## **Case Report**

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# Recurrent glomus jugulotympanicum: a case report

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### **ABSTRACT**

Glomus tumors, also known as paragangliomas and chemodectomas, of head and neck are slow growing benign tumors with relatively good prognosis. The term Glomus is misnomer as it arises from paraganglions. Due to their critical location at the skull base, these tumors may have significant morbidity and mortality if not identified and managed in time. The common extra adrenal sites in head and neck are carotid body, jugular foramen, middle ear and the vagus nerve. The term glomus jugulotympanicum is used when the tumor crosses the distinction of being confined either to middle ear or the jugular foramen. Management aims at early detection and complete surgical excision preserving surrounding vital neurovascular structures as far as possible. Larger tumors in advanced stage may not be amenable to complete surgical excision.

Keywords: Glomus tympanicum, Paraganglioma, Catecholamines

## INTRODUCTION

Anatomy of the jugular foramen is complex as it contains lower cranial nerves and major vessels. The tumours developing in the foramen or extending into it pose significant management challenges. Paragangliomas, also known as glomus tumours and chemodectomas, originate from the paraganglia in the carotid body at the carotid bifurcation, vagal nerve, jugular foramen and the middle ear. Marchand, in 1891, reported first paragangliomas of the carotid body.1 The classic, adrenal pheochromocytoma is catecholamine producing paragangliomas arising in the adrenal medulla. Although, majority of these non adrenal tumours are non secretory, catecholamine producing paragangliomas of the head and neck have also been described.<sup>2</sup> Paragangliomas are named as per the site of their origin. The paragangliomas are typically described as benign, slow growing masses with peak incidence in the fifth and sixth decade of life.

Depending on the origin and the spread, paragangliomas can have a varied presentation. Glomus tympanicum commonly present with pulsatile tinnitus, hearing loss and otalgia.<sup>3</sup> Bleeding, despite the tumour being highly vascular, is an uncommon presentation.

Biopsy is condemned. Surgical excision is the traditional treatment of choice, but radiotherapy is also often used in conjunction with surgery for larger tumours or in cases of unresectable slow growing tumors. Glomus tympanicum often recurs after resection, which makes a key consideration while making management decision. Treatment planning, therefore, depends on determination of tumour size, type, extent, intracranial extension, and relation to major neurovascular structures.

#### **CASE REPORT**

A 59 years female, known hypertensive on medications, presented with complaints of pulsatile tinnitus and progressive hearing loss right ear for one year duration. There was no history of otorrhoea, vertigo, bleeding from the ear, dysphagia, hoarseness of voice, difficulty in mastication or speech, headache, visual disturbances or symptoms of nasal regurgitation. She had presented with similar complaints in 2003 and was diagnosed as glomus

tympanicum right for which she underwent surgical excision. General examination was essentially normal. No pallor, edema or lymphedenopathy was detected. ENT examination revealed a well healed right postauricular scar mark of previous surgery. Otoscopy showed a well epithelized mastoid cavity with high facial ridge. A reddish-blue pulsatile mass 1x1.5cm was seen in deep external auditory canal completely obscuring view of the tympanic membrane. Brown's sign was positive. Tuning fork tests were suggestive of moderate grade conductive hearing loss on right side. Otoneurological examination was normal. Systemic examination was Routine hematological essentially normal. biochemical parameters were within normal limits. Serum catecholamines and urinary Vanillylmandelic acid (VMA) levels were not raised. Computed tomgraphy (CT) and Magnetic resonance imaging (MRI) of the temporal bones and neck revealed post operative status with soft tissue opacification of mastoid bowl, well defined lobulated soft tissue density lesion in middle ear cavity centered on cochlear promontory and extending into all middle ear recess as well as epitympanum, hypotympanum and the jugular fossa. Preoperative embolisation was not done as the tumor was small. Also, the surgical planes were disturbed because of the previous surgery.

The patient was taken up for surgical excision of the tumor under general anesthesia. A 'C' shaped curvilinear incision was given in the right post-auricular region with neck extension. Intra-operatively, distal control of the internal jugular vein (IJV) was obtained in the neck. Temporalis fascia graft harvested. Cavity of previous mastoidectomy was seen with high a facial ridge. The cavity was revised; proximal control of the sigmoid sinus was obtained. The facial ridge was lowered completely. The posterior tympanic cavity and posterior attic space was free of tumor. Partially eroded incus and the malleus were removed. Tensor tympani muscle tendon was divided. Drilling was continued anterior to the facial nerve to gain access to the tumor without displacing or transposing the nerve. Glomus tumor was seen involving anterior epitympanum, mesotympanum, oval window, round window, protmpanum area, and jugular bulb and hypotympanic air cells. The tumor was freed from all directions with careful drilling and electrocautery ensuring hemostasis. Jugulo-carotid crest was partially eroded by the tumor and was drilled to get inferior clearance of tumor. Anteriorly the petrous part of temporal bone was drilled up to the ICA to obtain the clearance. The tumor was seen attached to the ICA as well as the jugular bulb by fibrous bands which were carefully divided. The stapes was removed with its footplate keeping the membranous labyrinth intact as the tumor was seen involving the footplate area. After complete removal of the tumor, autologous stapes and temporalis fascia was used to cover the oval window area. Temporalis fascia was used to cover the tympanic cavity. Post operative period was uneventful. Follow up at 2 months revealed a well healed mastoid cavity.



Figure 1: Oto-endoscopic image of the right ear revealing postop status. A well healed mastoid cavity with a high facial ridge is visible. Also a reddish fleshy mass can be seen filling the deep EAC (arrow).



Figure 2: The reddish mass filling the deep EAC obscuring view of the tympanic membrane.

The mass was pulsatile.

## **DISCUSSION**

Paragangliomas are benign neoplasms arising from the glomus bodies, composed of cells of neural crest origin, widely dispersed throughout the body. Extra adrenal paragangliomas are found in four locations in head and neck: the carotid body (at common carotid artery bifurcation), jugular foramen (glomus jugulare or glomus vagale) and the middle ear (glomus tympanicum). The paragangliomas of the temporal bone are closely related to the Arnold's nerve and Jacobson's nerve. 50-55% of tumors are seen in the dome of jugular bulb or along the nerve paths. About 25% tumors are seen along the Jacobson's nerve over the promontory<sup>4,5</sup>. The term glomus is a misnomer; the more apt term paragangliomas was coined by Glenner and Grimley, which is currently used to describe this lesions.<sup>6</sup>

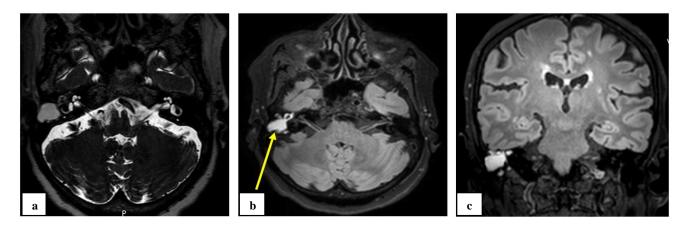


Figure 3: MRI of the temporal bone: postop status. Well defined soft tissue density lesion in middle ear filling all the recesses as well as hypotympanum and epitympanum. It showed a hyper-intense signal on T2W image and dense enhancement on post contrast images (arrow). The lesion was seen extending into the right EAC and the right jugular fossa.

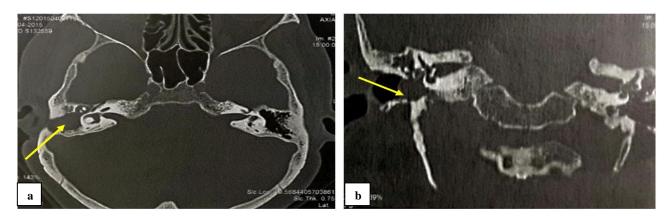


Figure 4: CT scans of the temporal bone: Soft tissue density lesion filling the middle ear and external auditory canal (arrow) eroding the floor into the jugular fossa (arrow).



Figure 5: The distal control of the IJV was obtained in the neck. Internal jugular vein (a) and Spinal accessory nerve (b) are seen here.

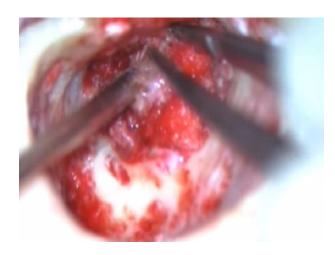


Figure 6: The facial ridge was lowered and the tumor was carefully dissected from its attachments.

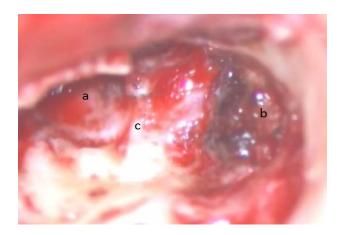


Figure 7: The jugular fossa was drilled. The attachments of the tumor with the jugular bulb (b) and the ICA (a) were carefully divided. Jugulo carotid crest (c) was partially eroded.

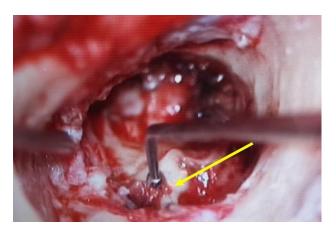


Figure 8: The tumor was seen involving the stapes footplate area and was going under the arch of the stapes suprastructure (arrow). The stapes was removed carefully keeping the membranous labyrinth intact to obtain tumor clearance.

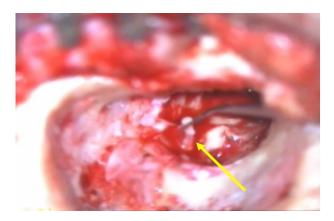


Figure 9: The autologous stapes (arrow) and Temporalis fascia graft was used to cover the exposed oval window. The cavity was lined with Temporalis fascia graft and the tympanomeatal skin flap.



Figure 10: The jugular fossa was drilled. The attachments of the tumor with the jugular bulb (b) and the ICA (a) were carefully divided. Jugulo carotid crest (c) was partially eroded.

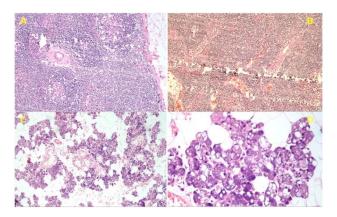


Figure 11: HPE: Nests of epitheloid cells separated by reticulin fibres of subendothelial layers of vascular spaces. Seen here in different magnification fields.

Paragangliomas have an annual incidence of 1 per 1.3 million people. These tumours are commonly seen in 5<sup>th</sup> and 6<sup>th</sup> decade of life, with a female preponderance. Glomus jugulare tumors arising in the jugular foramen spread typically along the paths of least resistance, including mastoid air cells, vessels, eustachian tube and neural foramen. Glomus tympanicum may be confined to the middle ear or may spread along the paths of least resistance to the mastoid or even up to the nasopharynx. Vagal paragangliomas (glomus vagale) can be 'dumbbell' shaped, with superior spread to posterior fossa and inferior spread into the infratemporal fossa. If the tumor is large and involves the jugular bulb, the term jugulotympanicum applies. In the present case, the tumor was involving the middle ear as well as the jugular fossa.

Microscopically, they consist of clusters of type I catecholamine containing chief cells (Zellballen) and type II sustentacular cells (modifies Schwann cells) with rich vascular network <sup>10</sup>. Catecholamines secreting functional tumors are found in 1-3% of cases. Multicentricity is seen in around 5-15% of non familial patients.

Paragangliomas are typically described as benign and slow growing. Depending upon the spread of the tumor, it may present with pulsatile tinnitus with hearing loss. Other symptoms may include bloody otorrhoea, otalgia and facial nerve palsy. Brown's sign is the pulsations elicited by pneumatic compression of the tumour, while Aquino's sign describes the blanching of the mass with gentle reassure over the carotid artery. A large neglected tumor may spread to the jugular fossa, neck and even intracranial cavity giving rise to lower cranial nerve palsies and features of raised intracranial tension and occupying lesion. Hypertension, tremors. tachycardia and headaches indicate towards the possibility of a functional tumour.

Imaging is the primary modality of investigation for glomus tumours of head and neck. A combination of contrast enhanced computed tomography (CT), magnetic resonance imaging (MRI) and angiography is ideal for the diagnosis and localization of glomus tumours. CT scan demonstrates strongly enhancing mass at its site of origin which is typical in the diagnosis of glomus tumors<sup>11</sup>. It is also best for evaluating bony erosion and destruction which is commonly seen in glomus jugulotympanicum. Similar to CT, contrast enhanced MRI demonstrates strongly enhancing soft tissue mass, which is typical in diagnosis, at characteristic locations. 12,13 A typical salt and pepper pattern of the tumour mass is seen on MRI. Angiography identifies the primary feeding vessels to the lesion, helps in detecting multicentric tumours and allows for possible preoperative embolisation. Preoperative embolisation of head and neck paragangliomas is very safe adjuvant to surgical excision.14

Surgical resection is the treatment of choice traditionally. The surgical approach depends on the size and extent of the tumour. In the present case, the anatomical course of facial nerve was not disturbed and the tumour was approached by drilling anterior to the nerve. However, radiotherapy is often used for larger slow growing tumours with an aim to restrict growth in unresectable tumours. Preoperative evaluation with CT, MRI and angiography has been proposed as essential for optimal planning. A careful evaluation of hypotympanum and jugular bulb is important; as its involvement alters the surgical approach. Facial nerve transposition may not be required in cases of smaller tumours, as was seen in our case.

A conservative approach applying both the modalities has also been advocated in elderly and medically infirm patients. <sup>16</sup> Radiation therapy, particularly gamma knife radio surgery, has shown good growth control with low risk of therapy related cranial nerve weaknes. <sup>17,18</sup>

#### **CONCLUSION**

Recurrent or residual paragangliomas present rare but challenging situations to the surgeon. The choice of treatment is made on case to case basis, depending upon patient's age, health, location and size of tumour and status of lower cranial nerves at presentations. The goal of treatment is to ensure complete tumour control with a low morbidity.

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