Abstract: Glomus tumors are the most common neurotologic tumor next to acoustic neuroma. When it is confined to the middle ear it is termed as Glomus tympanicum, which is a very rare tumor of the head and neck but most common tumor of the middle ear. We are presenting here a rare presentation of recurrent glomus tympanicum in a middle aged woman presented with facial nerve palsy, which was removed surgically.

Keyword: Glomus tympanicum, Facial palsy, Recurrent glomus, Middle ear

INTRODUCTION
Glomus tumors are the tumors arising from the special neural crest elements, the paraganglion cells which along with autonomic ganglion cells forms the paraganglia. Glomus tumors can be classified according to their location- glomus jugulare, glomus vagale, glomus tympanicum and carotid body tumors. Glomus jugulare is the most common tumor of the head and neck while glomus tympanicum is very rare tumor of the head and neck. They are slowly progressive, highly vascular and low grade malignant tumors causing problem due to neurological deficit and its complex location at the skull base.

CASE REPORT
A 40 years old female presented to our outpatient department with complaints of dizziness for the past 6 months, which was persistent, headache for the past 6 months, which was localized to the right frontal and temporal region. Profuse watery ear discharge in the right side for the past 4 months, Pricking right ear pain radiating to the right side of the face for the past 4 months. Facial deviation, inability to close the right eye and dribbling of saliva for the past 2 months. Patient was apparently normal till 1998, when she had hard of hearing, tinnitus, discharge and pain in the right ear, giddiness and headache for which she was investigated in Government General Hospital, Chennai in 2006. Diagnosed as glomus tympanicum. Excision of Right side middle ear mass with Type I tympanoplasty was done. Post operatively pt was relieved from all symptoms except for hard of hearing and tinnitus in the right side. On clinical examination, there was smooth pulsatile grey white mass occupying entreateral auditory canal and tympanic membrane was not visualized in the right ear. Rinnie test was negative and weber lateralized to right ear. Absolute bone conduction was reduced in the right side. Left ear was found to be normal. Vestibular functions were normal. Clinical examination of the cranial nerves was normal except for the lower motor neuron type of facial nerve palsy. Examination of other systems was found to be normal. Investigation of the patient in pure tone audiometry showed profound mixed hearing loss in the right ear while normal hearing in the left. Nerve conduction study for facial nerve was normal. HRCT temporal bone showed evidence of complete soft tissue opacification of right middle ear, mastoid and external auditory canal.

MRI temporal bone showed avidly enhancing mass lesion with no obvious bone destruction and epicentre located in the middle ear possibly glomustympanicum right ear, right mastoiditis. MRA was done to identify the blood supply to the tumor and the feeding vessel.

Surgical procedure: After clamping the external carotid artery to control bleeding, tumour was found to occupy mesotympanum, hypotympanum, attic, antrum and granulation tissue was seen over the horizontal portion of fallopian canal. Modified radical mastoidectomy with excision of glomustumour with facial nerve decompression was done, using 0 rod Hopkin endoscope.
On 3rd postoperative day patient was able to close her right eye completely. Electrical stimulation response to galvanic current - neuropraxia on right side. Nerve conduction study was normal. Histopathological examination revealed features of glomustympanicum - Clusters of polygonal cells with focal organoid pattern, cells exhibiting vesicular nuclei interspersed with numerous congested vessels, fibrovascular septa.

D I S C U S S I O N :
Glomustympanicum are slowly progressive nonchromaffin paraganglia associated with the middle ear, no carotid artery, facial nerve, stapes footplate, or round window. S T resection for advanced disease that was adherent to the petrous bone and facial nerve. Evidence of endocrine activity should be looked for by urinary assay of metabolites dopamine and VMA. Histologically the tumor resembles glomus bodies with epithelioid cells interspersed in a highly vascular stroma of capillary and precapillary vessels.

Classification is done by Fisch classification.

- Type A – Tumor localized to the middle ear.
- Type B – Tympanomastoid tumors with no destruction of bone in the infralabyrinthine compartment of temporal bone.
- Type C – Tumors invading the bone of the infralabyrinthine compartment of temporal bone.
- Type D – Tumor with intracranial extension.

C L I N I C A L  P R E S E N T A T I O N :
The most common presenting symptom is pulsatile tinnitus (80%) followed by hearing loss. Tympanic membrane erosion and bleeding are late signs. Facial nerve weakness signals advanced disease, and an indication of poor facial nerve prognosis. Observation of the drum under the microscope show a pulsation of mass - soft and often blanches on palpation. Hearing loss is conductive. Neurological assessment of the cranial nerves reveals the extent of tumor. Differential diagnosis includes high jugular bulb and an aberrant internal carotid artery. A more extensive spread involving the external auditory meatus may appear to be a squamous cell carcinoma which can bleed profusely.

I N V E S T I G A T I O N S:
Mycroscopy and biopsy are to be avoided. CT scan is mandatory to know the bone erosion and extent of tumor to bony structure of the ear which is vital in operative planning. MRI with gadolinium enhancement is done to identify the soft tissue involvement. Angiography or MRA indicates blood supply of tumor.

The treatment plan is based on tumor size, tumor location, patient’s age and health.

1. surgical excision
2. primary radiotherapy
3. surgical excision with planned adjunctive radiotherapy
4. no active treatment and continuous observation

O b j e c t i v e of the surgery is total resection of tumor where possible and without increasing the patient's neurological deficit.

R E V I E W  O F  L I T E R A T U R E:
In a study conducted by Carlson M Let al, the clinical presentation, surgical management, and outcomes of a large consecutive cohort of patients with glomustympanicum (GT) tumors managed at a single tertiary referral group over 4 decade was studied. 90.4% were women with mean age of 55.2 years. (93.9%) patients underwent gross total removal, while 6.1% received less than complete resection for advanced disease that was adherent to the petrous carotid artery, facial nerve, stapes footplate, or round window. S T Subha et al, presented two cases of glomustympanicum with no recurrence. One case presented with giddiness while the other presented with pulsatile tinnitus and progressive hearing loss. Gauravkumar et al, studied in a case, an unusual presentation of glomustympanicum with new bone formation with resultant ossicular fixation and conductive hearing loss. S Hirunpat et al, presented a very rare case of nasopharyngeal extension of recurrent glomustympanicum in a 61 year old woman with recurrent episodes of epistaxis. P Subashini et al, presented a case of glomustympanicum with unilateral conductive hearing loss as the only symptom. While the usual presentation of glomus tumor in the middle ear is pulsatile tinnitus. Bierry et al, studied the features mimicking glomustympanicum among Middle ear adenomatous tumor (MEAT). MEAT and GT appeared as tissular lesion with significant enhancement on CT and MRI. A vascular blush was present on angiography in all cases of GT and absent from all cases of MEAT. A close relationship between the tumor and the Jacobson's nerve or its branches was identified in all cases of GT. Pulsatile tinnitus was present in all patients with GT and absent in all patients with MEAT.

R E F E R E N C E S: