtotal removed was attained in 1 case due to extensive bleeding. 2 cases received radiotherapy postoperatively. One of the patients had a preoperative ENoG reduction of 83%, although his facial nerve function was normal (H-B grade I) with occasional facial spasm. During the operation, the mass was found to surround the mastoid segment of the facial nerve. To achieve total tumor removal, the facial nerve was sacrificed, and then a greater auricular nerve graft was performed.



Complications

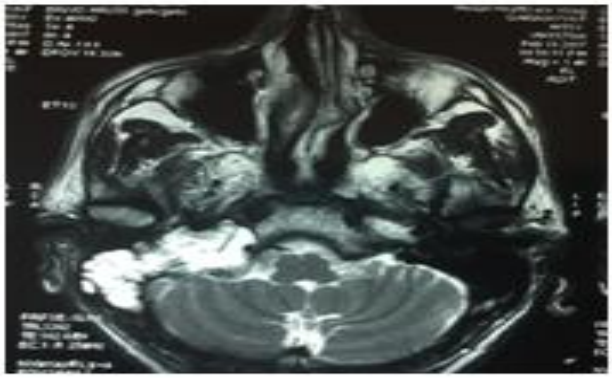
Two patients, including a facial nerve schwannoma case experienced postoperative cerebrospinal fluid leakage, which was managed successfully using a conservative strategy. No patients developed new lower cranial nerve palsy postoperatively.

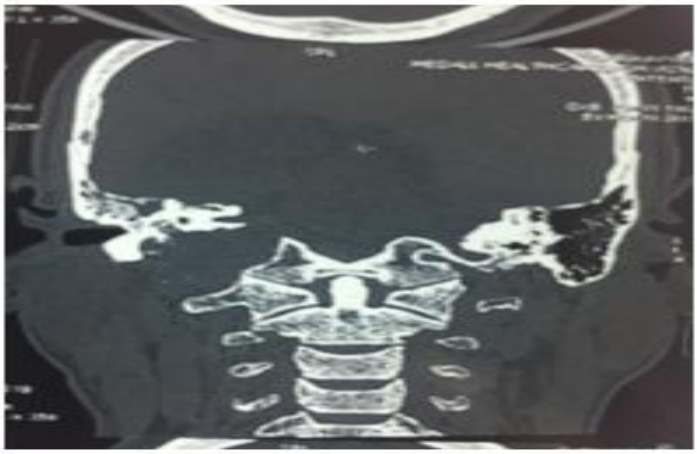
Prognosis :

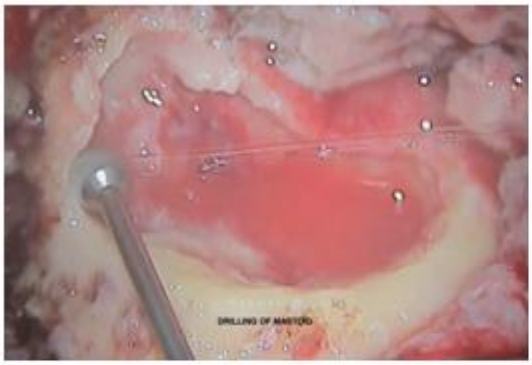
GJ tumor and JF schwannoma with subtotal removal, no obvious growth was revealed by clinical and radiological follow-up. No recurrence or death was noted in the other patients.

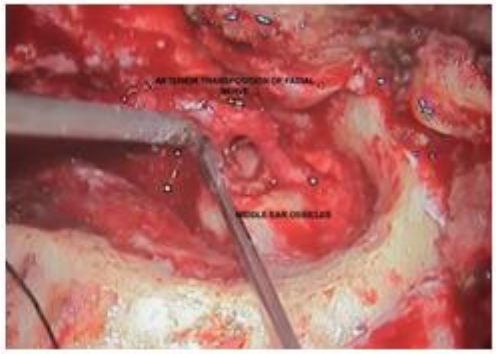
ILLUSTARTIVE CASES:

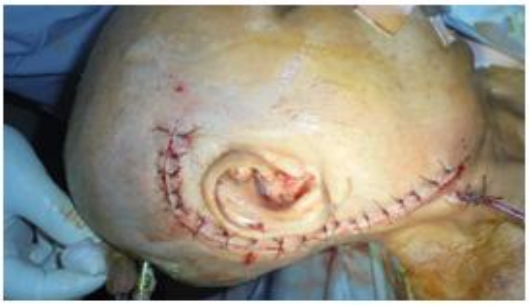
Case 1:Pre Operative Mri Contrast Brain Rt Schwannoma Of Jugular Fossa Operative Ct Bone Window Showing Widening Of Jugular Fossa On Rt Side. Trans Mastoid With Neck Dissection

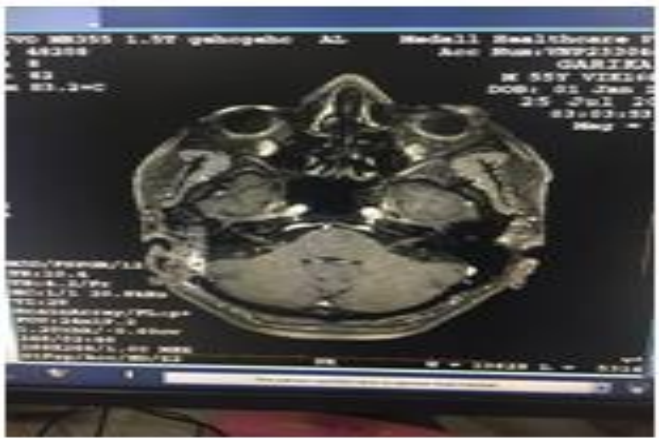












POST OP MRI BRAIN SHOW COMPLETE EXCISION OF TUMOUR

IV. Discussion

Paragangliomas of the Jugular Foramen

In 1941, Guild4 described carotid body–like structures in the temporal bone and named them "glomus jugulare bodies." In 1945, Rosenwasser5 published the first description of a middle ear paraganglioma and associated these tumors with the glomus jugulare bodies. In 1962,Alford and Guilford classified these lesions as glomus tympanicum and glomus jugulare tumors. Multiple terms have been used to describe paragangliomas—glomus tumor, chemodectoma, glomerocytoma, nonchromaffin tumor, tympanic body tumor, and receptoma are the most common.6

Chemoreceptor cells are nonchromaffin-staining para- ganglion cells developed from the neural crest region during embryogenesis.7 These structures are found in the adventitia of the jugular bulb beneath the floor of the middle ear, in the bone walls of the tympanic canals related to the tympanic branches of the glossopharyngeal and vag- us nerves, in the bone of promontory, close to the mucosal lining of the middle ear, in the ciliary ganglion, the ganglion nodosum of the vagus nerve, and in the walls of large arteries.8 The arterial blood supply to these bodies comes from the ascending pharyngeal artery through its inferior tympanic branch. The histological features of these bodies are the same in the glomus caroticum and glomus aorticum; histologically, paragangliomas are sim- ilar in all locations. Familial cases present with a much higher incidence of multicentricity;9 in these cases the paternal allele is the determining factor.10 There is a 50% incidence of male carrier offspring and 0% female carrier offspring with paraganglioma. This genetic abnormality is created by mutations located on chromosome 11 at 11q13 and q2. The most frequent initial symptoms are pulsatile tinnitus, hearing loss, and cranial nerve involvement. Diagnosis is usually made by ear, nose, and throat surgeons, and neu- rosurgeons usually see patients with large intracranial ex- tensions. Hypertension, headache, arrhythmias, nausea,and palpitations occur when the tumor secretes norepine- phrine. Perioperative treatment with -blockers is neces- sary for these patients.

Other Jugular Foramen Tumors

Schwannomas of lower cranial nerves, meningiomas, chondrosarcomas, and high-grade malignant tumors are other lesions originating in the jugular foramen. Hearing loss and paralysis of lower cranial nerves are often the ini- tial symptoms of these tumors. Most jugular foramen schwannomas are benign, noninfiltrative lesions.11 Men- ingiomas arise from arachnoid cells in the jugular bulb and infiltrate surrounding bone and nerve tissue.12 In this series all patients with meningiomas presented with lower cranial nerve deficits and those with malignant lesions had pain in the ear and mastoid region. Clinical signs and symptoms may not, however, accurately indicate the ex- tension of the lesion.

Surgical Treatment

Surgical exposure of this region requires understanding of the complex jugular foramen anatomy. 13 Anatomical studies have been conducted but the anatomy of this structure remains poorly understood.14Classically, the jugular foramen is described as consisting of two portions: the nervous part, containing the hypoglossal nerve, the inferior petrosal sinus, and the meningeal branches of the ascending pharyngeal artery; and the venous part, containing the sigmoid sinus and the vagus and accessory nerves. Anatomical studies have shown that the arrangement of the neurovascular structures within the jugular foramen does not conform to the compartmentalization into an anteromedial pars nervosa and a posterolateral pars venosa.

Katsuta, et al.,divided the jugular foramen into three portions: two venous and one nervous (intrajugular). The nervous portion is localized between the two venous ones and it contains the ninth, 10th, and 11th cranial nerves. The course of the cranial nerves within the jugular foramen presents many anatomical variations. Sen, et al.,described the cranial nerves running anteromedially to the jugular bulb and maintaining a multifascicular histoarchitecture (particularly the 10th cranial nerve). The vagus nerve is formed by multiple fascicles and the glossopharyngeal and accessory nerves by one and two fascicles, respectively. The lower cranial nerves (ninth, 10th, and 11th) run through a septum of connective tissue in continuity with the duramater and the pericranium. The tympanic branch of glossopharyngeal nerve (Jacobson nerve) and the auricular branch of the vagus nerve (Arnold nerve) cross the jugular foramen.

Preoperative evaluation includes complete otological and neurological examination, CT scanning, MR imaging, MR angiography, and digital subtraction angiography in cases of highly vascularized lesions. Balloon test occlusion is performed if the ICA is involved or encased. A high-flow shunt (saphenous vein or radial artery graft) is used to reconstruct this vessel. Preoperative embolization is performed in cases of highly vascularized tumors, and is useful to reduce bleeding and surgical time.15

Resection of these lesions is difficult for the following reasons: jugular foramen tumors are deeply located; some are highly vascularized and may involve or infiltrate cranial nerves, vessels, brainstem, and the bone structures at the cranial base, producing a large surgical defect; and some may present with major posterior

fossa extension. Despite these difficulties, radical removal of a benign jugular foramen tumor may be curative and remains the treatment of choice. The size of the lesion is directly related to the complication rate.Total resection of benign paragangliomas with large intracranial extension may be impossible or associated with unacceptable com- plications.Paragangliomas may invade cranial nerves that still function, and total resection is only possible with sacrifice of these nerves.

The multidisciplinary approach gives the best chance of radical removal with preservation of cranial nerves, vessels, and the brainstem. To avoid postoperative complications, an adequate surgical exposure and reconstruction of the cranial base are required. Surgical morbidity and mortality are usually associated with damage to the lower cranial nerves.Identification and dissection in the neck and at the brainstem is helpful in the preservation of these nerves. When they are infiltrated yet still functioning, our strategy is to leave a small piece of tumor around them and if necessary (if there is proven postoperative residual tumor growth) administer radiotherapy.

Tracheotomy should be performed as early as possible if clinical signs of persistent aspiration are present. Cases of CSF leakage are common and may be a serious complication after resection of jugular foramen lesions.The cranial base is usually reconstructed with abdominal fat graft, rotation of local and distant pedicled muscle flaps, and free muscle flaps vascularized with microsurgical vessel anastomosis.

Postoperative Treatment

Some authors recommend radiotherapy as the initial treatment for paragangliomas, because resection of these tumors is associated with damage to the lower cranial nerves. Series in which more than 80% tumor control is reported after radiotherapy are presented. The effect of radiation appears to be on the blood vessels and fibrous elements of the lesion, and the beneficial effects of radiotherapy for these tumors remain unclear. Radiation-induced neoplasm has been described after radiotherapy for benign glomus jugulare tumors. Radiosurgery with gamma knife or linear accelerator modalities may be more beneficial in reducing the side effects of irradiation. Chemotherapy is indicated for some malignant lesions.

V. Conclusions

Neurologic examination of low cranial nerve palsy is crucial since most patients had at least one low cranial nerve palsy. All tumors were detected in advanced stage due to slow growing nature and lack of symptom. Angiography with embolization is crucial for successful tumor removal without massive bleeding. Transmastoid retro auricular approach can be considered as a safe, satisfactory approach for removal of jugular foramen tumours. In tumors with intracranial extension, combined approach is recommended in that it provides better surgical view and can maintain the compliance of the patients.

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