GLOMUS JUGULARE TUMOR: A CASE REPORT AND LITERATURE REVIEW

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ABSTRACT
Glomus Jugular tumors develop within the jugular foramen of the temporal lobe. Medical and surgical management of these cases very rare, slow developing and more vascular tumors are always difficult to handle. Patients that have such type of tumors will have their inferior cranial nerves affected. With the contribution of inferior cranial nerves, the patients may have different functions of swallowing, chewing, hearing, speaking, balance and coordination. The prevalence rates of these tumors are very uncommon as 1 case per 1.3 million. Although they are consider benign. Clinical diagnosis are based on imagining study such as Computed tomography, magnetic resonance imagining digital subtraction angiography are mostly used. The management is notorious and can include surgical excision, stereotactic radiosurgery and embolization. A patient of 38 year old was evaluated for the glomus jugulare and surgical management. Patient was discharge after 8th postoperative day with decreased presenting sign and symptom on 6 month follow up.

Key words: glomus tumor, paragangolima, carotid, tympanic juglare

INTRODUCTION
Glomus tumors (GT) also termed as paragangliomas and chemodectomas are uncommon tumors. The incidence rate is 1 case per 1.3 million people 0.6% of head and neck tumors 0.03% of all tumors. In Pakistan the occurrence rate is 1 case in 1 million. Typically, these growths are diagnosed in the middle of the fourth and six decades of life. In females are 3-6 more frequent than in males in 50-70 decades in particular, especially on the left side .25-50% is in family cases, the gene responsible for these cases is located on 11q23 band[2]. Jugulare foramen is complex as it contains lower cranial nerves and major vessels. This tumor gets its name from the nerves called glomus bodies, and their location in the small opening in the temporal bone known as the jugular foramin. Glomus malignant tumors mostly exist in two position cervical (carotid, vagal) and cranium base (jugular and tympanic). These tumors seen have benign features and a very sluggish progress rate of 1 mm per year[7].

The classic adrenal origin pheochromocytoma is catecholamine producing paragangliomas arising in the adrenal medulla. Although, majority of these non-adrenal tumors are non-secretory, catecholamine producing paragangliomas of the head and neck[3].

The typical growth pattern of glomus jugulare tumors leads to cranial nerve damage with related symptoms facial palsy, pulsatile tinnitus, hearing loss, ear fullness, hemorrhage, bruit, vertigo, difficulty in swallowing and loss of voice and in larger tumors brainstem compression can be seen. While clinical signs are essential as an initial clue to a mass located in the temporojugular area. Clinical diagnosis based on imagining study such as Computed tomography, magnetic resonance imagining digital subtraction angiography is mostly used[7].

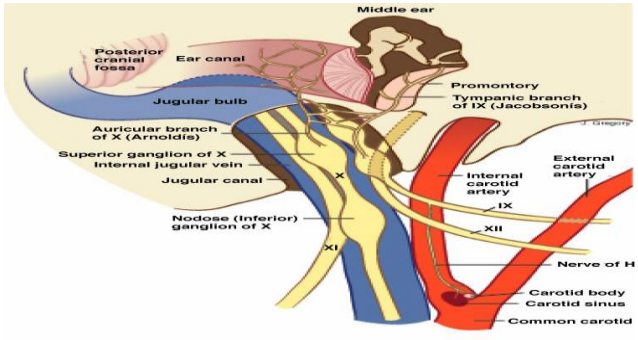
Treatment Complete surgical removal is good option but not always possible because to closeness of essential neurovascular bundle and difficult approach. Some studies shows these tumors can be removed by surgery called gamma knife surgery show that there is no significant difference in tumor volume and size. Radioactive branded meta- iodobenzyl guanidine is used by the neurosecretory cells because of physical resemblance to noradrenaline. For that reason 123 IMIBG scan can help in diagnosis secretory paragangliomas. The area selected by the scan can be targeted with 131 I-MIBG loaded with substantial radioactivity in malignant cases. Sometime Direct biopsy is required to exclude a squamous cell carcinoma[1].

CASE PRESENTATION
A 38 year old female admitted in public hospital with of Left ear bleeding, hearing loss and facial of the same side. Three year back she suddenly experience an

episode of fresh bleeding from Left ear it happen Again 6 month later. She experienced loss of hearing of the same ear which was gradually in onset and progressive one year ago. Physical examination revealed normal vital sign. Glass glow coma scale 15/15. Pupils equal in size and reactive to light. Cranial nerves V, VII, VIII affected. An enlarge tumor palpable back of the ear. There was no history of any previous surgery or any medical disease. Family history of same disease not significant. She was married from 9 year but no child. Pathology shows H.b= 10.8 g/dl decrease and all other labs were in normal range. Computerized tomo angiography shows tumor blush is noted the region of left petrous temporal and medial to mandibular region with neovascularity. Magnetic resonance imagining shows that mass 3.3x2.5x2.5 cm with the involment of internal auditory canal signals isointense to hyper intense on T2W1 and T2 FLAIR with homogenious post contrast enhancement. Mass shows numerous nternal flow of voids. Inferiorly the mass reaching up to the intratemporal fossa with the involment of temporal lobs. All these findings suggested most likely glomus juglre. Treatment patient was treated by surgery (Craniotomy and Excision). Histopathology report shows begnin tumor. Post operative medication was antibiotic injection ceftriaxone 1g I/V BID, injection Augmentin 1.2g I/V TDS, 2 tab panadol SOS, injection dexa 4mg I/V TDS. Symptom was decrease patient discharge after 01 week with improving condition.

DISCUSSION

Glomus tumors also called as paragangliomas or chemodectomas are benign neuroendocrine growths resulting from the glomus cells of the vegetative



nervous system. This type of growth is more common in females, with prevalence of up to six times more than male. Most common symptoms of glomus jugular are associated to anatomical presentation and related structures affected by the tumor. 80% patient present with pulsatile tinnitus this is common symptom of this tumor. Classically, this type of lesion has pulsatie tinnitus and hearing loss this is complete by audiometry and visualized through vascular membrane as vascular tympanic mass on the promontory. Glomus jugulare tumors are pinpointed on the jugular bulb and may cause palsies of the ninth to eleven cranial nerves.

(Representation of head and neck paragangliomas and their relationship with the lower cranial nerves and major vessels[5].

Glomus jugular tumors develop from the jugular foramen alongside the way of less resistance and mostly expand into the pneumatized portions of the temporal bone, related vascular and neural foramina and eustachian tube. Characteristically, in advance stage jugular paragangliomas increase causing in a "moth-eaten" appearance of the temporal bone due to related bony erosion. This discriminates from tympanic paragangliomas to jugular paragangliomas, which are normally less destructive, sparing the jugular bulb andossicless[10].

Histologically these structures are composed of chemoreceptors type I and type II cells support a thick matrix of connective tissue between nerve fascicles and highly vascularized showing a mass arterial drains in a vast peripheral venous system. Chemoreceptors Type I and II secrete dopamine, noradrenaline, tumoral secretion and the most important blood supply is coming from ascending pharyngeal artery. Large tumors can receive extended branches of the internal carotid or vertebral artery Tumors left from these tissues are called equal chemodectomas, paragangliomas noncromaphin[9].

Radiological imaging techniques, especially magnetic resonance imaging (MRI), are the major methods for the diagnosis of glomus juglare tumor. There are very few reports of ultrasound in the diagnosis of glomus juglare being located in the skull base[3].

We present the case of a rare giant GJT, which extended within the internal auditory canal. And was detected by CT scan first. We highlight the initial CT scan findings with the involment of CT angiography and MRI findings. Advances in technology resulted in better surgical outcomes. The main reasons for this were introduction of the operating microscope, bipolar cautery, microdissection techniques, various skull base approaches better imaging (computed tomography CT and magnetic resonance image)[8].

Stereotactic radiosurgery may be applied as the treatment of choice. In paragangliomas this method results in lower morbidity than surgery with a possibility of clinical improvement. It provides a good long-term tumor control, with 61% of the tumors stable in size. Recent studies have shown a long-term neurological improvement in 42% of patients and

aggravation of clinical symptoms can treated with stereotactic radiosurgery.As far as endovascular therapy is concerned, it is confined to preparation for surgery and palliative applications but it is not curative. As regards preoperative embolization through feeding vessels its role is to decrease blood loss during surgical procedures and to help perform resections in a more secure manner. In case of head and neck paragangliomas both Tran's arterial embolization with polyvinyl alcohol (PVA) particles and direct percutaneous embolization with n-butyl cyanoacrylate (NBCA) or ethylene vinyl alcohol polymer are used. Either way, the procedure of presurgical adjuvant endovascular embolization is safe and effective, or the application of this form of therapy followed by surgical excision of paragangliomas is widely accepted[7].

Excision of the portion of the glomus jugulare helps decompress the brain stem and cranial nerves and thus eliminates any threat to life from the lesion. The extracranial portion is removed using the infratemporal fossa approach up to the distal limit of the lesion. This "middle path" helps reducing intraoperative bleeding and operative time. Moreover, postoperative morbidity is also reduced. Sometimes, the petrous portion is small and can be excised without extensive bone work[8].

However, neurological consequences of surgical resections are significant and mainly include persistent cranial nerve deficits. Surgery may be contraindicated due to age and general physical condition of patient. This relates especially to glomus jugular para-gangliomas even in experienced centers, the results are not satisfactory and the overall mortality rate ranges from 1.2% to 6.4%[9].

In general, surgery is the best option for treatment of carotid body growth; however, radiation therapy is the number one choice for jugular and vagal tumor. Because of the complication of paragangoliomas it become difficult how to treat these type of case, a multidisciplinary algorithmic approach should be used for management of paragangliomas. The approach should highlight single-modality management which produces outstanding rates of tumor preventation, short rates of morbidity and the protection of long-term purpose in these patient residents[6].

CONCLUSION

Glomus jugular tumor is very uncommon tumor found at jugular foramin. Growth configuration of these tumor affect cranial nerve damage and common sign symptom are hearing loss, pulsatile tinnitus and facial palasy. This tumor diagnosed on the base of radiology test like Computerized tomography, magnetic resonance imagining. Treatment depends on the size of the tumor. Small sized tumor can treat with radio therapy, surgical treatment also used if there is no other medical disorder

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