MedMentor EDU

ENT Study Notes | MBBS / NEET-PG

TUMOURS OF THE

MIDDLE EAR & MASTOID

Based on: Dhingra | Ramalingam | Logan Turner

Paragangliomas • Glomus Tumours • SCC • Facial Nerve Schwannoma

SECTION 1 | INTRODUCTION & OVERVIEW

1.1 Definition

Tumours of the middle ear and mastoid are neoplastic lesions arising from the various tissues of the tympanic cavity, mastoid air cells, temporal bone, and adjacent skull base. They may be primary (arising from local tissues) or secondary (metastatic).

1.2 Epidemiology

- **Rare overall** — middle ear tumours constitute <0.1% of head and neck neoplasms
- **Glomus tumours** are the most common benign tumours of the middle ear
- **Squamous cell carcinoma (SCC)** is the most common malignant tumour
- **Female predominance** in glomus tumours (3–6:1 female-to-male ratio)
- Peak incidence of glomus tumours: 5th–6th decade
- **Rhabdomyosarcoma** is the most common middle ear malignancy in children

1.3 Relevant Anatomy

1.3.1 Jugulotympanic Anatomy

- Jugular bulb lies in the jugular fossa below the floor of the middle ear
- Jugular foramen transmits: CN IX, X, XI and the internal jugular vein
- Inferior tympanic canaliculus transmits the tympanic branch of CN IX (Jacobson nerve)
- Mastoid canaliculus transmits the auricular branch of CN X (Arnold nerve)
- **Hypotympanum**: the floor of the tympanic cavity — immediately above the jugular bulb
- Carotid canal lies anteromedial to the jugular fossa — important surgical landmark

?? Figure 1.1: Jugulotympanic anatomy — jugular bulb, carotid canal, hypotympanum relationships

1.3.2 Paraganglion System Anatomy

- Paraganglia are clusters of neuroendocrine cells derived from neural crest
- **In the temporal bone/skull base**: found along Jacobson nerve (tympanic plexus) and Arnold nerve
- **Tympanic paraganglia**: on the promontory — give rise to glomus tympanicum
- **Jugular paraganglia**: in the jugular bulb adventitia — give rise to glomus jugulare
- Part of the diffuse neuroendocrine system (DNES) / APUD system

?? Figure 1.2: Distribution of paraganglia along the tympanic and vagal plexuses

1.3.3 Neural Crest Origin — Key Points

- Glomus tumours (paragangliomas) arise from neural crest-derived paraganglia
- Neural crest cells migrate to form adrenal medulla, peripheral ganglia, and dispersed paraganglia
- **APUD cell origin**: Amine Precursor Uptake and Decarboxylation — explains catecholamine-secreting potential
- SDH gene mutations (succinate dehydrogenase) are the commonest germline mutations in familial paraganglioma

SECTION 2 | CLASSIFICATION OF TUMOURS

2.1 Overall Classification

Category	Tumour Type
Benign	Glomus tympanicum, Glomus jugulare, Middle ear adenoma, Hemangioma, Facial nerve schwannon Cholesterol granuloma, Endolymphatic sac tumour, Giant cell tumour, Chondroblastoma, Eosinophil granuloma
Malignant	Squamous cell carcinoma (most common), Adenocarcinoma, Rhabdomyosarcoma, Langerhans cell histiocytosis, Metastatic tumours
Locally Aggressive	Paraganglioma (glomus jugulare — can invade skull base)

2.2 Benign Tumours — Summary

Tumour	Origin / Tissue	Key Feature

Glomus tympanicum	Paraganglia (Jacobson nerve)	Pulsatile mass on promontory
Glomus jugulare	Paraganglia (jugular bulb)	Jugular foramen erosion, CN palsy
Middle ear adenoma	Glandular epithelium	Neuroendocrine differentiation
Hemangioma	Vascular endothelium	Geniculate ganglion most common site
Cholesterol granuloma	Chronic haemorrhage / obstruction	Blue-black mass, cholesterol crystals
Facial nerve schwannoma	Schwann cells of CN VII	Facial palsy + middle ear mass
Endolymphatic sac tumour	Endolymphatic sac epithelium	Associated with VHL syndrome
Giant cell tumour	Osteoclast-like cells	Rare; temporal bone involvement
Chondroblastoma	Cartilage	Temporal bone, young patients
Eosinophilic granuloma	Langerhans cells	Bone destruction, part of LCH spectrum

2.3 Malignant Tumours — Summary

Tumour	Age Group	Key Feature
Squamous cell carcinoma	Adult (>50 yr)	Most common; chronic otitis predisposes
Adenocarcinoma	Adult	Rare; from middle ear glands
Rhabdomyosarcoma	Children (<10 yr)	Most common middle ear malignancy in children; ra
Langerhans cell histiocytosis	Children	Punched-out bone lesions; systemic disease
Metastatic tumours	Variable	From breast, kidney, lung, prostate, thyroid

SECTION 3 | GLOMUS TUMOURS (PARAGANGLIOMAS)

3.1 General Features

3.1.1 Definition

Glomus tumours (also called **paragangliomas** of the temporal bone) are benign, highly vascular neoplasms arising from paraganglia associated with the tympanic plexus (Jacobson nerve) or the jugular bulb (Arnold nerve). They are the **most common benign tumour of the middle ear** and the **second most common tumour of the temporal bone** (after acoustic neuroma).

? COMMON MCQ

- Glomus tumour = most common benign tumour of middle ear
- Glomus jugulare = most common tumour of jugular foramen
- Female predominance: 3–6:1 (peak: 5th–6th decade)
- Most are BENIGN; malignant transformation in <5%

3.1.2 Origin and Biology

- **Arising from:** paraganglia — chemoreceptor tissue derived from neural crest
- **Chromaffin-negative**: unlike adrenal pheochromocytoma — most do NOT secrete catecholamines (but ~1–3% may)
- **Highly vascular:** fed by multiple branches — ascending pharyngeal, posterior auricular, occipital arteries
- **Slow growing**: but locally invasive — erodes bone, tracks along nerves, invades vascular structures
- **Multicentric tumours**: bilateral or multiple paragangliomas in 10% (familial cases: up to 25–30%)

3.1.3 Female Predominance

- F:M ratio = 3–6:1
- Oestrogen receptors demonstrated in some glomus tumours
- Postmenopausal decline in some cases supports hormonal influence

3.1.4 Familial Paraganglioma

- **SDH gene mutations**: most important — SDHB, SDHC, SDHD subunits of succinate dehydrogenase
- **SDHD mutation**: commonest; autosomal dominant with genomic imprinting — maternal silencing; expressed only with paternal transmission
- **SDHB mutation**: highest risk of malignant transformation and metastasis
- VHL (von Hippel-Lindau) syndrome: endolymphatic sac tumour + paraganglioma
- Carney triad: gastric GIST + paraganglioma + pulmonary chondroma (rare, non-hereditary)

3.1.5 Catecholamine-Secreting Tumours

- Only ~1–3% of head and neck paragangliomas are functional (secrete catecholamines)
- **Suspect if**: hypertension, headache, palpitations, diaphoresis, anxiety attacks
- **Investigations**: 24-hour urinary catecholamines, metanephrines, vanillylmandelic acid (VMA)
- **CRITICAL**: do not operate without controlling catecholamine excess — risk of hypertensive crisis

3.2 Blood Supply

- **Primary feeder:** Ascending pharyngeal artery (ICA branch of external carotid — via inferior tympanic branch)
- **Posterior auricular artery**: supplies mastoid component
- **Occipital artery**: contributes to posterior fossa extension
- **Internal carotid branches**: caroticotympanic arteries in advanced disease
- **AICA/PICA**: supply intracranial extensions

?? Figure 3.1: Angiographic feeder vessels of glomus jugulare — ascending pharyngeal artery blush

3.3 Classification

3.3.1 Fisch Classification

Most widely used surgical classification — based on extent of disease

Type	Extent	Key Features
Type A	Tympanic cavity only	Confined to middle ear; arising from tympanic plexus (promontory)
Type B	Tympanomastoid	Tympanic cavity + mastoid; no infralabyrinthine extension; bone of hypotympanum intact
Type C	Infralabyrinthine (jugular bulb + ICA)	C1: minimal carotid canal involvement; C2: partial ICA; C3: full IC cavernous ICA
Type D	Intracranial extension	D1: <2 cm extradural; D2: >2 cm extradural; De: intradural

3.3.2 Glasscock-Jackson Classification

Glomus Tympanicum

Type	Description
Type I	Small mass limited to promontory

Type II	Tumour completely filling middle ear space
Type III	Tumour filling middle ear + extending into mastoid
Type IV	Tumour filling middle ear, extending into mastoid, EAC; may extend anterior to ICA

Glomus Jugulare

Type	Description
Type I	Small tumour involving jugular bulb, middle ear, mastoid
Type II	Extends below IAC; may involve ICA intrapetrous
Type III	Extends into petrous apex; may extend into cavernous sinus
Type IV	Extends beyond petrous apex to clivus / infratemporal fossa

? VERY HIGH-YIELD — CLASSIFICATION

- Fisch classification most important for surgery — Type A = tympanic only; Type D = intracranial
- Glomus tympanicum Type I = small promontory mass; most curable
- Glomus jugulare Type III/IV = skull base invasion; requires combined approach

SECTION 4 | ETIOLOGY & PATHOGENESIS

4.1 Etiology

- **Sporadic (most cases — >90%):** no identifiable genetic cause
- **Genetic/Familial:** SDH mutations, VHL gene, NF1, RET proto-oncogene
- **Radiation exposure**: prior radiation to head and neck is a risk factor (dose-dependent)
- **Chronic hypoxia**: carotid body and jugulotympanic paraganglia are chemoreceptors — chronic hypoxia (high-altitude dwelling) may promote hyperplasia

4.2 Pathogenesis — Flowchart

Neural crest-derived paraganglia (tympanic / jugular)

?

Genetic trigger (SDH mutation / VHL) OR sporadic factors

?

Monoclonal neoplastic proliferation of chief cells

?

Slow but progressive expansion

?

Bone erosion via osteoclast activation + pressure necrosis

?

Vascular recruitment — hypervascular tumour

?

Spread along paths of least resistance:

?

Nerves ? Air cells ? Vascular structures ? Skull base

?

Intracranial extension (Fisch Type D)

SECTION 5 | PATHOLOGY

5.1 Gross Pathology

- **Appearance**: reddish-brown, lobulated, highly vascular tumour — bleeds profusely on biopsy
- **Consistency**: firm to soft; highly friable — DO NOT biopsy blindly (risk of catastrophic haemorrhage)
- **Bone erosion**: smooth, expanding margins; erodes jugular foramen, carotid canal, hypotympanum

?? Figure 5.1: Gross specimen glomus jugulare — reddish-brown hypervascular tumour with bone destruction

5.2 Histopathology — Zellballen Pattern

? FAVOURITE EXAM QUESTION — HISTOPATHOLOGY

- Zellballen pattern = pathognomonic of paraganglioma
- Zell = cells; Ballen = balls ? nests of chief cells surrounded by sustentacular cells
- Abundant thin-walled vascular sinusoids between nests

- **Zellballen pattern**: **nests of chief cells** (type I cells) surrounded by **sustentacular cells** (type II cells), embedded in a rich fibrovascular stroma

- **Chief cells (type I)**: large polygonal cells; eosinophilic granular cytoplasm; round nuclei; contain neurosecretory granules; immunopositive for synaptophysin, chromogranin A, NSE
- **Sustentacular cells (type II)**: spindle-shaped; support cells around nests; S-100 positive
- **Vascularity**: abundant thin-walled sinusoidal vessels throughout — explains dramatic bleeds
- **Malignant paraganglioma**: NO reliable histological criteria; malignancy defined by **distant metastasis** (bone, liver, lung, lymph nodes)

?? Figure 5.2: Histopathology — Zellballen pattern, chief cells, sustentacular cells, sinusoidal vascularity (H&E stain)

5.2.1 Immunohistochemistry

Marker	Cell Type	Result
Synaptophysin	Chief cells	Positive (+)
Chromogranin A	Chief cells	Positive (+)
NSE (Neuron-specific enolase)	Chief cells	Positive (+)
S-100 protein	Sustentacular cells	Positive (+)
Cytokeratin	Carcinoma	Negative (differentiates from carcinoma)

5.3 Pathways of Spread

- **Local spread**: expands along aerated spaces, air cells, and fascial planes

- **Jugular foramen spread**: erodes floor of middle ear ? hypotympanum ? jugular bulb ? jugular foramen ? CN IX, X, XI palsies
- **Carotid canal involvement**: tracks along carotid sheath — may encircle or invade ICA
- **Sigmoid sinus**: tumour may occlude sigmoid sinus ? venous hypertension / intracranial complications
- **Skull base extension**: petrous apex, cavernous sinus, clivus — Fisch C3/C4
- **Intracranial extension**: usually extradural; intradural invasion (Type De) — contact with posterior fossa structures
- **Lower cranial nerve involvement**: CN IX–XII within jugular foramen and posterior fossa — causes Jugular Foramen Syndrome

?? Figure 5.3: Pathways of spread — jugular foramen erosion, ICA involvement, sigmoid sinus, intracranial extension

SECTION 6 | CLINICAL FEATURES

6.1 Symptoms

Early Symptoms

- **Pulsatile tinnitus**: **most common and earliest symptom** — synchronous with pulse; low-pitched
- **Conductive hearing loss**: due to middle ear involvement and ossicular involvement
- Aural fullness / pressure sensation
- **Otalgia**: mild, intermittent

Late / Advanced Symptoms

- **Otorrhagia**: blood-stained discharge — tumour ulceration through TM or EAC
- **Facial palsy (CN VII)**: tympanic segment involvement — 5–10% of cases
- **Dysphagia**: CN IX/X involvement — swallowing difficulty
- **Hoarseness**: CN X involvement — vocal cord palsy
- **Aspiration**: due to combined CN IX/X palsy
- **Vertigo**: inner ear invasion (rare)
- **Raised ICP symptoms**: headache, vomiting — with intracranial extension or sigmoid sinus occlusion

6.2 Classical Signs

Brown Sign

BROWN SIGN (Pulsatile mass blanching sign): On pneumatic otoscopy — compression of air blanches the red pulsatile mass; release ? refills and pulsates. Specific for glomus tumour. Also known as the **blanching sign** or vascular compressibility sign.

Rising Sun Sign

RISING SUN SIGN: Seen on otoscopy — a reddish/bluish pulsatile mass visible through the intact tympanic membrane (particularly in the posteroinferior quadrant), resembling a rising sun appearing above the horizon. Characteristic of glomus tympanicum.

6.3. Jugular Foramen Syndrome (Vernet Syndrome)

Cranial Nerve	Deficit
CN IX (Glossopharyngeal)	Loss of taste posterior 1/3 tongue; reduced gag reflex
CN X (Vagus)	Hoarseness; dysphagia; ipsilateral vocal cord palsy
CN XI (Accessory)	Weakness of sternocleidomastoid and trapezius

Syndrome Name	Nerves Affected	Cause/Location
Vernet (Jugular foramen)	CN IX, X, XI	Jugular foramen
Collet-Sicard	CN IX, X, XI, XII	Posterior condylar area
Villaret	CN IX, X, XI, XII + Horner	Retroparotid space

Jackson	CN X, XI, XII	Posterior fossa / medulla

SECTION 7 | DIFFERENTIAL DIAGNOSIS

7.1 Causes of Pulsatile Tinnitus

- **Vascular:** glomus tumour, aberrant ICA, high/dehiscent jugular bulb, AV fistula, carotid artery stenosis
- **Non-vascular:** palatal myoclonus, patulous Eustachian tube, middle ear effusion

7.2 Red Pulsatile Retrotympanic Mass — Differentials

Condition	Key Differentiating Features
Glomus tympanicum	Most common; promontory origin; Brown sign positive; CT shows no jugular foramen eros
Glomus jugulare	Jugular foramen erosion on CT; larger; lower CN palsies
Aberrant internal carotid artery	Pulsatile; anterior/inferior TM; ICA does not lie medially on CT — lateral displacement
Dehiscent/high jugular bulb	Blue-tinged; posteroinferior location; no erosion; CT shows high riding bulb

Hemangioma	Geniculate fossa; facial nerve palsy; honeycomb CT pattern
Cholesterol granuloma	Blue-black, non-pulsatile; CT shows non-enhancing fluid-filled cavity
Aural polyp	Non-pulsatile; pale/grey; associated with chronic otitis

? VERY HIGH-YIELD — DIFFERENTIALS

- NEVER biopsy a pulsatile middle ear mass without imaging — aberrant ICA mimics glomus perfectly
- Aberrant ICA: risk of catastrophic haemorrhage if biopsied
- Brown sign: specific for glomus; absent in aberrant carotid
- CT shows: Glomus = jugular foramen erosion; Aberrant ICA = lateral carotid canal

SECTION 8 | INVESTIGATIONS

8.1 Clinical Investigations

- **Otoscopy**: reddish pulsatile mass on promontory; Brown sign; rising sun appearance
- **Pneumatic otoscopy**: Brown sign (blanching on compression)
- **Pure tone audiometry (PTA)**: conductive hearing loss (ossicular involvement); sensorineural loss (inner ear invasion)
- **Tympanometry**: Type As (stiff) or flat type B curve; pulsatile movements on tympanogram

8.2 Imaging — HRCT Temporal Bone

- **Investigation of choice** for initial assessment and classification
- **Glomus tympanicum**: soft tissue mass on promontory; jugular foramen — INTACT
- **Glomus jugulare**: irregular **'moth-eaten' / permeative erosion** of jugular foramen, hypotympanum
- **Carotid canal**: check for ICA involvement
- **Sigmoid sinus**: check for sinus involvement
- Bone detail: assess tegmen, otic capsule, facial canal

?? Figure 8.1: HRCT temporal bone — permeative/moth-eaten erosion of jugular foramen in glomus jugulare

8.3 MRI with Gadolinium — Salt-and-Pepper Appearance

SALT-AND-PEPPER SIGN (MRI): Characteristic MRI appearance of glomus tumours — multiple flow voids (dark = 'pepper') within the hypervascular tumour matrix (bright = 'salt') on T1/T2 imaging. Pathognomonic of paraganglioma.

- **T1:** Intermediate signal with flow voids (pepper); enhances avidly with gadolinium
- **T2:** Heterogeneous; flow voids prominent
- **T1 + Gad:** Intense enhancement — most helpful for tumour delineation and intracranial extension
- **MRV (MR venography):** Assess sigmoid sinus and jugular vein patency — critical for surgical planning
- **MRA:** Assess ICA displacement/encasement

?? Figure 8.2: MRI T2 — salt-and-pepper appearance of glomus jugulare; multiple flow voids within heterogeneous tumour

8.4 Angiography and DSA

- **Digital Subtraction Angiography (DSA)**: gold standard for vascular mapping prior to embolization + surgery
- **Four-vessel angiography**: bilateral carotid (ICA + ECA) + vertebral — assess all feeders and contralateral collaterals
- **Angiographic blush**: hypervascular tumour stain — characteristic pattern with early arterial filling
- **Balloon occlusion test (BOT)**: temporary ICA balloon occlusion under neurological monitoring — assesses safety of ICA sacrifice if encased by tumour

?? Figure 8.3: DSA — angiographic blush from ascending pharyngeal artery feeder vessels in glomus jugulare

8.5 Functional Imaging

- **Urinary catecholamines**: 24-hour urine for adrenaline, noradrenaline, dopamine, VMA, metanephrines — screen all glomus tumours
- **Octreotide scintigraphy (In-111 octreotide scan / OctreoScan)**: paragangliomas express somatostatin receptors — positive scan; useful for multi-focal/metastatic disease
- **DOPA-PET / Ga-68 DOTATATE PET-CT**: most sensitive for detecting multifocal and metastatic paraganglioma — superseding octreotide scan in modern practice

SECTION 9 | TREATMENT OF GLOMUS TUMOURS

9.1 Treatment Options Overview

Modality	Indication	Goal
Observation (Watch & Wait)	Elderly, infirm, small asymptomatic tumours, bilateral tumours	Monitor growth; avoid morbidity
Surgery	Fisch A/B; selected C/D; younger patients; functional	Complete resection; cure
Radiotherapy / SRS	Elderly, inoperable, residual/recurrent, poor surgical candidates	Tumour control (not cure); halt growth
Embolization	Preoperative for all glomus jugulare; palliative for inoperable	Reduce blood loss; shrink feeders

9.2 Observation (Watch & Wait)

- **Indications**: elderly patients (>70 yr), significant comorbidities, small asymptomatic tympanicum, bilateral tumours (Fisch A), slow/no growth on serial MRI
- MRI every 6–12 months for first 2 years; then annually
- If growing or symptoms develop ? active treatment

9.3 Surgical Treatment

9.3.1 Glomus Tympanicum — Surgical Approaches

- **Transcanal / Trans-tympanic approach**: Fisch Type A / Glasscock-Jackson Type I — simple, performed under local or general anaesthesia; excellent view of promontory
- **Extended transcanal (hypotympanotomy)**: for Type II tumours — remove scutum, lower anterior EAC wall
- **Facial recess approach**: Type III — posterior tympanotomy for posterior extension
- **Mastoidectomy + tympanotomy**: for mastoid extension

9.3.2 Glomus Jugulare — Surgical Approaches

- **Infratemporal fossa approach Type A (ITFA-A)**: Fisch approach — most commonly used for glomus jugulare; anterior transposition of facial nerve; blind sac closure of EAC; removal of ICA infrapetrous; sigmoid sinus control
- **Infratemporal fossa approach Type B/C**: for petrous apex and cavernous sinus extension
- **Combined skull base approaches**: multidisciplinary team (neurosurgeon + skull base ENT) for intracranial extension

?? Figure 9.1: Fisch infratemporal fossa approach Type A — facial nerve anterior transposition, jugular bulb exposure, ICA mobilisation

? HIGH-YIELD — SURGERY

- Infratemporal fossa approach Type A = standard approach for glomus jugulare
- Facial nerve is ANTERIORLY transposed in ITFA — causes temporary paresis
- EAC is closed as 'blind sac' to prevent CSF leak
- Preoperative embolization always done 24–48 hours before surgery
- ICA control must be obtained before tumour removal

9.4 Preoperative Embolization

- **Timing**: 24–48 hours before surgery (allows tumour oedema/necrosis without allowing new vessel formation)
- **Technique**: superselective catheterisation of feeding vessels (ascending pharyngeal, posterior auricular, occipital); PVA particles / coils used
- **Benefits**: reduces intraoperative blood loss significantly (up to 50–70%); improves tumour devascularisation; easier dissection
- **Risks of embolization**: stroke (ICA/ECA anastomoses), cranial nerve infarction, tumour swelling causing worsening CN palsy, non-target embolization
- **Contraindications to ICA branch embolization**: dangerous anastomoses between ECA and ICA (e.g., petrous ICA)

9.5 Radiotherapy and Stereotactic Radiosurgery

Modality	Dose	Indication	Outcome
Conventional RT (EBRT)	45–50 Gy in fractions	Large tumours, poor surgical candidates, residual disease	Tumour growth arrest ~
Gamma Knife (SRS)	12–15 Gy single fraction	Fisch A/B, small C; elderly; residual; good results	Growth control >95% at preservation good
CyberKnife (SBRT)	18–25 Gy in 3–5 fractions	Larger lesions near critical structures	Similar to gamma knife; reduces CN risk

- **Mechanism**: radiation causes vascular endothelial damage ? progressive fibrosis ? growth arrest (not cell kill)
- **NOT curative**: tumour persists but does not grow; viable cells remain on biopsy

- **Complication**: radiation-induced temporal bone osteonecrosis (rare); CN injury (generally low with SRS)

SECTION 10 | COMPLICATIONS

10.1 Tumour-Related Complications

- **Facial nerve palsy (CN VII)**: tympanic segment invasion — most feared otological complication
- **Lower cranial nerve palsy (CN IX–XII)**: jugular foramen syndrome — dysphagia, hoarseness, aspiration risk
- **Intracranial extension**: venous hypertension, raised ICP, posterior fossa compression
- **ICA encasement**: risk of catastrophic haemorrhage, stroke
- **Sensorineural hearing loss**: inner ear invasion

10.2 Surgical Complications

- **Massive intraoperative haemorrhage**: from ICA or sigmoid sinus injury — primary cause of operative mortality
- **CSF leak**: dural breach — managed with fat packing, lumbar drain
- **Facial nerve injury**: dehiscent nerve, tumour adherence, traction injury — temporary palsy common with ITFA; permanent in 5–15%
- **Lower CN injury**: especially CN IX/X/XII during jugular fossa dissection
- **Stroke**: ICA manipulation, embolization complication
- **Aspiration**: combined CN IX/X injury — may require nasogastric tube / PEG
- **Infection / meningitis**: CSF leak + contamination
- **Recurrence**: 5–15% in completely resected tumours; higher with incomplete removal

SECTION 11 | PROGNOSIS

11.1 Prognosis of Glomus Tumours

- **Overall survival**: excellent for benign disease — 10-year survival >90%
- **Glomus tympanicum**: nearly 100% cure with complete resection (Fisch Type A)
- **Glomus jugulare**: long-term control with surgery ± SRS — 5-yr tumour control 85–95%
- **Recurrence rates**: 5–15% after surgery alone; SRS reduces recurrence in residual disease
- **Malignant transformation**: rare (<5%) — SDHB mutations carry highest risk of metastasis
- **CN recovery**: pre-existing lower CN palsies have limited recovery; facial nerve generally recovers well if preserved

SECTION 12 | OTHER MIDDLE EAR TUMOURS

12.1 Middle Ear Adenoma (Adenomatous Neuroendocrine Tumour)

- **Origin**: secretory/respiratory epithelium of middle ear mucosa; shows BOTH glandular and neuroendocrine differentiation
- **Clinical**: conductive hearing loss; middle ear mass; rarely facial palsy; NO pulsatility
- **Histology**: glandular structures + neuroendocrine markers (chromogranin, synaptophysin) — dual differentiation
- **Treatment**: surgical excision; recurrence possible; rarely malignant transformation (carcinoid tumour)

12.2 Hemangioma of Temporal Bone

- **Location**: most common at geniculate ganglion; also IAC, jugular foramen
- **Clinical**: facial palsy + conductive hearing loss; pulsatile tinnitus (if vascular)
- **CT**: honeycomb pattern — spiculated bone with interspersed vascular channels; diagnostic
- **MRI**: T2 bright; enhances with gadolinium
- **Treatment**: surgical excision — often requires facial nerve mobilisation; good functional recovery if nerve anatomically preserved

?? Figure 12.1: HRCT temporal bone — honeycomb pattern of hemangioma at geniculate ganglion

12.3 Cholesterol Granuloma

- **Not a true neoplasm**: a reactive lesion from chronic haemorrhage + obstruction in pneumatised spaces
- **Petrous apex**: most common site; also middle ear / mastoid
- **Pathology**: cholesterol crystals + haemorrhage products + foreign body giant cells + fibrous tissue

- **Clinical**: blue-black middle ear mass (if middle ear); sensorineural/conductive HL; facial palsy; headache
- **MRI**: T1 **BRIGHT** (methaemoglobin/cholesterol) + T2 bright — **unique MRI appearance** (no flow voids unlike glomus)
- **Treatment**: surgical drainage and ventilation — drainage via infralabyrinthine or translabyrinthine approach

12.4 Facial Nerve Schwannoma

- **Origin**: Schwann cells of any segment of intratemporal facial nerve
- **Most common segment**: geniculate ganglion / labyrinthine / tympanic segment
- **Clinical**: slowly progressive facial palsy (in 50–60%); conductive/SNHL; middle ear mass
- **Paradox**: facial function may be NORMAL or only mildly affected despite large tumour — because slow growth allows adaptation
- **CT**: fusiform expansion of facial canal — smooth, well-defined margins
- **MRI**: T1 isointense; T2 bright; enhances with gadolinium; along course of CN VII
- **Treatment**: observation if function good; surgery (cable graft) if significant palsy; SRS for small lesions

12.5 Endolymphatic Sac Tumour (ELST)

- **Association**: strongly associated with **Von Hippel-Lindau (VHL) syndrome** — VHL gene mutation (chromosome 3p)
- 2–16% of VHL patients develop ELST
- **Clinical**: SNHL, vertigo, tinnitus — mimics Meniere's disease; facial palsy in advanced cases
- **CT**: retrolabyrinthine mass with irregular bone erosion; may show calcification
- **MRI**: heterogeneous T1/T2 signal; avid enhancement
- **Histology**: papillary adenocarcinoma — locally aggressive, rarely metastasizes
- **Treatment**: surgical excision (translabyrinthine or retrosigmoid approach); screen VHL patients routinely

SECTION 13 | MALIGNANT TUMOURS OF MIDDLE EAR

13.1 Squamous Cell Carcinoma (SCC) of Middle Ear

13.1.1 Introduction

- **Most common malignant tumour** of the middle ear and temporal bone
- Incidence: 1–6 per million population annually
- Peak age: 6th–7th decade; slight male predominance

13.1.2 Etiology / Risk Factors

- **Chronic suppurative otitis media (CSOM)**: strongest association — chronic inflammation, keratin accumulation, carcinogenic byproducts
- **Cholesteatoma**: some debate — chronic inflammation with enzymatic bone destruction may predispose
- **Radiation**: prior therapeutic radiation
- Human papillomavirus (HPV): emerging association
- Arsenic or kerosene exposure (historical)

? COMMON MCQ — SCC MIDDLE EAR

- Most common malignant tumour of middle ear = SCC
- Strongest association = Chronic suppurative otitis media
- Suspect malignancy if: blood-stained discharge + otalgia + facial palsy in CSOM
- Pittsburgh staging system most used for middle ear SCC

13.1.3 Pathology

- **Gross**: irregular, ulcerated, friable mass within middle ear; extends to EAC, mastoid, petrous apex

- **Histology**: well-differentiated to poorly differentiated SCC; keratin pearls; intercellular bridges
- Bone invasion: permeative erosion of temporal bone
- Lymph node metastasis: to preauricular, parotid, and upper deep cervical nodes

?? Figure 13.1: Histopathology SCC — keratin pearls, intercellular bridges, invasive nests of squamous cells (H&E)

13.1.4 Clinical Features

- **Early**: blood-stained otorrhoea, otalgia, conductive hearing loss — often mistaken for CSOM exacerbation
- **Late**: facial palsy (CN VII), trismus (pterygoid involvement), vertigo (inner ear invasion), lower CN palsies, meningitis
- **Key warning signs**: otalgia out of proportion to discharge; blood-stained discharge in CSOM; **facial palsy in CSOM** — must biopsy

13.1.5 Pittsburgh Staging System for Temporal Bone SCC

Stage	Description
T1	Tumour limited to EAC without bony erosion or <0.5 cm soft tissue involvement
T2	Tumour with bone erosion of EAC <0.5 cm or middle ear extension
T3	Full-thickness erosion of EAC + limited middle ear / mastoid extension; facial palsy
T4	Tumour erodes cochlea, petrous apex; dura; carotid; jugular; infratemporal fossa; parotid

13.1.6 Treatment of SCC Middle Ear

- **Surgery**: **lateral temporal bone resection (LTBR)** for T1/T2; **subtotal / total temporal bone resection** for T3/T4 (combined with neurosurgery)
- **Radiotherapy**: adjuvant post-op — 60–65 Gy; primary RT for inoperable cases
- **Chemoradiation**: concurrent cisplatin-based regimen for advanced/unresectable
- **Parotidectomy + neck dissection**: if parotid or nodal involvement

13.1.7 Prognosis

- 5-year survival: T1/T2 — 60–80%; T3 — 40–50%; T4 — <20%
- Poor prognostic factors: dural invasion, carotid involvement, facial palsy, nodal metastasis

13.2 Rhabdomyosarcoma

- **Most common middle ear malignancy in children**: peak age 2–5 years
- **Histology**: embryonal type most common in head and neck region
- **Clinical**: rapid-onset otorrhoea + polyp + facial palsy in a child — DIAGNOSTIC TRIAD
- **Radiology**: extensive bone destruction; soft tissue mass
- **Treatment**: multimodal — chemotherapy (VAC = vincristine + actinomycin + cyclophosphamide) + radiation + limited surgery for residual disease
- **Prognosis**: improved with modern protocols — 5-year survival ~70% in localised disease

? IMPORTANT — PAEDIATRIC

- Any child with aural polyp + facial palsy + rapid onset ? RULE OUT rhabdomyosarcoma
- Embryonal RMS most common histological type in head and neck
- Treatment: chemotherapy is the mainstay (unlike adult SCC where surgery leads)

13.3 Langerhans Cell Histiocytosis (LCH)

- **Previously called**: Histiocytosis X / Eosinophilic granuloma (localised bone disease)
- **Middle ear LCH**: lytic ('punched-out') lesions of temporal bone; otorrhoea, aural polyp, HL
- **Histology**: Langerhans cells + eosinophils; CD1a+ and S-100+ and CD207 (Langerin)+

- **Radiological**: 'punched-out' lytic bone lesion — characteristically **geographic** bone destruction
- **Treatment**: systemic steroids, vinblastine + prednisolone (multi-system); curettage/radiation for localised bone disease

13.4 Metastatic Tumours

- **Haematogenous spread**: from breast, kidney (renal cell carcinoma), lung, prostate, thyroid
- **Clinical**: rapidly progressive SNHL or facial palsy; no primary middle ear disease history
- **Management**: directed at primary malignancy + palliative radiotherapy to temporal bone

SECTION 14 | MASTER COMPARISON TABLES

14.1 Glomus Tympanicum vs Glomus Jugulare

Feature	Glomus Tympanicum	Glomus Jugulare
Origin	Jacobson nerve on promontory	Paraganglia of jugular bulb
Location	Middle ear (promontory)	Jugular foramen + middle ear
Jugular foramen erosion	ABSENT	PRESENT (moth-eaten)
Fisch Type	Type A (confined to middle ear)	Type C/D (jugular/intracranial)
Lower CN palsy	Absent	Present (IX, X, XI, XII)
CT finding	Promontory soft tissue mass; intact floor	Permeative jugular fossa erosion
Primary treatment	Transcanal excision	Infratemporal fossa approach Type A
Cure rate	Near 100% (Type A)	85–95% control with surgery + SRS

14.2 Benign vs Malignant Middle Ear Tumours

Feature	Benign (Glomus)	Malignant (SCC)
Growth rate	Slow (years to decades)	Rapid
Symptoms onset	Gradual	Rapid deterioration
Otoscopy	Pulsatile red mass, Brown sign	Irregular mass, granulations, ulceration
Discharge	Absent / blood-stained (late)	Blood-stained, foul-smelling
Otalgia	Mild	Severe
Bone erosion CT	Smooth expanding erosion	Permeative / irregular moth-eaten
Metastasis	Rare (<5%)	To parotid and neck nodes
Treatment	Surgery / SRS / observation	Surgery + adjuvant RT/CRT

14.3 Lower Cranial Nerve Palsy Syndromes

Syndrome	CN Involved	Site of Lesion
Vernet (Jugular foramen)	IX, X, XI	Jugular foramen
Collet-Sicard	IX, X, XI, XII	Posterior condylar / retroparotid
Villaret	IX, X, XI, XII + Horner	Retroparotid space
Jackson	X, XI, XII	Posterior fossa / dorsal medulla
Tapia	X, XII	Parapharyngeal or vagal ganglion

14.4 Surgical Approaches — Quick Reference

Approach	Fisch Type	Features
Transcanal	Type A (small)	Minimal; local/GA; promontory only
Extended transcanal (hypotympanotomy)	Type A/B (medium)	Scutum removal; inferoposterior approach

Facial recess approach	Type B	Posterior tympanotomy
Mastoidectomy + tympanotomy	Type B	For mastoid extension
Infratemporal fossa Type A (ITFA-A)	Type C/D	Anterior CN VII transposition; ICA control; sigmoid ligatio
ITFA Type B	Petrous apex	Mandibular condyle removed; ICA petrous exposure
Combined skull base	Type De (intradural)	Multidisciplinary; neurosurgery + ENT

14.5 Radiology Summary Table

Investigation	Glomus Tympanicum	Glomus Jugulare	SCC
CT finding	Soft tissue mass on promontory; jugular fossa intact	Moth-eaten/permeative erosion jugular foramen	Irregular perme bone destructio
MRI T1+Gad	Intense enhancement	Salt-and-pepper appearance; intense enhancement	Enhancing irreg invasion
DSA	Minimal blush	Florid angiographic blush; ascending pharyngeal feeder	Not routinely u

PET-CT	Ga-DOTATATE positive	Ga-DOTATATE positive; use for multifocal/met	FDG-PET for s

SECTION 15 | HIGH-YIELD EXAM PEARLS

15.1 Must-Know Facts — Glomus Tumours

2 GLOMUS TUMOURS — EXAM ESSENTIALS

- Glomus tumour = most common benign tumour of middle ear; 2nd most common temporal bone tumour
- Glomus jugulare = most common tumour of jugular foramen
- Female predominance: 3–6:1; peak 5th–6th decade
- Arise from paraganglia (neural crest origin); part of APUD system
- Most are BENIGN; malignancy = distant metastasis (no histological criteria)
- Pulsatile tinnitus = most common presenting symptom
- Brown sign = blanching of pulsatile mass on pneumatic otoscopy
- Rising sun sign = reddish pulsatile mass visible through intact TM
- Salt-and-pepper MRI = flow voids within enhancing tumour — pathognomonic
- Zellballen pattern = nests of chief cells + sustentacular cells — histological hallmark
- NEVER biopsy a pulsatile middle ear mass without imaging — risk of massive haemorrhage
- Fisch Type A = middle ear only ? transcanal excision ? near 100% cure
- Fisch Type C/D = jugular/intracranial ? ITFA Type A ? surgery + SRS
- Preoperative embolization: 24–48 hours before surgery; ascending pharyngeal artery primary feeder
- Familial paraganglioma: SDHD mutation most common; SDHB = highest malignant risk
- Catecholamine screen (24-hr urine) all glomus patients before surgery

15.2 Must-Know Facts — Malignant Tumours

2 MALIGNANT TUMOURS — EXAM ESSENTIALS

- SCC = most common malignant tumour of middle ear; associated with CSOM
- Rhabdomyosarcoma = most common middle ear malignancy in CHILDREN
- Blood-stained discharge + otalgia + facial palsy in CSOM ? suspect SCC
- Rapid onset otorrhoea + aural polyp + facial palsy in CHILD ? suspect Rhabdomyosarcoma

- Pittsburgh staging for temporal bone SCC — T4 = carotid/dura/infratemporal fossa
- Lateral temporal bone resection = surgery for T1/T2 SCC
- LCH: punched-out geographic lytic bone lesions; CD1a+ / S-100+ / Langerin+
- Endolymphatic sac tumour ? VHL syndrome; screen all VHL patients for ELST
- Facial nerve schwannoma: slowly progressive facial palsy + fusiform expansion of facial canal on CT
- Hemangioma: geniculate ganglion + honeycomb CT pattern
- Cholesterol granuloma: T1-bright (bright on MRI) — unique; no flow voids

15.3 Important Mnemonics

GLOMUS — Features:

- **G** — Gadolinium MRI = Salt-and-Pepper appearance
- **L** — Low-pitched pulsatile tinnitus
- **O** — Origin = paraganglia (neural crest)
- **M** — Most common benign middle ear tumour
- **U** — Under Brown sign ? blanches with pressure
- **S** — SDH mutation in familial cases; SDHB = malignant risk

Fisch Classification Memory Aid:

- **A** = Attic/middle ear ONLY
- **B** = Both middle ear + mastoid (Bone of hypotympanum intact)
- **C** = Carotid / Infralabyrinthine extension
- **D** = Dural / Intracranial extension

15.4 Important Viva Questions

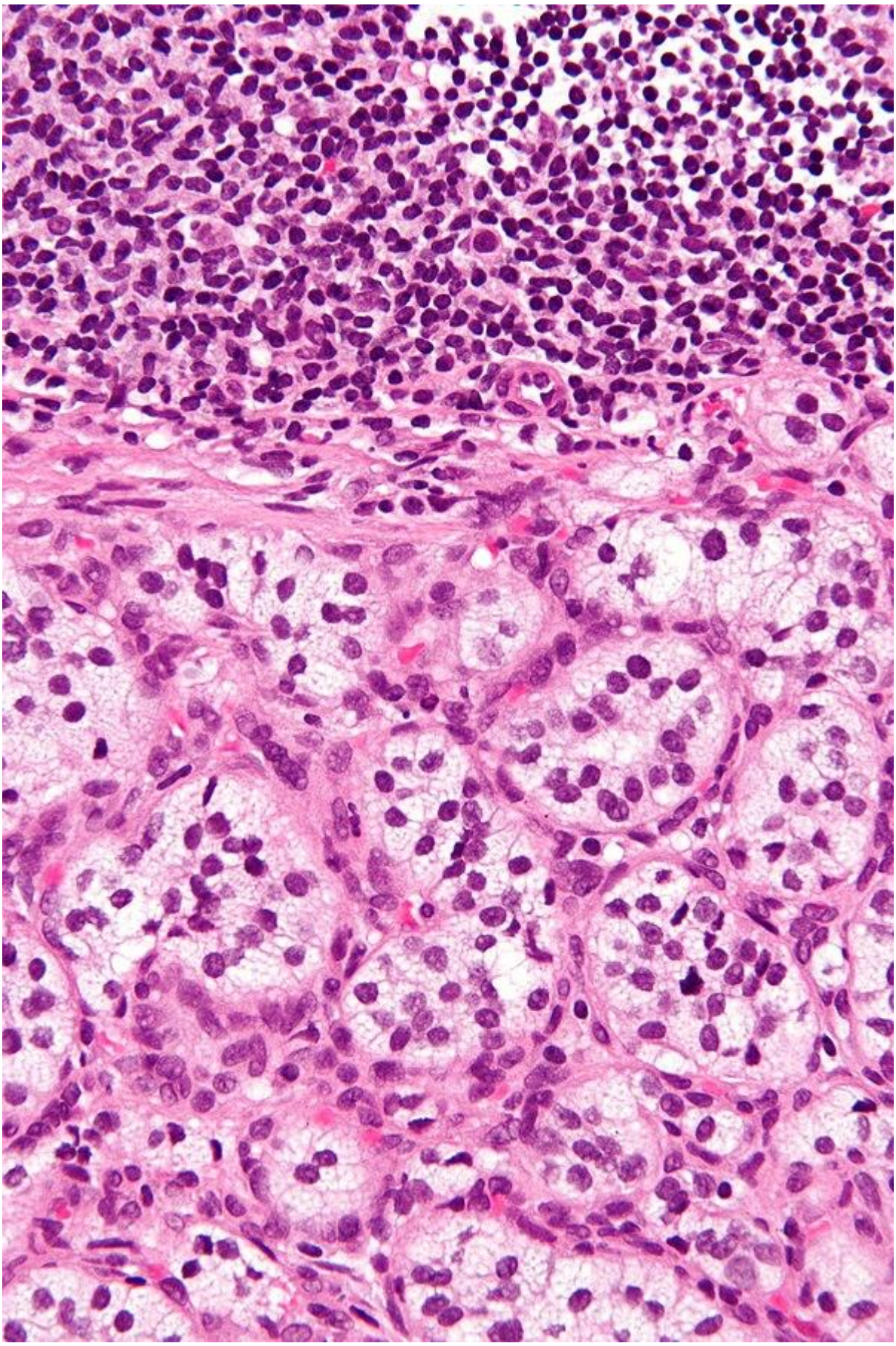
- Q: What is the most common benign tumour of the middle ear? ? Glomus tympanicum
- Q: What is the Brown sign? ? Blanching of pulsatile middle ear mass on pneumatic otoscopy
- Q: What is the salt-and-pepper sign on MRI? ? Flow voids within hypervascular paraganglioma
- Q: Name the histological hallmark of glomus tumour ? Zellballen pattern

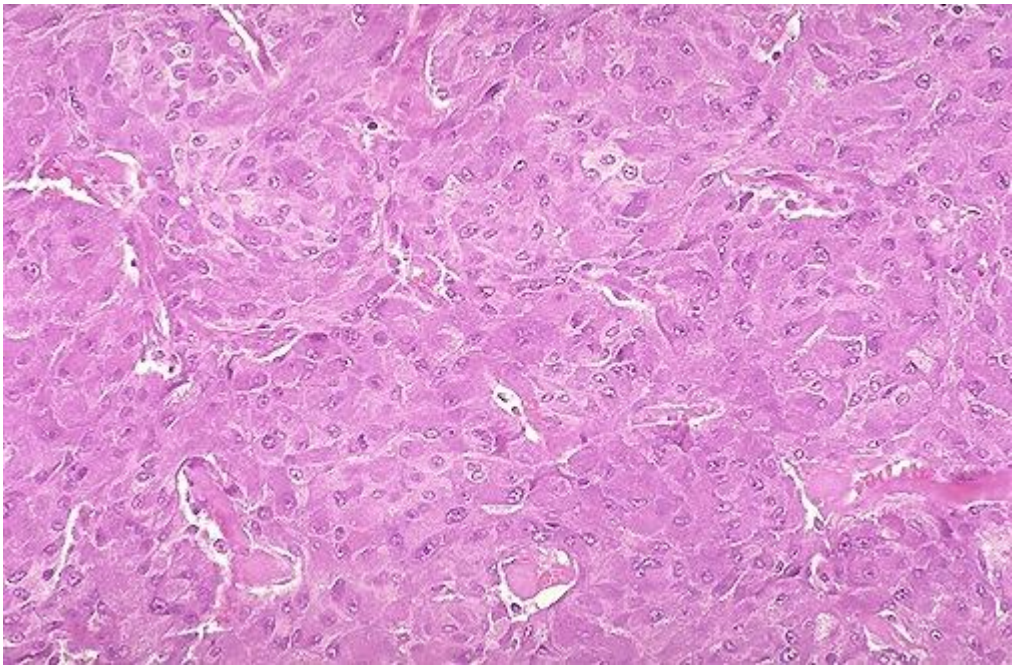
- Q: What is the surgical approach of choice for glomus jugulare? ? Fisch Infratemporal Fossa Approach Type A
- Q: Why should you not biopsy a pulsatile middle ear mass? ? Risk of catastrophic haemorrhage from aberrant ICA or glomus
- Q: What is the most common malignant middle ear tumour in children? ? Rhabdomyosarcoma
- Q: What gene mutation is associated with familial paraganglioma? ? SDHD (most common); SDHB (malignant risk)
- Q: What syndrome is associated with endolymphatic sac tumour? ? Von Hippel-Lindau (VHL) syndrome
- Q: What is the Vernet syndrome? ? CN IX, X, XI palsy due to jugular foramen lesion

IMPORTANT MICROBIOLOGY / HISTOPATHOLOGY SLIDES

Image 1

- Zellballen architecture
- Chief cells
- Sustentacular cells





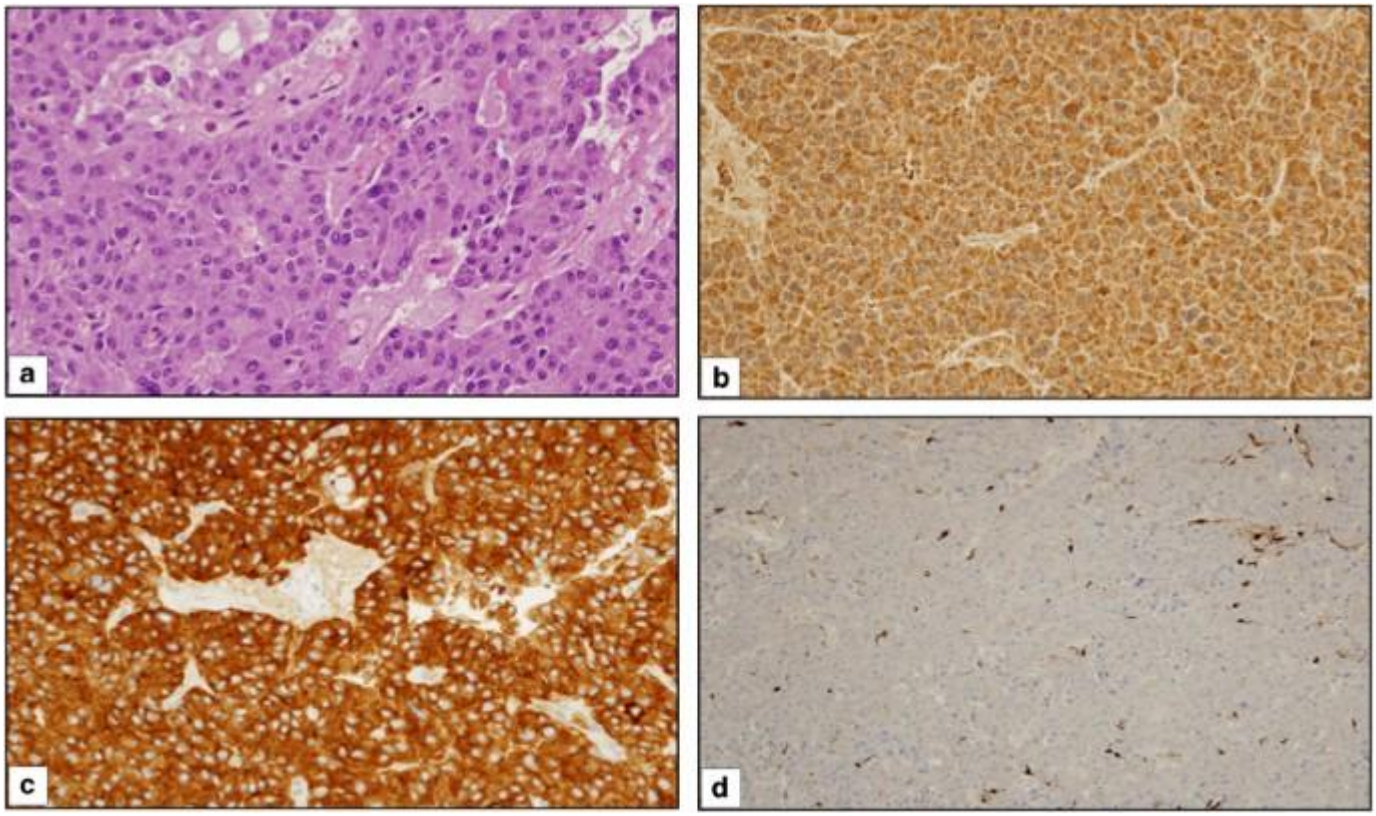
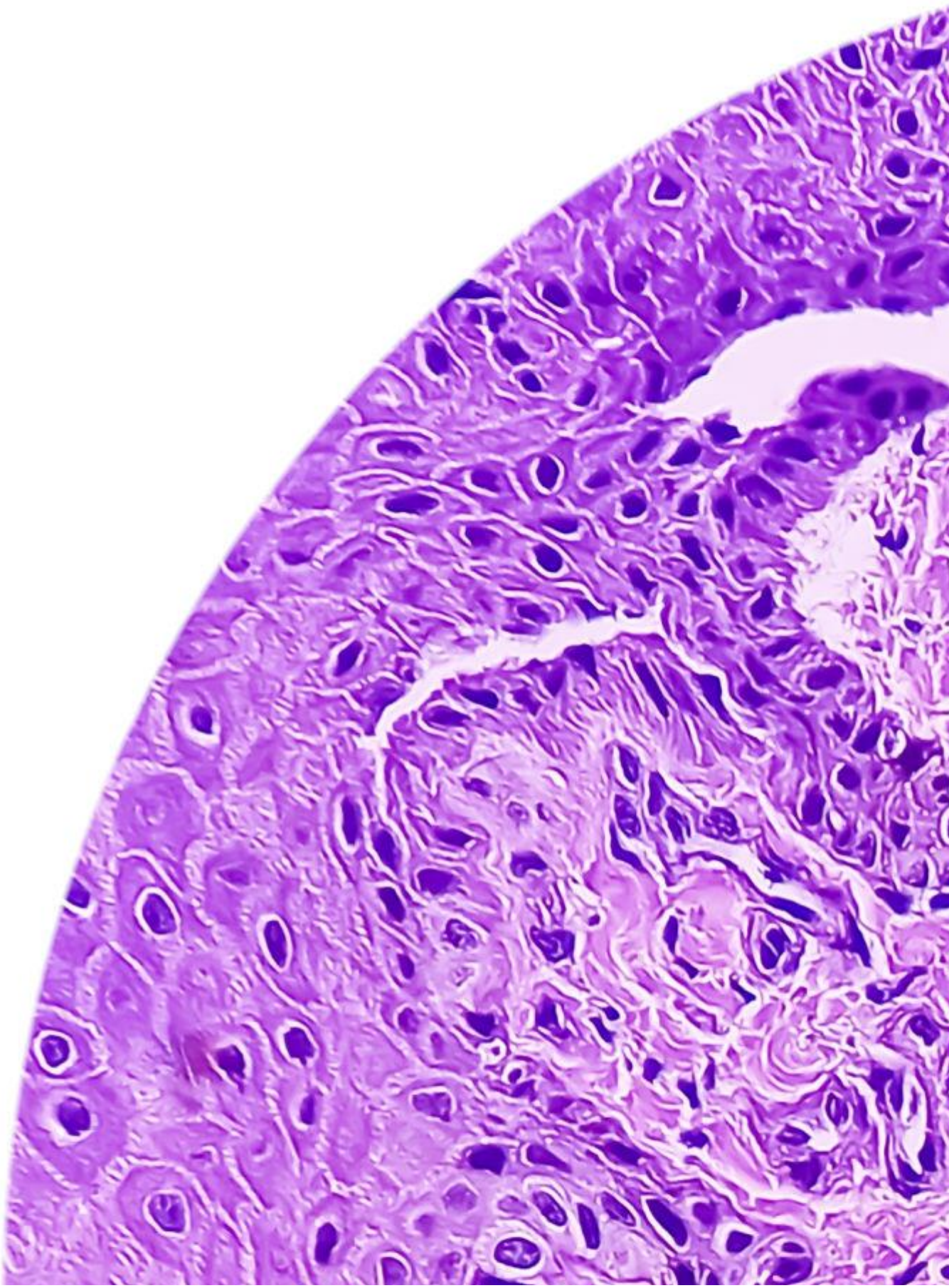
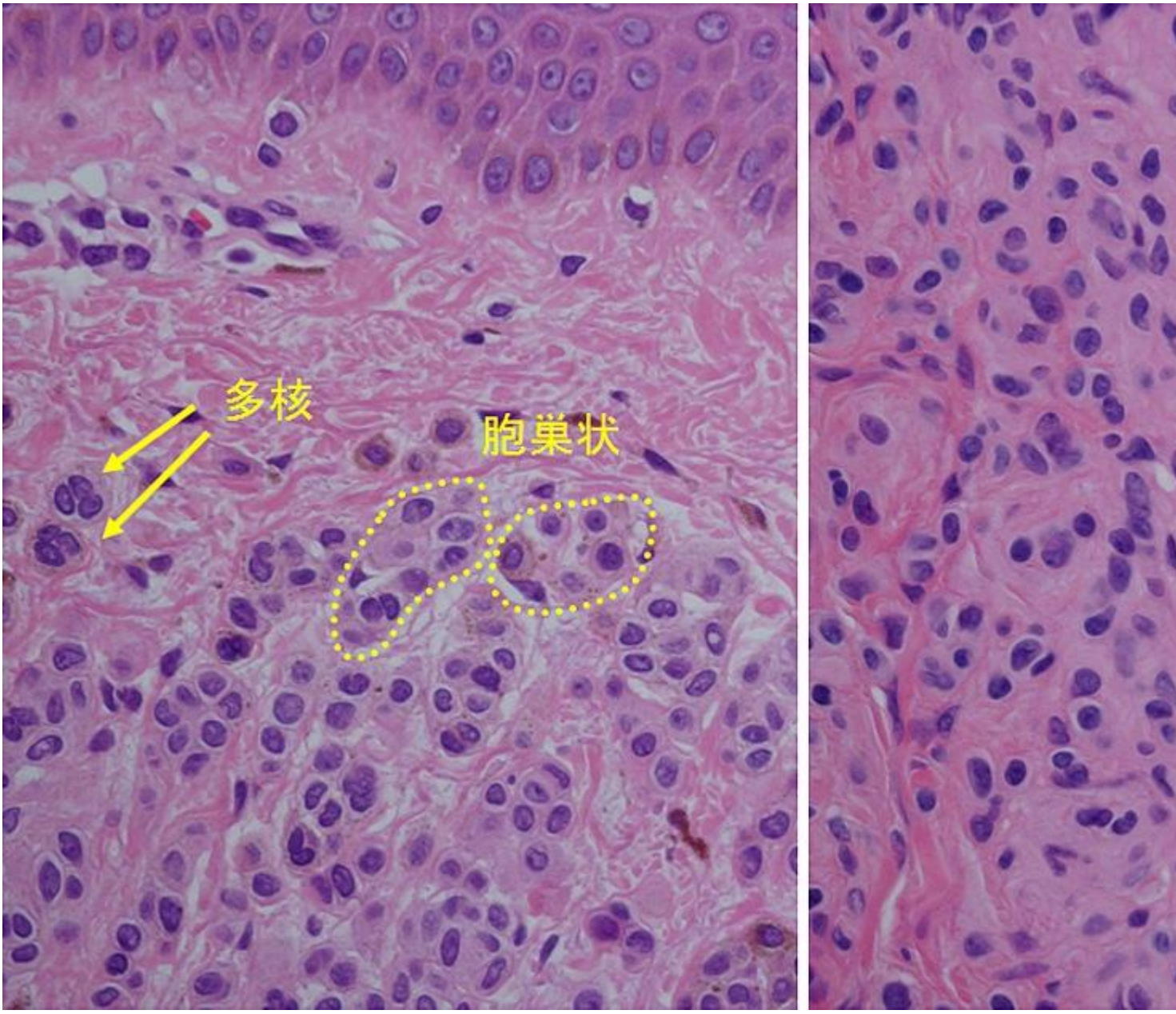


Image 2

- Hypervascular tumour histology

- SCC histology
- Adenocarcinoma




多核
胞巢状

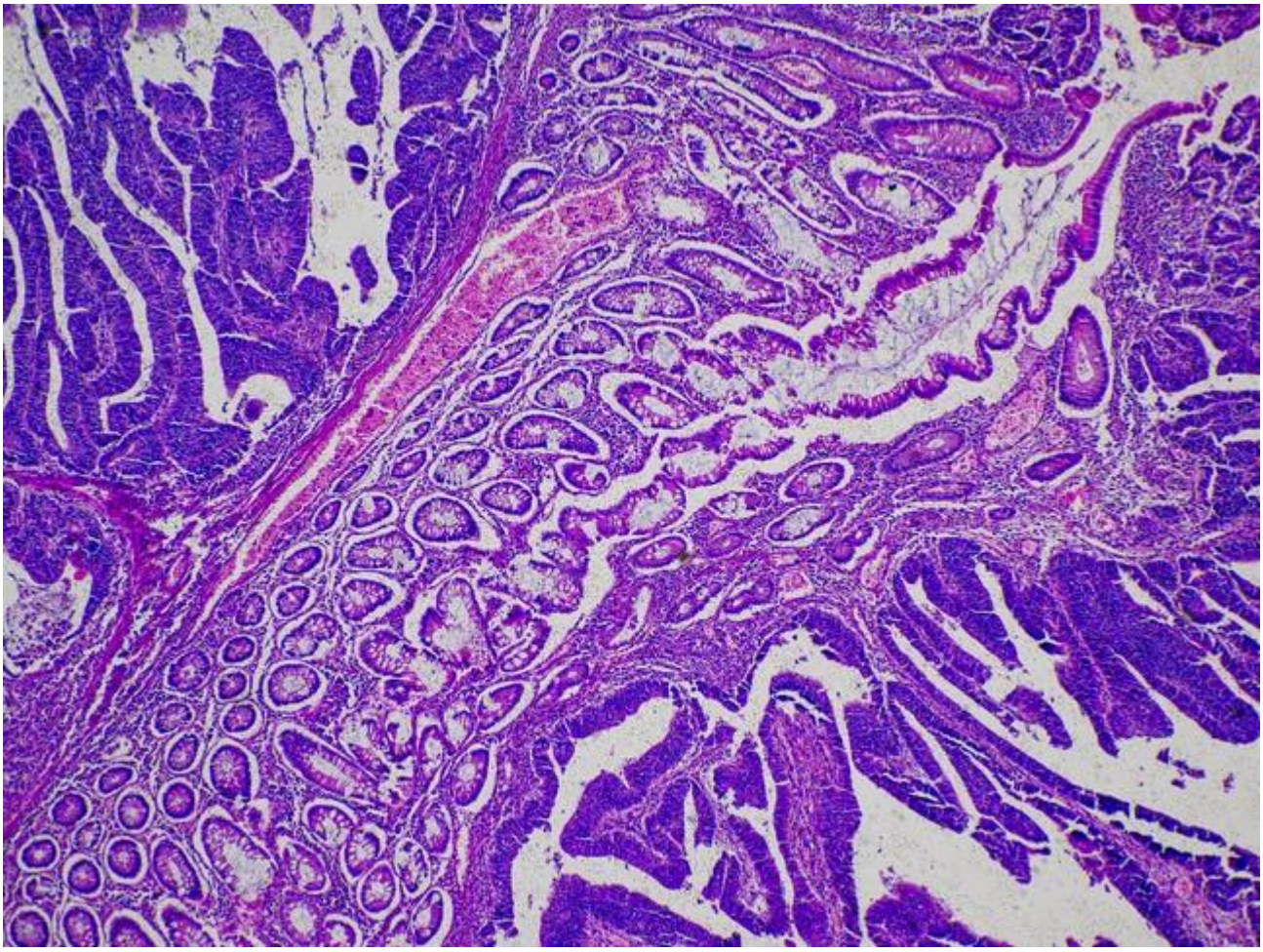
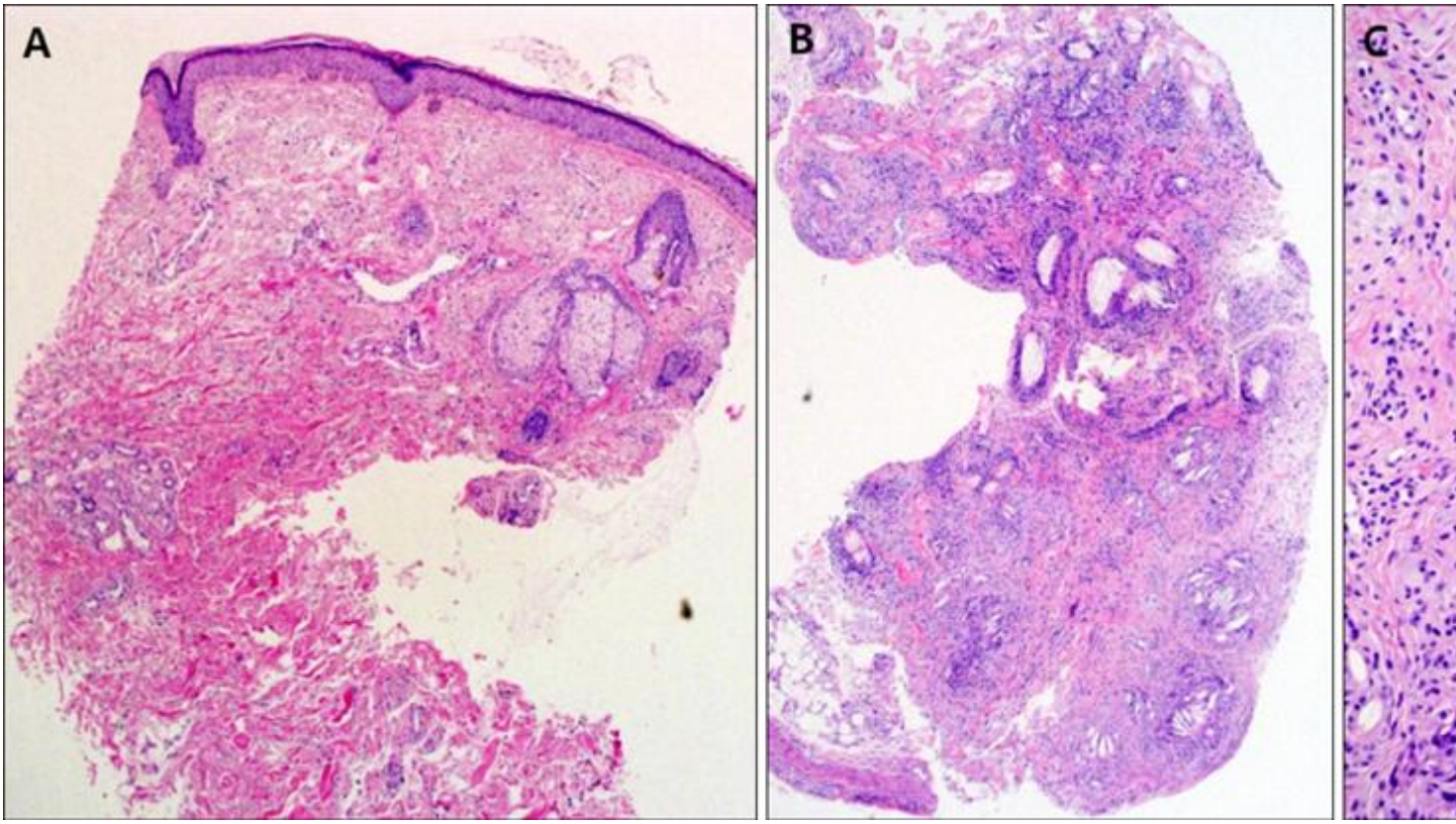


Image 3

- Rhabdomyosarcoma
- Cholesterol granuloma

DIAGNOSIS OF RHABDOMOYSARCOMA:

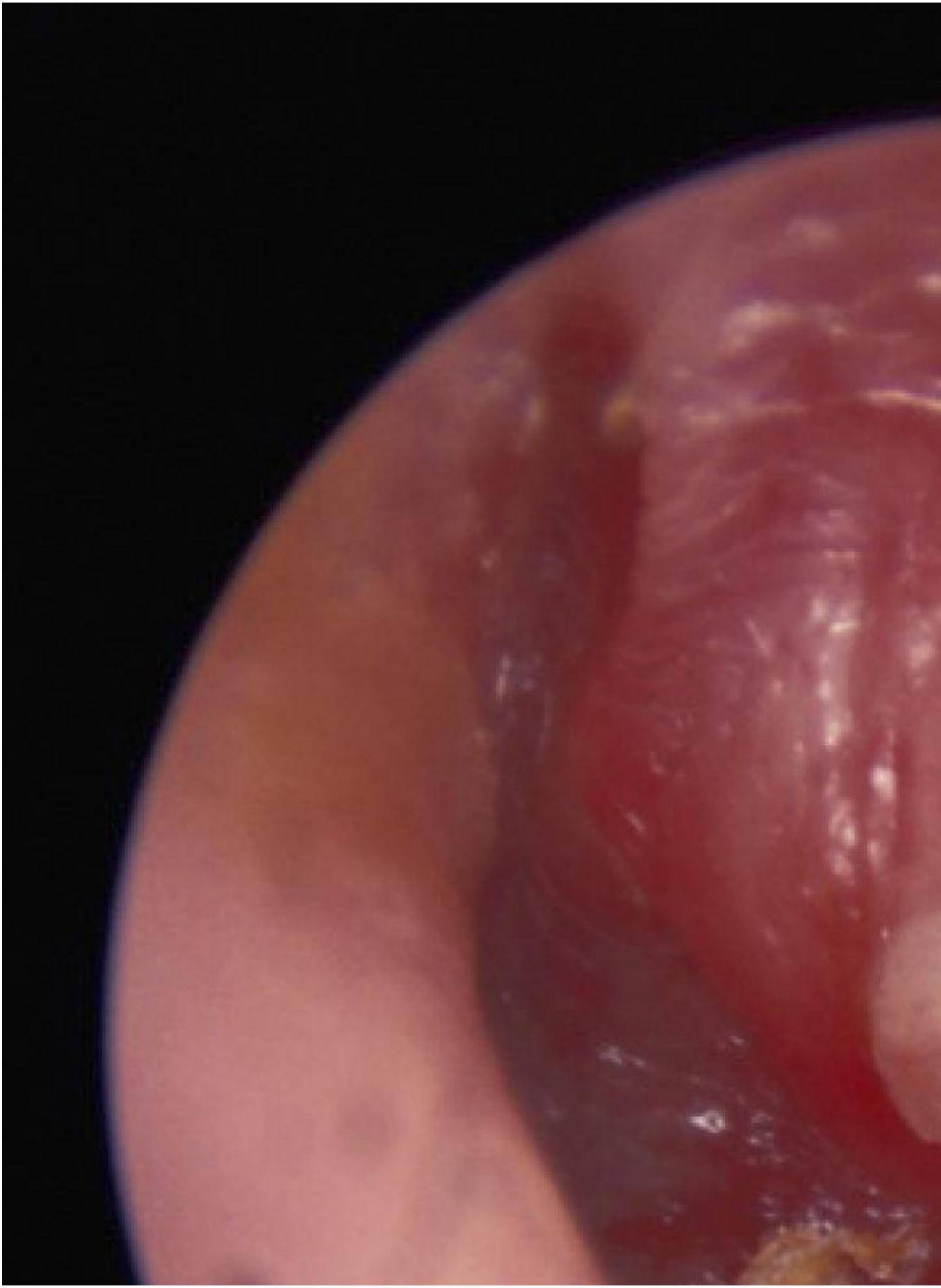
- Difficult to diagnosis due to its similarities to other cancers and varying levels of differentiation. It is loosely classified as one of the "small, round, blue-cell cancer of childhood" due to its appearance on an H&E stain
- The defining diagnostic trait for RMS is confirmation of malignant skeletal muscle differentiation with myogenesis (presenting as a plump, pink cytoplasm) under light microscopy.
- The alveolar type of RMS tends to have stronger muscle-specific protein staining
- Classification into types and subtypes is accomplished through further analysis of cellular morphology (alveolar spacings, presence of cambium layer, aneuploidy, etc.) as well as genetic sequencing of tumor cells.
- Radiologic evaluation should include plain radiographs of the primary site as well as a computed tomography (CT) scan of the primary and surrounding structures

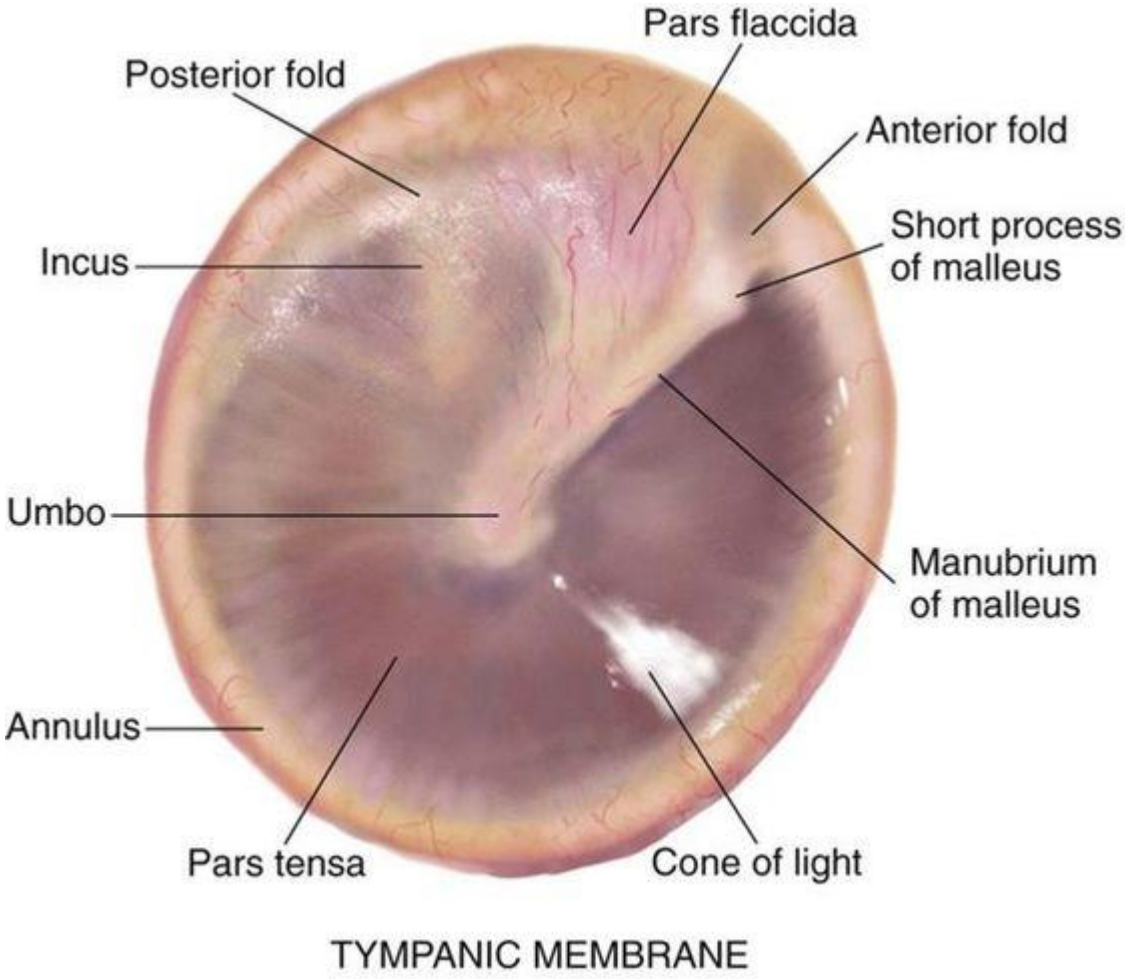


IMPORTANT CLINICAL PHOTOGRAPHS

Image 1

- Red pulsatile retrotympanic mass
- Brown sign
- Glomus tumour otoscopy

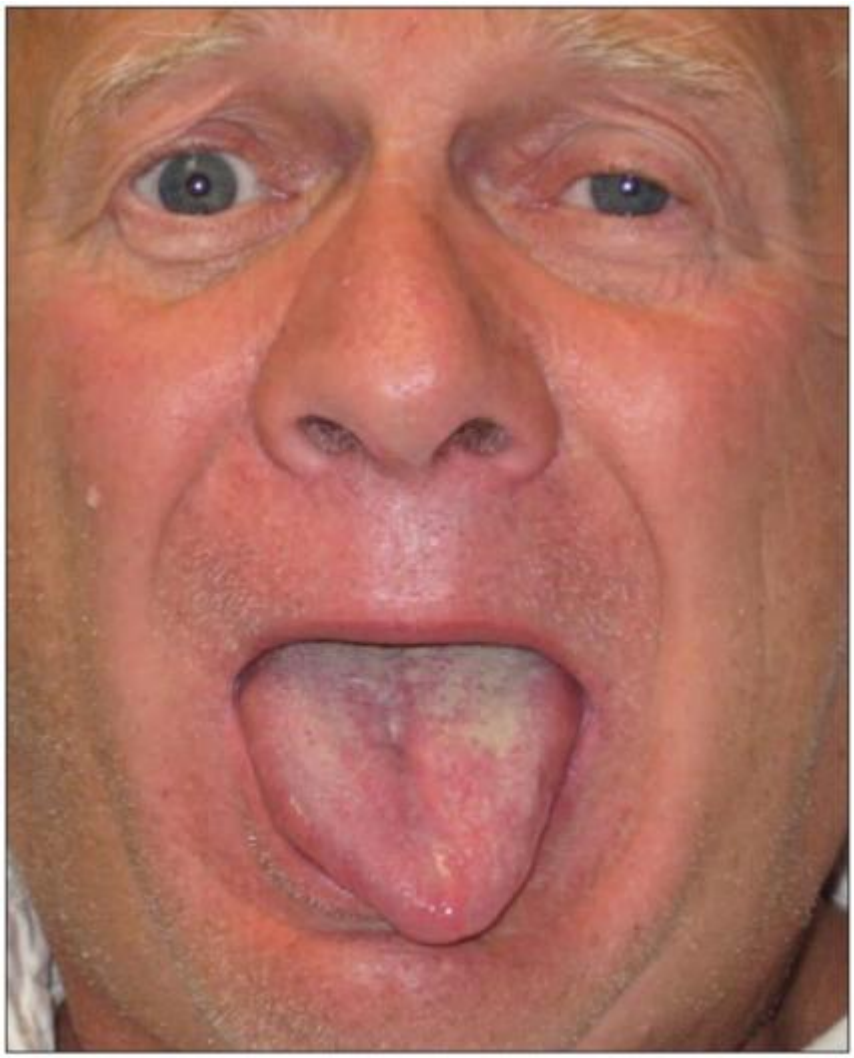


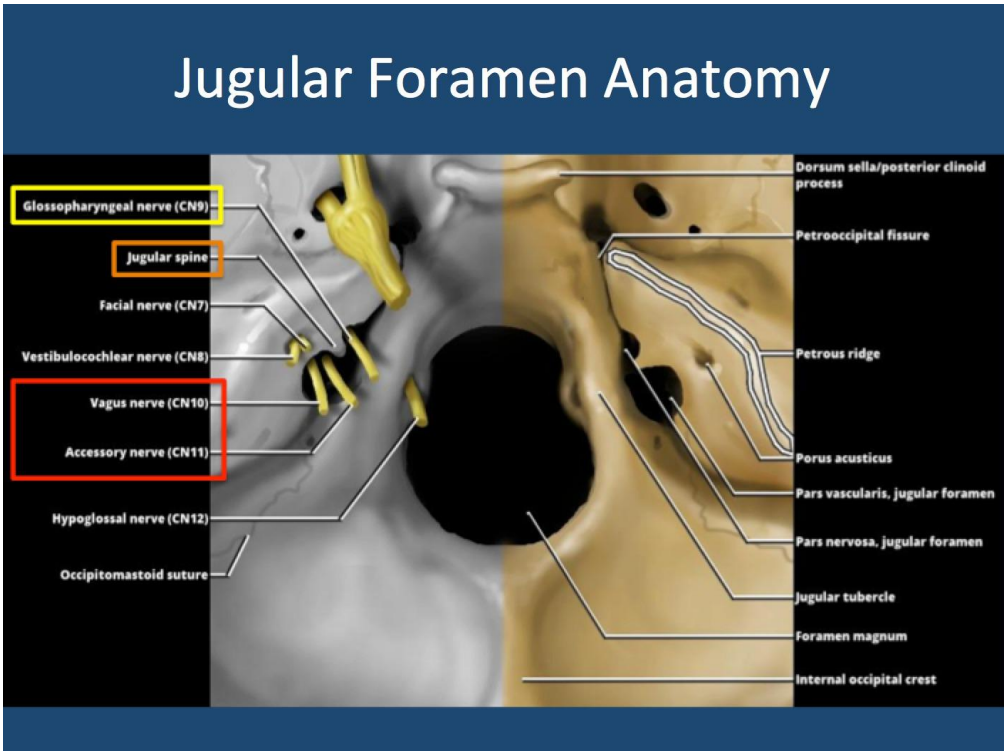


TYMPANIC MEMBRANE

Image 2

- Facial palsy
- Jugular foramen syndrome
- Angiography




Jugular Foramen Anatomy
Glossopharyngeal nerve (CN9)
Jugular spine
Facial nerve (CN7)
Vestibulocochlear nerve (CN8)
Vagus nerve (CN10)
Accessory nerve (CN11)
Hypoglossal nerve (CN12)
Occipitomastoid suture
Dorsum sella/posterior clinoid process
Petrooccipital fissure
Petrous ridge
Porus acusticus
Pars vascularis, jugular foramen
Pars nervosa, jugular foramen
Jugular tubercle
Foramen magnum
Internal occipital crest

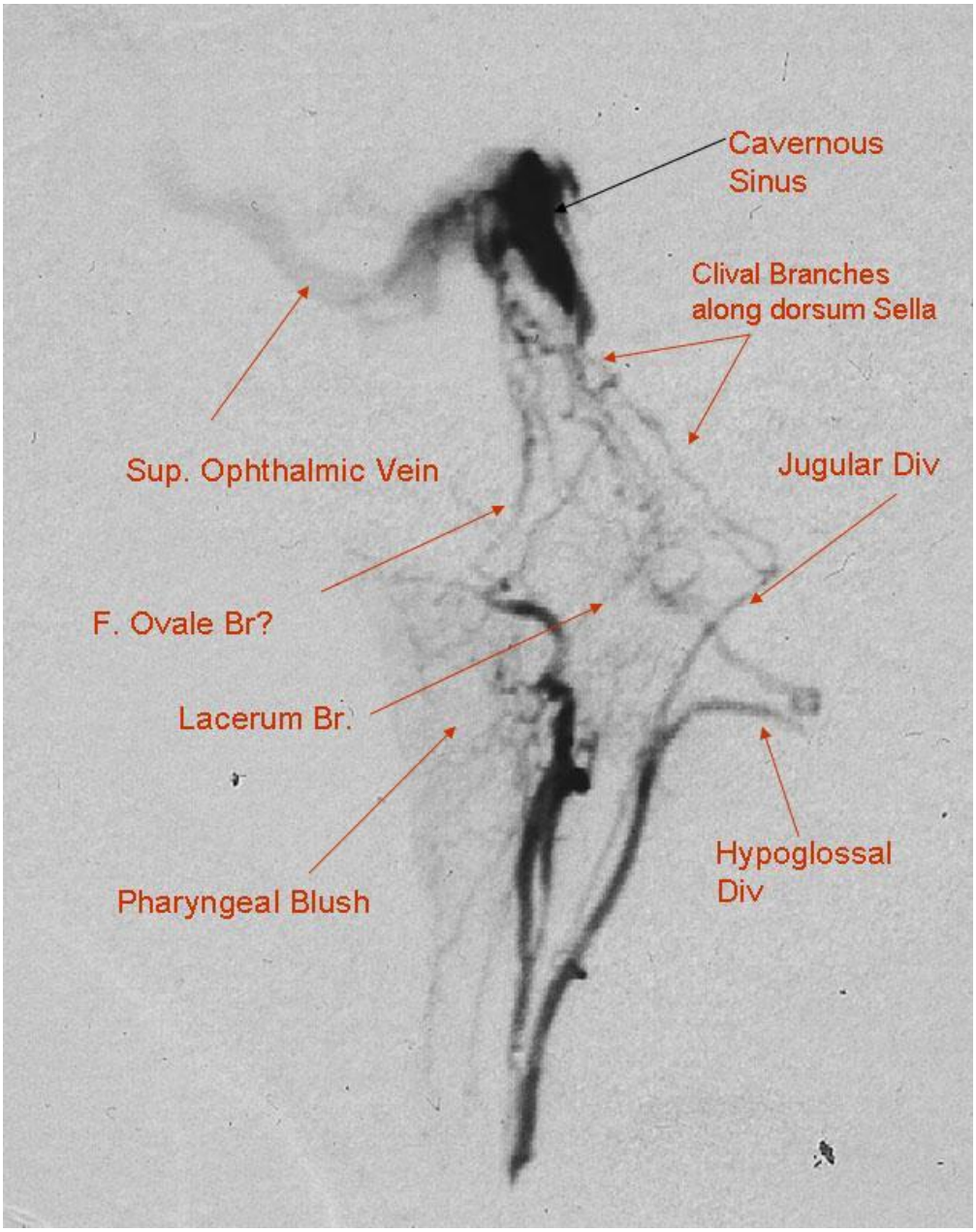


Image 3

- Embolization
- Postoperative mastoid cavity