Glomus Jugulare

SYNONYMS
- Paragangliomas, Chemodectoma, Ganglia tympanica, vascular tumors of middle ear.

DEFINITION
- Glomus tumors are benign slow growing vascular tumors arising from GLOMUS BODIES present on dome of Jugular bulb, hypotympanum, promontory and along the Tympanic plexus.
- In other words collection of ganglionic tissue within the temporal bone in the adventitial layer of the jugular bulb.
- They are Non Chromaffin Paragangliomas.
- Paragangliomas are most common true neoplasms of Middle ear.
- Supplied by the 9th and 10th nerves (tympanic branch of glossopharyngeal or auricular branch of Vagus (i.e. Jacobsons and Arnold nerves). They are supplied by branches from the Ascending Pharyngeal Artery.
- Paraganglia are cells derived from neural crest and are found widely distributed in ANS (carotid, ciliary, vagal bodies, along aorta, glomus jugulare complex, adrenal medulla and paraadrenal glands).
- (Adrenal medulla = Chromaffin Paraganglia as it secretes Adrenaline and Nor Adrenaline and stain positive for chromaffin)
- Glomus jugulare is physiologically inactive chemoreceptor.

HISTOLOGICALLY
- Macroscopically – deep red, firm, rubbery masses that bleed profusely during manipulation.
- Microscopically – Clusters of epithelioid cells called Chief Cells (Zellballen) enclosed by fibrous septa and supporting cells interspaced in a highly vascular stroma of capillary and precapillary vessels. The proportion of the cellular and stromal components vary. Scarce unmyelinated nerve fibers also found.
- GUILD classified glomus tumors into two types depending on the amount of cellular and stromal components:
  1. Cellular glomus bodies – When the cellular component is predominant
  2. Vascular glomus bodies – When the vascular stromal component predominates.
- Most glomus bodies are ovoid in shape.
- Paragangliomas of the temporal bone divided into (as per Lundgrens Classification):
  - Glomus Tympanicum – originating within middle ear.
  - Glomus Jugulare – originating within the jugular fossa.

INCIDENCE
- 1 in 1,00,000  F:M = 6:1
- AD inheritance. The gene responsible for hereditary paragangliomas has been localized to band 11q23.
Most commonly seen in 5th decade of life but can occur from childhood to adulthood.

**ENDOCRINE ACTIVITY**

- Even though these tumors are considered non chromaffin paragangliomas with no endocrine activity, some cases with endocrine activity by these tumors have been reported. It is hence important to look for evidence of endocrine activity by urine estimation of VMA (Vanillylmandelic acid).
- Glomus tumors sometimes may show multicentric presentation i.e. present in both ears, or in conjunction with other paragangliomas.

**PATHOPHYSIOLOGY**

- Glomus tumors are **encapsulated, highly vascular, and locally invasive 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 the eustachian tube. They also invade and erode bone in a lobular fashion, can affect Facial nerve but they often spare the ossicular chain.
- Great propensity to infiltrate mastoid air cell system.

**SPREAD OF TUMOUR**

- Anteriorly ➔ Eustachian tube to nasopharynx
- Inferiorly ➔ Via lumen of IJV and Sigmoid Sinus to as far as atria. Carotid sheath lends access to neck. Neural infiltration is possible.
- Medially ➔ Labyrinth invasion along internal acoustic meatus
- Superiorly ➔ Via Skull base foramina.
- Laterally ➔ Perforation of TM may lead extension of tumour mass in EAC
- Posteriorly ➔ Mastoid air cell invasion occurs later in course

Ossicles remain intact or displaced. Extension along facial nerve can occur along fallopian canal

**PRESENTATION**

- These tumors are slow growing, with very little symptoms.
- The diagnosis may easily be missed. In fact the average delay between the onset of symptoms and diagnosis varied from **6 years to 15 years**.

**Symptoms of Glomus Jugulare:**

1. **Deafness** – 69% (CHL mostly, SNHL only with large tumors)
2. **Pulsatile tinnitus** – 55% (reduced on compression over jugular vein in neck.)
3. **Aural pain**
4. **Aural fullness**
5. **Imbalance** – 8%
6. **Otorrhoea** – 5% (usually blood stained)
7. **Facial palsy** – 8%
8. **Endocrine syndrome** – 3%
9. **Cranial nerve deficits**
   - Hoarseness – 16%
Dysphagia – 16%
10. **Headache** – 15 %
11. **Visual disturbance** – 6 %
12. **Mass in External auditory canal** (in case of rupture of TM and extension as polyp)
14. **Jugular Foramen Syndrome** – Eg: Vernet syndrome – Simultaneous paresis of 9\(^{th}\), 10\(^{th}\), 11\(^{th}\) with or without 12\(^{th}\) nerve. (Causes Glomus tumors (most frequently), Meningiomas, Schwannomas (Acoustic neuroma), Metastatic tumors located at the cerebellopontine angle, Trauma, Infections, Cholesteatoma (very rare), Obstruction of the jugular foramen due to bone diseases).
15. Hypoglossal nerve palsy in case of occipital extension
16. **Horners Syndrome** – due to involvement of Superior Cervical Ganglion (Sympathetic)

Presence of headache indicates intracranial extension

Dural sinuses may be involved may mimic sinus thrombosis

**On Otoscopy:**
- Bluish tinge of tympanic membrane
- **RISING SUN SIGN** - A reddish-blue mass behind an intact ear drum.
- **BROWN'S SIGN** – Blanching of mass on siegalization.
- Polyp might be seen coming out of External auditory canal.
- **AQUINO's SIGN** – Blanching of Tympanic membrane on gentle pressure of ipsilateral carotid artery.
- 30% of cases cranial nerve palsies. Mostly facial nerve affected

**CHAUSE OTOSCOPIC STAGES**
- Hyper vascular stage ➔ TM vessels dilated
- Tympanic Stage ➔ Rising Sun Sign
- Polypoidal or Haemorrhagic Stage ➔ Ear drum destroyed and tumour extends along the EAC.

**CLASSIFICATION:**

**LUNDGREN CLASSIFICATION**
1. Glomus tympanicum
2. Glomus Jugulare

**GLASSCOCK – JACKSON CLASSIFICATION** of temporal bone paragangliomas:

**GLOMUS JUGULARE**
- 1. Type I : Small tumor involving the **jugular bulb, middle ear and mastoid.**
- 2. Type II : Tumor extending **under the internal auditory canal**. There may be intracranial extension.
- 3. Type III : Tumor extending into the **Petrous Apex**. There may be intracranial extension.
- 4. Type IV : Tumor extending beyond the petrous apex into the **Clivus and Infratemporal Fossa.** There may be intracranial extension.
GLOMUS TYMPANICUM

- Type I → Only promontory
- Type II → Involves Middle ear space
- Type III → Middle ear and mastoid
- Type IV → Middle ear mastoid and EAC

THE FISCH CLASSIFICATION (MC) of glomus tumors is based on extension of the tumor to surrounding anatomic structures and is closely related to mortality and morbidity.

1. Type A tumor - Tumor limited to middle ear cleft (The globus tympanicus tumours, best prognosis)
2. Type B tumor - Tumor limited to the tympanomastoid area with no infralabyrinthine compartment involvement
3. Type C tumor - Tumor involving the infralabyrinthine compartment of temporal bone with extension to petrous apex

This is divided into three types: C1, C2 & C3.

- Type C1 - Tumor with limited involvement of the vertical portion of the carotid canal
- Type C2 - Tumor invading the vertical portion of the carotid canal
- Type C3 - Tumor invasion of the horizontal portion of the carotid canal

4. Type D tumor has 2 types
   - Type D1 - Tumor with an intracranial extension less than 2 cm in diameter
   - Type D2 - Tumor with an intracranial extension greater than 2 cm in diameter

D/D

- High Jugular Bulb
- Aberrant Carotid Artery
- Idiopathic Haemotympanum
- Cholesterol Granuloma
- Primary Brain Tumors.

INVESTIGATIONS:

- 2 conditions mimic the rising sun sign i.e. a high jugular bulb or aberrant carotid artery.
- Otology: look for pulsations of mass
- PTA = Shows mostly conductive hearing loss.

Radiology:

- Plain X-ray (Jugular Foramen view) – shows unilateral jugular foramen enlargement in A and B types of tumors with type C showing bony erosions.
- PHELP’s SIGN (On lateral Tomography) – Loss of Crest of bone between Jugular foramen and Carotid canal.
- Axial CT scan → Jugular fossa enlarged (with cortical erosion think of Glomus jugulare, without cortical erosion think of high jugular bulb) → IF Normal jugular fossa then go for coronal CT →
Normal carotid canal (Globus Tympanicus) or if laterally placed carotid canal then aberrant carotid artery.

- **MRI** with gadolinium enhancement (GOLD STANDARD) ➔ **Salt and Pepper Appearance**
- Better indication of soft tissue involvement but more difficult to correlate with anatomy
- **Carotid Angiography**: For tumour embolisation to prevent intra op bleeding
- For all cases positive for VMA, **prophylactic alpha blockade** should precede angiography.
- **Retrograde venography** of internal jugular vein was popular to differentiate between glomus jugulare and glomus tympanicus with glomus jugulare showing a filling defect (**no longer done**)
- **Endocrine assessment**: before surgery It is important to rule out any vasoactive hormone secretion by the tumour (mostly in cases with elevated BP). So 24hr VMA assessed.
- **Biopsy not indicated. Only done if SCC suspected.**

**MANAGEMENT:**

- Presently radiotherapy is only an adjunct to surgery.
- Current treatment plan:
  1. No treatment and continuous observation.
  2. Surgical resection
  3. SURGERY + RT
- **Observation**: > 60yrs and minimal growth.
- RT only has some effect on slowing tumour growth. (☞ Cerebral Necrosis or Radionecrosis of temporal bone can occur).
- For extensive tumours surgery usually not possible and in such cases RT is best option

**SURGERY**

- Aim is complete resection with minimal neurological deficit
- Type A tumours – i.e. Glomus tympanicus approached via EAC (Transmeatal Tympano Tomy Approach)
- Type B tumours - Intact canal wall procedure (combined approach i.e. Extended Facial Recess Approach)
- Type C tumours - Skull base approach utilizing Upper Cervical Dissection And Transmastoid Approach, Fisch Infratemporal Fossa Approach
- Type D tumours - Skull Base Approach With Posterior Fossa Craniotomy(some do as 2 stage procedure)

**Reducing Tumour Vascularity:**

- Radiation – causes stromal fibrosis, decrease vascularity and thus easier tumour dissection
- Pre operative tumour embolization (Gelfoam or Poly Vinyl Alcohol sponge i.e. ivalon) done 4 to 8 days prior to planned resection using TIVA after selective angiography.

**TRANSMEATAL APPRAOCH : (Type A tumours Tympanotomy)**

- Directly doing tympanotomy, sometimes dissecting malleus handle free of drum might be required
- Additional exposure provided by lowering inferior annulus in hypotympanic approach by removing mastoid tip and mobilizing vertical portion of facial nerve.
EXTENDED FACIAL RECESS APPROACH:

- For large **type A** or moderate **type B** tumours.
- Intact canal wall approach. By extending the facial recess inferiorly, reasonably good access is obtained to the hypotympanum particularly if chorda tympani is sacrificed. Even better exposure on skeletonising vertical portion of facial nerve, sigmoid sinus and Posterior SCC.
- Total tumour removal is possible without sacrificing Ossicular function.

INFRATEMPORAL FOSSA APPROACH (LATERAL APPROACH):

- Resection of jugular bulb after ligating internal jugular vein in neck and packing off sigmoid sinus superiorly with anterior transposition of facial nerve allow direct access to the jugular bulb.
- **FISCH's Infratemporal Fossa** approach:
  - **TYPE A/B/C**
    - Type A ➔ Access to temporal bone upto petrous apex.
    - Type B ➔ Anterior approach across the apex to basiocciput to clivus.
    - Type C ➔ Remove lesions of nasopharynx and parasellar regions

STEPS

- Postaural incision extended both superiorly and inferiorly in neck
- Facial flap and pinna raised and reflected Anteriorly
- Cartilagenous meatus is transected and closed off as blind ending sac.
- Parotid region is dissected to mobilize the peripheral branches of facial nerve, and nerves and vessels of upper neck are mobilized upto skull base.
- Control ligatures placed on Internal Jugular vein and Internal Carotid artery but not tied at this stage.
- Complete mastoidectomy (subtotal petrosectomy) done removing all air cells, posterior meatal wall, drum, malleus, incus.
- Facial nerve skeletonized along both vertical and horizontal portions, dissected off canal and permanently transposed Anteriorly
- If invaded then portion removed with sural nerve grafting.
- Sigmoid sinus ligated at sinodural angle and internal jugular vein ligated in neck.
- Tumour now mobilized first peripherally then centrally.
- If possible the medial wall of sigmoid sinus is preserved.
- At completion of surgery any dural defect is repaired with fascia graft.
- Eustachian tube closed off with bone wax and whole cavity filled with free fat graft.
- Anterior transposition of facial can lead to temp paresis which usually recovers in 2 to 3 months.
- Intracranial extensions more than 2 cm are best managed by second stage procedure to reduce chances of CSF leak and meningitis.

POSTEROLATERAL APPROACH:

- Modification to infratemporal approach of Fisch
- Without transposition of facial nerve
- Posterior craniotomy + Infratemporal approach of Fisch
- Radical mastoidectomy and facial nerve transposition was only required in these procedures with extensive tumour around ICA.

**MANAGEMENT OF SECRETORY GLOMUS JUGULARE TUMOURS:**

- Initial HTN requires both alpha and beta blockers
- Following embolisation and surgical resection loss of vasoconstrictor tone may result in circulatory collapse needing correction with massive i.v. fluids.
- So only 24 hr VMA necessary