Glomus Jugulare tumour:

- **Vascular tumour** of glomus body (sympathetic paraganglia like structure containing cells of neural crest origin with eosinophilic granules possibly functions as chemoreceptor and capable of secreting neurotransmitters such as norepinephrine, dopamine, serotonin and vasoactive intestinal peptide).Nerve supply is tympanic branch of IX (Jacobson) and auricular branch of X (Arnold). Similar to carotid body glomus tympanicum and glomus vagal tumours. **Most common middle ear tumour**. Does not produce epinephrine (lacks the enzyme that converts norepinephrine to epinephrine).
- 1/1000000. **F: M 6:1**. Peak of onset 50-60 years. 7% associated with carotid body tumour and in 10% multiple paragangliomas. Some cases are familial and transmitted as autosomal dominant with disease developing only if gene transmitted from the father (**gene imprinted during oogenesis**).
- Patients present with **conductive hearing loss**, otalgia, and tinnitus (pulsatile). Cranial nerves palsy (V, VII, VIII, IX, X, XI and XII). **1-3% of the tumours are hormonally active** (palpitations, labile blood pressure, perspiration and explosive diarrhoea similar to pheochromocytoma).
- Investigations:

1. Brain CT scan with bone windows to assess the degree of bone destruction or remodelling
2. MRI: enhancing tumours with **flow voids**. Differential diagnosis include meningiomas, vestibular schwannomas, chordomas, chondrosarcoma, metastasis, plasmocytoma and rhabdomyosarcoma
3. Cerebral angiogram: vascular blush, main blood supply is from external carotid artery in particular ascending pharyngeal branch. Consider embolisation
4. Balloon test occlusion with SPECT or Xenon CT scan if the tumour involves ICA
5. Serum and 24 hour urine for catecholamine and VMA(vanellylmendalic acid).Patients with active tumours
6. FBC, electrolytes, coagulation, cross match
7. Otoscopy: pink lesions behind the eardrum

- Pathology: encapsulated, lobulated tumours microscopically resemble the architecture of sympathetic ganglia (Clusters of epithelioid cells "Zellballen" found within highly vascular stroma
- Treatment:

1. Surgery: 1-3% mortality, 4% permanent VII paresis, 10% recurrence. Patients with hormonally active tumours can be dehydrated and anaemic and require preparation with alpha and beta blockers and hydration before surgery. Surgery through retrosigmoid infratemporal approach
2. Stereotactic radiosurgery:9 patients , 8 remained stable and 1 regressed at 4 years follow up

3. Conventional radiotherapy: 80% stabilisation rate. Reported serious complications include radionecrosis, delayed bone and dural necrosis – CSF leak, hypopituitarism, malignancy

- Glomus tympanicum: margin around the tumour 360 degrees on otoscopy. Bone between the tumour and jugular bulb