

# **Invasive Glomus Jugulare Tumor (Type D2) in a Male - A Rare Case Report**

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**Case Report**

Volume 1 Issue 2

**Received Date:** July 25, 2017

**Published Date:** August 28, 2017

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## **Abstract**

**Study Design:** A rare case report in a male and review of literature.

**Objectives:** To describe a case report of invasive glomus jugulare tumor with intracranial extension.

**Methods:** A 35 year old male complaint of pulsating tinnitus on his left ear, vertigo and progressive ipsilateral hearing loss for 4 years. Audiological examination and radiological assessment including Computerized Tomography and Magnetic Resonance Image were carried out. We diagnosed it as invasive glomus jugulare tumor with intracranial extension (type D2).

**Conclusion:** Glomus tumors are uncommon slow growing and hypervascularized benign tumor that arise within the jugular foramen of temporal bone but often locally aggressive and produce permeative pattern of bone destruction. It is commoner in females (F: M ratio 3-6:1) and estimated annual incidence of 1 case per 1.3 million people. In most cases diagnosis confirmed by imaging studies.

**Keywords:** Magnetic Resonance Imaging (MRI); Computed Tomography (CT); Glomus

## **Introduction**

Glomus jugulare tumors are rare, slow growing, hyper vascular tumors that arise within the jugular foramen of temporal bone. These tumors grow from paraganglion or glomus bodies. Gaffey called them chemodectomas, he believed that jugulotympanic paraganglions had chemoreceptor properties and associated them with aortic and carotid bodies, which could modulate cardiopulmonary reflexes from changes in pO<sub>2</sub> and blood pressure. Glomus bodies originate from nonchromaffin cells; these cells originating from primitive neural crest compose the extra-adrenal neuroendocrine system [1].

They may be found in the carotid body, adrenal medulla, and roof of the jugular bulb, along Jacobson and Arnold's nerves, from the jugular fossa to the promontory of the middle ear. Therefore, glomus tumors must be named according to their origin: glomus tympanicum, glomus jugularis, glomus vagale and glomus carotid [2]. According to Gaffey, paraganglions have chemoreceptor properties, and are named chemodectoma. This may cause increased catecholamine synthesis [3].

In 1945, Rosenwaser described the first patient diagnosed with glomus jugulare tumor [4]. The patient

survived until 1987. Vascular tumors of the middle ear had previously been reported, but Rosenwasser was the first to recognize the origin of these tumors from the glomus jugulare. He provided the first description of the surgical removal of a glomus jugulare tumor.

### Case Report

A male patient of 35 years old complaining of pulsating tinnitus on his left ear, vertigo and progressive ipsilateral hearing loss for 4 years. He also complains headache and dysphasia for 3 months. Otoscopy showed polyp in the external auditory canal and bruit is seen on mastoid antrum on clinical examination. Plain radiography of skull showed an osteolytic area in petro-mastoid region (Figure 1). CT scan showed bone destruction in the region of jugular fossa, mastoid and postero-inferior petrous bone

and adjacent occipital bone (Figure 2a & b). Doppler ultrasound of mastoid region showed a mixed echogenic mass at mastoid region having high vascularity (Figure 3). Gd-MR scan of brain showed soft tissue mixed signal intensity on T1WI and T2WI with intermixed high intensity signal and signal voids (i.e salt and pepper appearance) at left petro-occipital region (Figure 4a-c) extending into middle ear and posterior fossa region abutting left cerebellar hemisphere causing mild effacement of 4th ventricle. MR scan also revealed this tumor also extending into inner ear (Figure 4d) but sparing internal auditory canal.

He underwent surgery followed by radiotherapy. Histopathologic examination confirmed the diagnosis of glomus tumor.



Figure 1: Plain radiography of skull showing irregular lytic area at left petro-mastoid region.

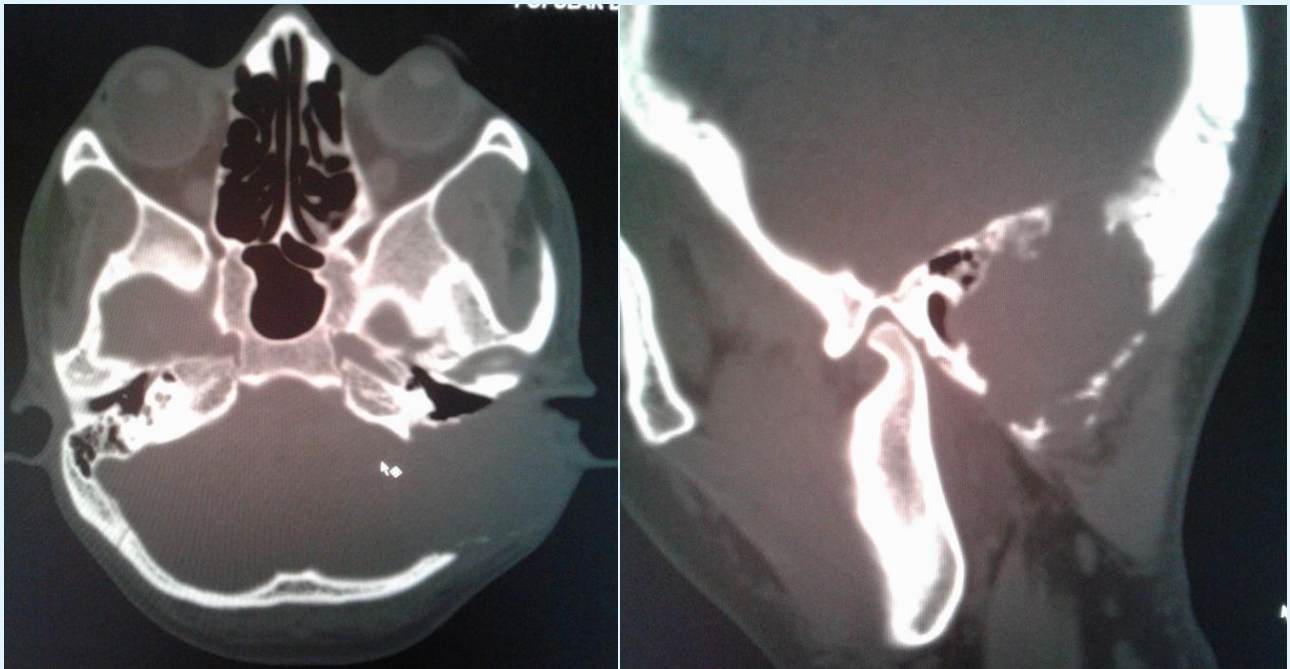


Figure 2 (a, b): CT imaging demonstrates the extension of bony destruction by the tumor involving petro-mastoid and adjacent occipital bone.

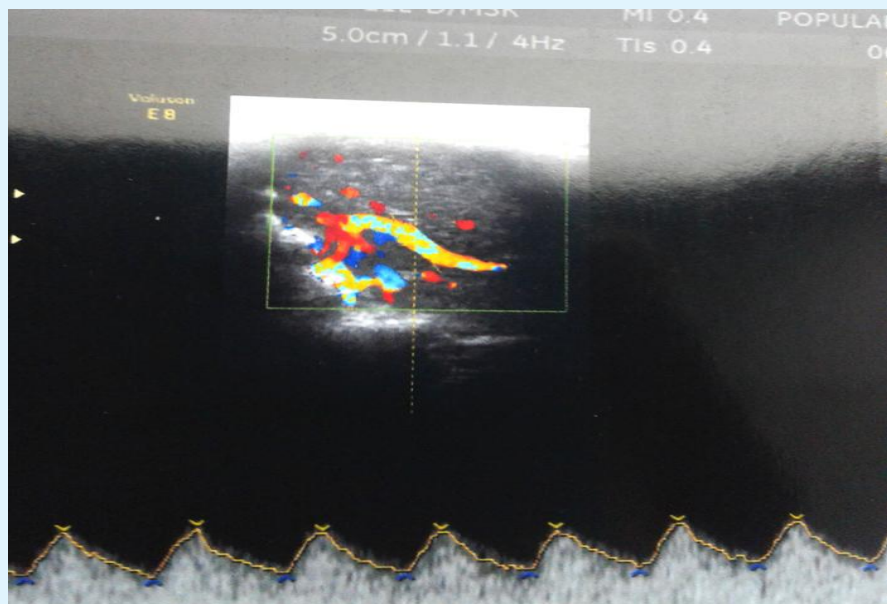
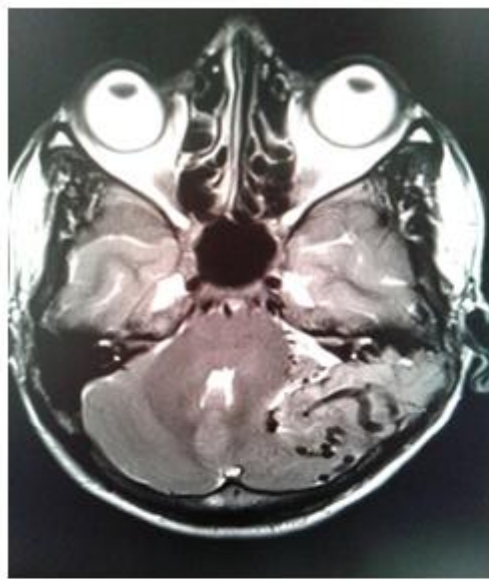
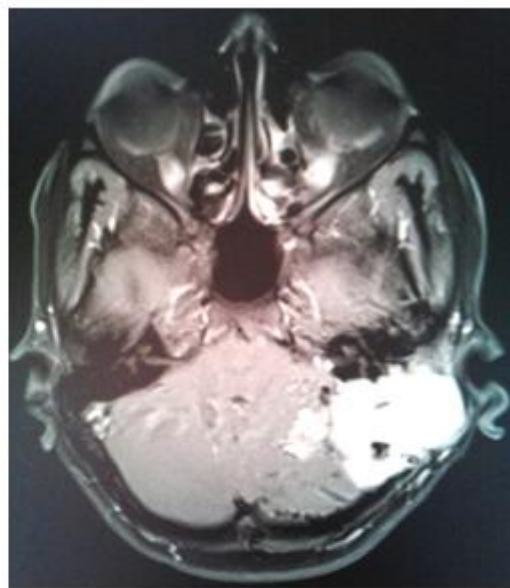


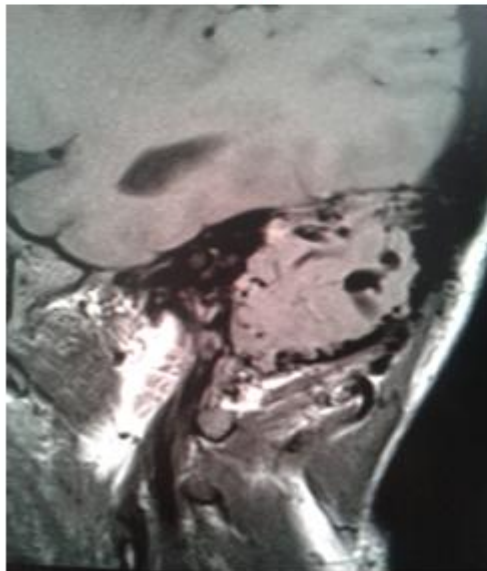
Figure 3: Doppler ultrasound of mastoid shows the vascularity of the tumor.



4a



4b



4c



4d

Figure 4a-d: Sagittal and axial Gd-MR showing a large glomus jugulare tumor and its upper and lower limits in the posterior cranial fossa and neck sparing internal acoustic meatus (4a, 4b, 4c). This tumor also extending into inner ear (4d) is abutting the semicircular canals.

## Discussion

Glomus jugulare tumors occur predominantly in women (female to male ratio is 3-6:1) in the fifth and

sixth decades of life. The right ear was most commonly affected in previous literature [5]. This may be linked to the anatomy of the jugular gulf, more elevated and dilated on the right ear. In our case it occurred in male patient of



35 years old involving left ear, that's why it is a rare presentation.

The most frequent symptoms are hearing loss and pulsating tinnitus, usually unilateral; adjacent structures are usually eroded due to massive vascularization. Other aural signs and symptoms are ear fullness, otorrhea, hemorrhage, bruit and presence of middle ear mass. Significant ear pain is uncommon. Involvement of the inner ear produces vertigo and sensorineural hearing loss. Cranial nerve involvement produces hoarseness and dysphagia. The presence of jugular foramen syndrome (paresis of cranial nerve IX-XI) is pathognomonic for this tumor. Our patient presenting with pulsating tinnitus, sensorineural hearing loss and dysphagia and clinical examination reveals bruit over mastoid region.

In case of small lesion otoscopic examination reveals a characteristic, pulsatile, reddish-blue tumor behind the tympanic membrane that is often the beginning of more extensive findings (i.e, the tip of the iceberg). In case of larger masses, the tumor represents as polyps in the external ear and they may present massive bleeding when handled this findings is similar of our patient [6,7].

Diagnosis is confirmed through imaging studies because biopsy is usually contraindicated. Axial and coronal computed tomography (CT) scanning with thin sections are superior at demonstration the extent of bone destruction and Magnetic resonance imaging (MRI) with gadolinium-diethylenetriaminepentaacetic acid (Gd-DTPA) contrast is best for delineating intracranial invasion, as well as neck and large vessels involvement. Initially, the skull base erodes in the region of the jugular fossa and postero-inferior petrous bone, with subsequent extension to the mastoid and adjacent occipital bone. Significant intracranial and extracranial extension may occur as well as extension within the sigmoid and inferior petrosalsinuses; almost all features seen in our case having posterior fossa extension of about 4 cm (Figure 4a).

CT scan of this case shows bone destruction in the region of jugular fossa, mastoid and postero-inferior petrous bone and adjacent into occipital bone. Gd- MR scan of brain showing characteristics soft tissue mixed intensity with intermixed high intensity signals and signal voids on T1WI and T2WI at left jugular fossa with extension into middle ear, inner ear and posterior fossa in lobular fashion. For tumors with large intracranial

extension, vertebral angiography is advised to exclude arterial feeders from posterior circulation.

The Glasscock-Jackson and Fisch classifications of glomus tumors are widely used. The Fisch classification of glomus tumors is based on extension of the tumor to surrounding anatomic structures and is closely related to mortality and morbidity. According to Fisch classification our case is type D2 glomus jugulare tumor.

Type A tumor - Tumor limited to the middle ear cleft (glomus tympanicum)

Type B tumor - Tumor limited to the tympanomastoid area with no infralabyrinthine compartment involvement

Type C tumor - Tumor involving the infralabyrinthine compartment of the temporal bone and extending into the petrous apex

Type C1 tumor - Tumor with limited involvement of the vertical portion of the carotid canal

Type C2 tumor - Tumor invading the vertical portion of the carotid canal

Type C3 tumor - Tumor invasion of the horizontal portion of the carotid canal

Type D1 tumor - Tumor with an intracranial extension less than 2 cm in diameter

Type D2 tumor - Tumor with an intracranial extension greater than 2 cm in diameter

Incision biopsies and paracentesis are contraindicated because there is high risk for complications resulting from bleeding. Surgical resection and histopathologic examinations are necessary to differentiate it from other tumors. When there is no contraindication, all patients must undergo surgery [3,6,8].

## Conclusion

Glomus tumors are encapsulated, slowly growing, highly vascular and locally aggressive tumors. These tumors tend to expand within the temporal bone via the pathways of least resistance such as air cells, vascular lumens, skull base foramina and eustachian tube. Although most paragangliomas are sporadic, they can be familial with autosomal dominant inheritance and incomplete penetrance. The nonchromaffin paragangliomas have a familial tendency. So diagnosis is necessary for the patient and combination of CT as well as Gd-MRI is the imaging regimen for the diagnosis of glomus jugulare tumor.

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