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Case Report

Glomus Jugulare - A Rare Presentation

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Abstract

A case report on glomus jugulare from India.

We present a case of a 15-year-old male, with 6 months history of tinnitus and progressive hearing loss, presented with facial nerve palsy (House-Brackmann V). Computed tomography scans demonstrated an enhancing soft tissue lesion with erosion of jugular bulb, invasion of the petrous and tympanic bone, middle ear and vertical part of facial nerve with erosion of postero medial wall of left TM joint (type C2 of Fisch's classification for GJT). Intraoperatively the mastoid part of the facial nerve was identified involved by tumour and needed to be resected. Inter positioning cable grafting with greater auricular nerve, harvested at the beginning of the surgery is done. Total tumour resection is done with a remarkable postoperative course. The patient also presented with improvement in facial nerve function after 6 months. The patient consented with publication of his images.

Keywords: Glomus Jugulare; Paragangliomas; Facial Nerve

Introduction

Paragangliomas are tumours originating from the paraganglionic system (autonomic nervous system), mostly found at the region around the jugular bulb, for which reason they are also termed as glomus jugulare tumours (GJT). Glomus tumours are rare, non chromaffin paragangliomas. Incidence of about 1 per 1.3 million and a marked predisposition in women. The majority of these tumours are slow-growing with a clinical manifestation in the sixth to seventh decade of life. The typical expansive and locally destructive growth pattern of glomus jugulare tumours leads to cranial nerve damage with related symptoms (facial palsy, pulsatile tinnitus, hearing loss, difficulty in swallowing and loss of voice) and in larger tumours brainstem compression can be seen. These infiltrative tumours follow the path of least resistance, and in CT scans expansion and erosion of the temporal bone. Surgery is the main modality of treatment. Although these lesions appear to be histologically benign, clinically they present with great morbidity, especially due to invasion of nearby structures such as the lower cranial nerves. These are challenging tumours, as they need complex approaches and great knowledge of the skull base.

Case Report

15yrs old male came to ENT OPD with complaint of impaired hearing in left ear since 6 months, ringing sensation in left ear from 6 months, pain in left ear since 3 months, left sided facial weakness since 3 months. H/o bleeding from left ear. No h/o discharge from left ear.

Photo 1: Photo of Patient showing preoperative left facial palsy.

Examination

On examination of the Left ear there is a pinkish pulsatile mass seen behind bulging tympanic membrane. Grade 5 lower motor neuron facial nerve palsy on left side. Oto endoscopic examination shows a reddish pulsatile mass behind an intact bulging tympanic membrane seen in the floor of the middle ear. blanching is visualised on pneumo otoscopy. Rt. Ear Normal T.M.

Photo 2: Right ear – normal intact tympanic membrane.

Photo 3: Left ear there is a pinkish pulsatile mass seen behind bulging tympanic membrane.

Management

Pure tone audiometry showed - Rt: normal hearing level and Lt: moderately severe mixed hearing loss of 68db. Impedance audiometry showed Middle ear effusion in left ear, No middle ear pathology in right ear. High resolution computed tomography (HRCT) of temporal bones revealed enhancing soft tissue density lesion (1.1× 1.4×o.8cm) in the left hypo tympanum extending into the meso tympanum, lesion is seen bulging into EAC with erosion of jugular bulb e/o erosion of medial wall of mastoid and vertical segment of Lt.facial canal.

Photo 4: Photo of Pure Tone Audiometry.

Photo 5: Photo of Impedance Audiometry.

Photo 6: Photos of HRCT Temporal Bones.

Pre –op embolization was done to the feeding vessels arising from ECA with gelfoam particles.

Tumour excised by infra temporal fossa approach.

Photo 7: Photos of Embolization.

Photo 8: Intra operative Photos.

Photos of intra operative Large C shaped incision is made post auricularly, external auditory canal is transacted and over sewn.

Dissection is done initially in the neck, identifying the facial, glossopharyngeal, vagus, spinal accessory, hypoglossal nerves. Major vessels in the neck, carotid and its bifurcation, internal jugular vein are identified and ligated.

Radical Mastoidectomy, removal of mastoid tip are done. Facial nerve is completely skeletonised and vertical segment of facial nerve was infiltrated by the tumour, the proximal and distal segments of the involved facial nerve are decompressed.

Intraluminal packing of sigmoid sinus is done with gelfoam, dissection of the tumour from ICA, infra labyrinthine chamber and petrous apex was done. Perfect haemostasis is secured.

Skeletonisation of antero- inferior part of EAC is done for complete access to meso tympanum and complete dissection of the tympanic part of temporal bone and IAC to Eustachian tube is performed.

Tumour infiltrated part of facial nerve is resected and nerve repair is done by cable grafting with greater auricular nerve.

Photo 9: Intra Operative Photos and Cable Graft to the Damaged Facial Nerve.

Follow up

Blind sac procedure of left ear is seen on oto endoscopy on follow-up.

Patient is on Galvanic physiotherapy for Facial nerve palsy. Recovery was observed after 6 months post op. with improvement in facial nerve function

Discussion

Presentation of Glomus Jugulare in adolescent male is rare.

Involvement of facial nerve with glomus is also a rare presentation.

Kovacova., *et al*. [1] have described a case of 53-year old female with right sided hearing loss and right sided facial nerve palsy followed by hypoglossal nerve palsy, she was evaluated and diagnosed as Glomus Jugulare and was managed with Stereotactic radiosurgery. Arresting the tumour growth was the primary goal with no improvement of cranial nerve palsies.

Kotrashetti., *et al*. [2] described a case of glomus jugulare in a 4 year old female patient with complaints of left sided facial nerve palsy with drooping of left eye lid and difficulty in swallowing with left mastoid swelling and purulent foul smelling discharge. Child was managed with antibiotics and sent to higher centre for further management.

Conclusion

Although good results can be achieved with complete surgical removal, there is a significant risk of morbidity (0-39%) and mortality (0-2.7%) with this approach.

Radiotherapy is the treatment of choice for patients with intracranial extension, and patients with bilateral and multiple tumours, or patients who are inoperable. Glomus tumours respond to Radiotherapy (RT) slowly. Residual mass persisting after RT does not indicate treatment failure. Tumour may decrease in size, but rarely disappears.

Disease control is defined as the absence of progression of symptoms without any increase in size with physical examination or radiological control.

Glomus tumours are slow-growing lesions, therefore, it is necessary to be cautious about tumour control without increasing morbidity and mortality.

Follow-up was done every month for 6 months.

After 6 months there is improvement in facial nerve function and blind sac visualised in left EAC.

Photo 10: Photos of Patient with Post OP. Facial nerve palsy recovery after 6 months of surgery is seen.

Photo 11: Post OP. Oto endoscopy Photos.

Bibliography

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