Headmirror's ENT in a Nutshell
Jugular Paragangliomas (glomus jugulare)
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Presentation (0:30)

- Symptomatology
  - Pulsatile tinnitus
  - Conductive hearing losss
  - Lower cranial neuropathies (dysphonia, dysphagia, hypernasal speech) ~20%
  - Intermittent bloody otorrhea (late)
- Epidemiology
  - 40-60s (in contrast to familial paraganglioma which presents at younger age)
  - More common in women (6:1 ratio)
- Physical Examination
  - Red or violaceous color retrotympanic mass that can fill the inferior aspect of the tympanic membrane or extend to the entire middle ear space
  - **Rising sun sign** – recruit blood vessels of the ear canal that creates red hue of annulus with extension to the inferior aspect of EAC
  - **Brown's sign** – pneumatic otoscopy results in blanching of tumor
  - **Aquino's sign** – carotid compression decreases pulsations on otoscopy
  - Additional evaluation (cranial nerves)
    - Nasopharyngoscopy
    - Facial nerve assessment (not common)
    - High vagal injuries (palate incompetence with nasopharyngeal reflux, rhinolalia, ipsilateral vocal cord paralysis with lack of sensation)
    - Hypoglossal nerve weakness (hemiatrophy)
    - Accessory nerve involvement (not common)
  - Vernett syndrome – IX,X, XI deficits
  - Villaret's syndrome– IX, X, XI, XII, Horner's syndrome +/- facial nerve deficits
- Differential diagnosis
  - Facial nerve schwannoma
  - Endolymphatic sac tumor
  - Encephalocele
  - Middle ear adenoma
  - Inflammatory lesion (chronic otitis media with polyp)
  - High jugular bulb (purple hue – posterior and inferior)
  - Aberrant petrous carotid artery

Pathophysiology (8:35)

- Historic terminology
  - Chemodectomas (derived from chemoreceptors)→ Glomus tumors → Paragangliomas
- Paragangliomas
  - Jugular paraganglioma

    - Tympanic paraganglioma
    - Vagal paraganglioma
    - Carotid body tumor
- Derived from chief cells within paraganglia cells
    - Neuroendocrine in origin
    - Non-chromaffin staining cells, tend to cluster in nests (Zellballen)
- Functional (secreting tumors)
    - 2-5% of head and neck paragangliomas will secrete catecholamines
    - Uncontrolled hypertension, headache, diaphoresis, palpitations, arrythmia
- Risk of malignancy 1-3% (lower risk than paragangliomas outside of the head and neck)
- Diagnosis of malignancy is not based off histopathologic features
    - Have to find tumor in the lymph nodes to demonstrate metastasis

Workup (11:16)

- Imaging
    - High resolution thin sliced temporal bone CT scan
        - Jugular foramen involvement
            - Erosion of caroticojugular spine (bone that intervenes between carotid artery and jugular bulb)
            - Moth eating appearance of bone and poor margins
            - Can extend into petrous apex and intracranial extension
    - MRI
        - Demonstrate heterogenous salt and pepper appearance (prominent on T2)
        - Avidly enhance on T1, ill-defined borders or boundaries
    - Differential diagnosis on imaging
        - Meningioma – can involve jugular foramen or secondarily invade jugular foramen. Do not dilate jugular foramen. Have bone formation within tumor. Hyperostosis of surround base. Dural tails.
        - Schwannoma – dumb-bell appearance (wide within posterior fossa, constricted at jugular foramen and wide in the neck)
    - Conventional angiography
        - Prominent vascular blush
    - Abdomen and chest CT scan
        - Especially if multi-centric tumor or symptoms of excess catecholamines
        - Look for pheochromocytoma
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    - Dotatate-PET
        - >90% sensitivity and specificity
        - Whole body scan
- Laboratory Evaluation
    - Fractionated 24 urine metanephrines
        - 2-3 fold increase over baseline for this to be considered positive

        - Trending toward getting these on all jugular paraganglioma patients (even though secreting <5%)
    - Molecular genetic testing
        - Provide prognosis for disease and influence surveillance
        - Consider for malignant tumors, multi-centric tumors, secreting tumors, young male
        - MEN2A, MEN2B, von Hippel Lindau, NF1, familial paraganglioma syndrome
            - familial paraganglioma syndrome – autosomal dominant, succinate dehydrogenase mutation
                - type I-III benign disease course (non-secreting, non-malignant)
                - type IV more likely secreting and malignant

- Staging
    - **Glasscock-Jackson**
        - Type 1: small tumor involving jugular bulb, middle ear space and mastoid
        - Type 2: extends under internal auditory canal and may have intracranial extension
        - Type 3: erodes into petrous apex and may have intracranial extension
        - Type 4: extends beyond petrous apex to include clivus and infratemporal fossa and may have intracranial extension
    - **Fisch**
        - Type A: middle ear cleft (glomus tympanicum)
        - Type B: middle ear and mastoid
        - Type C1: carotid foramen
        - Type C2: vertical portion of the carotid
        - Type C3: horizontal portion of the carotid artery involving petrous apex
        - Type C4: more extension beyond petrous apex to involve paraclival area, foramen lacerum, and infratemporal fossa
        - Type D1: <2 cm intracranial extension
        - Type D2: >2 cm intracranial extension
        - Type D3: unresectable intracranial tumor

Treatment (21:12)

- Surgical resection
    - **Limited** resection (just middle ear component)
    - **Subtotal** resection (leave most critical portion in jugular foramen intact to prevent lower cranial neuropathy)
    - Gross **total** resection (historical gold standard but less common now)
        - More feasible with small tumor
        - 20-40% increase risk of developing lower cranial neuropathy
    - Other surgical considerations
        - Alpha blockade followed by beta blockade if catecholamine secreting tumor

        - Pre-operative embolization (ascending pharyngeal and occipital artery) with angiography almost always used to reduce intraoperative bleeding and need for transfusion
    - Infratemporal fossa type A approach: subtotal petrosectomy with ear canal closure + mobilization (anteriorly) of facial nerve
        - Newer techniques avoid or limit mobilization of facial nerve and closure of ear canal
- Radiosurgery
    - Gamma knife / Cyber knife
    - Risk of acquiring new or worsening cranial neuropathy with standard treatment dose is 1-10%
    - Tumor control after is >90% out to 10 years
    - Can be used primarily or adjuvant therapy or recurrence
    - Inferior extension is limited (down to C2)
- Observation
    - 30-40% of tumors will grow over 5 years
    - 20-30% chance of new or worsening lower cranial neuropathy
- Follow Up
    - Observation: Every 6 months, then yearly
    - Risk of recurrence is related to volume tumor left behind
        - Every year; more frequent if sub-total resection
        - Lifelong follow up needed
    - Radiosurgery: 9 months after treatment, then yearly
        - Lifelong follow up