Glomus Tumors

Synonyms

- Non chromaffin paraganglioma
- Chemodectoma

Definition

- Richly innervated, highly organized arterio-venous anastomosis forming tiny benign nodular focus in many parts of the body (promontory, jugular foramen, carotid body, nail beds, pads of fingers and toes, hands & feet)
- **Origin** – From widely distributed paraganglionic tissue which originates from neural crest
  - Glomus Tympanicum
    - Tympanic br of glossopharyngeal nerve (Jacobson nerve)
  - Glomus Jugulare
    - Jugular bulb

History

- 1941 → Guild first described "glomic tissue" in the dome of jugular bulb and on promontory
- 1945 → Rosenwasser reported "carotid body tumor"
- 1960 → Guilford and Alford coined the term glomus tympanicum
- 1978 → Fisch's classification
- 1981 → Glasscock and Jackson's classification → used currently

Epidemiology

- **Sex** → Male : Female  1:5 (Scott Brown)
- **Age** 50 - 60 yrs
- **Incidence**
  - Familial , although genetic model of inheritance is not agreed
  - No racial or ethnic predilection
  - 1 per 1.3 million (Moffat et al., laryngoscope, quoted in emedicine)
- **Malignancy** low (<5% of glomus tumors)
- **Catecholamine** secreting  1% - 3% of cases
Histology

- Gross
  - Encapsulated
  - Highly vascular
  - Locally invasive tumors
  - Main blood supply → ascending pharyngeal artery via inferior tympanic branch

- Microscopically
  - Granular eosinophilic cytoplasm
  - Small oval nucleus
  - Polymorphism & hyperchromatism frequent → not suggestive of malignancy
  - Abundance of thin walled sinuoids with no contractile muscular coat

Growth

- Middle ear
  - → Mastoid (VII N Palsy)
  - → TM (Bleeding Polyp)
  - → Inner Ear
    - VIII N Palsy
    - Labyrinth
    - Petrous Apex
  - → ET → Nasopharynx
  - → Jugular Foramen (IX, X, XI N Palsy)
  - → Intracranial → Middle & post. Cranial fossa

- Rarely metastatic spread to lungs & bones

Classification (FISCH)

- **Type A** - Tumors limited to middle ear cleft
- **Type B** - Tympanomastoid tumor no bone destruction in infralabyrinthine compartment
- **Type C** - Invading infralabyrinthine region and extending into petrous apex
  - **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
- **Type D1** - Intracranial extension <2cm in diameter
- **Type D2,D3** - Intracranial extension > 2cm in diameter

Classification (GLASSCOCK-JACKSON)

- **Glomus tympanicum**
  - **Type I** - Small mass limited to the promontory (Glomus Tympanicum)
  - **Type II** - Tumor completely filling middle ear space (Infralabyrinthine bone spared)
o **Type III** - Tumor filling middle ear and extending into mastoid process, petrous apex or infra labyrinthine bone destruction

o **Type IV** - Tumor filling EAC also &
  - A – Intracranial extradural extension
  - B – Intracranial intradural extension

- **Glomus jugulare**
  o **Type I** - Small tumor involving jugular bulb, middle ear, and mastoid process
  o **Type II** - Tumor extending under internal auditory canal; may have intracranial extension
  o **Type III** - Tumor extending into petrous apex; may have intracranial extension
  o **Type IV** - Tumor extending beyond petrous apex into clivus or infratemporal fossa; may have intracranial extension

**Clinical Features**

- **Pulsatile Tinnitus**
  o Swishing character synchronous with pulse
  o Clinically first symptom
  o 80 – 90 % cases → maybe the only complaint
  o 40 – 60 % → Disabling tinnitus which was previously neglected
  o Can be temporarily stopped with carotid pressure

- **Hearing loss**
  o Low frequency conductive hearing loss
  o Unilateral, slowly progressive
  o Gradually progresses to SNHL as inner ear invasion starts
  o 20 – 25 % present with mod mixed hearing loss at outset

- **Facial weakness**
  o Insidious onset, often incomplete
  o Develops when fallopian canal is invaded or tumor grows & compresses dehiscent facial nerve

- **Lower cranial nerve palsy**
  o Glomus jugulare → jugular foramen
    - IX, X
      - Dysphagia
      - Hoarseness ( U/l Vocal cord palsy)
      - U/l palsy soft palate, uvula
    - XI → Weakness of trapezius, SCM

- **Other symptoms**
  o Vertigo, chronic imbalance (invasion of otic capsule)
  o Headache (tumor mass effect)
  o Bleeding from EAC (polyp)
  o Otorrhea → when assoc with infection in middle ear cleft
  o Diplopia (petrous apex → 6th CN)
  o Nasal obstruction, epistaxis (ET → nasopharynx)
Examination
- Otoscopy
  - Retro tympanic reddish / bluish mass
  - Rising Sun – glomus jugulare → arising from pro tympanum above jugular bulb
  - Bulging TM may be present
  - Browne’s sign (Pulsation sign) – EAC pressure ↑ → Tumor pulsates vigorously → then blanches → reverse when pressure released
  - Aldering’s sign – pressure on ipsilateral carotid → blanching of tumor
  - Aural polyp might be present → bleeds on touch / spontaneously
- TFTs
  - Conductive hearing loss pre dominant
  - SNHL in advanced stage
- Other signs
  - Mastoid bruit
  - Lower cranial nerve palsies
  - Fistula sign +ve in semi-circular canal invasion
  - Some secrete catecholamine → Headache, flushing of face, HTN, anxiety
- Otomicroscopy – to confirm findings

Investigations
- PTA
  - Conductive hearing loss pre dominant
  - SNHL in advanced stage
- Baseline routine haemogram
- 24 hrs urinary catecholamine levels (VMA)
- Imaging (cornerstone of diagnosis)
  - HRCT temporal bone with contrast enhancement
    - Enhancing soft-tissue masses at the skull base
    - Erosion of the jugular foramen and petrous apex is often a key finding
    - → demonstrates
      - Site & type
      - Extent & normal landmarks
      - Pattern of spread
      - Phelp’s sign (erosion of jugulo carotico crest)
  - MRI
    - Intense tumor enhancement with possible intracranial / extra cranial extension
    - Salt-and-pepper fine vascular pattern on T2-weighted images
    - Coronal imaging → relationship to brainstem, skull base, and deep cervical soft-tissue structures
  - Angiography
    - Digital Subtraction Angiography can clearly delineate tumor
    - Feeding vessels → External carotid system
     → Internal carotid system
• Can be combine with pre-operative embolization within 48 hrs before surgery
  o MRA
    ▪ Non-invasive technique with equal efficiency
    ▪ Cross over vessels & morphology

• Biopsy is contraindicated
Algorithm for Diagnosis

Otoscopy findings → Suspicion of

Axial CT scan

Jugular fossa enlarged

Cortex eroded

Glomus jugulare

Cortex normal

High jugular bulb

Normal carotid canal, mass in middle ear

Glomus tympanicum

Normal jugular fossa

Coronal CT scan

Laterally placed carotid canal

Aberrant carotid artery
Management

Options → Wait & watch
→ Surgery
→ Radiotherapy
→ Gamma Knife Irradiation
→ Surgery + radiotherapy

• Wait, Watch & Follow up
  o For small tumors limited to promontory (Class 1)
  o No disabling tinnitus / hearing loss
  o Available for long follow up
  o Advanced age and poor surgical candidate
  o Denies surgery
  o Yearly CT scans to record progression

• Surgery
  o Pre op embolization
    ▪ Reduces intra op bleeding
    ▪ Some surgeons (Glasscock) → Risks outweigh benefits → do not advise
    ▪ Done 24 – 48 hrs prior to surgery
    ▪ Gel foam used routinely
    ▪ Absence of tumor blush confirms satisfactory embolization
    ▪ Usually combined with pre op angiography
  o Surgical options
    ▪ Fisch type A & Glasscock type 1 tumors
      • Trans meatal / peri meatal approach
    ▪ Fisch B,C1, C2 & Glasscock type 2 tumors
      • Trans Mastoid
      • Trans mastoid with extended facial recess approach with/ without re routing
    ▪ Fisch C3,D1 &Glasscock type 3 tumors
      • Transmastoid-infratemporal approach
      • Transtemoral-infratemporal approach
      • Fisch type A infratemoral fossa approach
    ▪ Fisch D2 & Glasscock type 4 tumors
      • Combined otologic and neurosurgical approach
      • Infratemoral approach with skull base resection and posterior fossa exploration
  o Complications
    ▪ Death
    ▪ Cranial nerve palsy
    ▪ Bleeding
    ▪ CSF leak
- Meningitis
- Uncontrollable hypotension/hypertension
- Tumor regrowth

**Radiotherapy**
- Proponents $\rightarrow$ surgical resection carries too high a price in terms of morbidity due mainly to iatrogenic cranial nerve deficits and CSF leaks
- Subnormal (28 – 32 Gy) in 20 # over 1 week
- Complications $\rightarrow$ osteoradionecrosis (ORN) of the temporal bone, brain necrosis, pituitary-hypothalamic insufficiency, and secondary malignancy, ICA thrombosis
- Additionally, Wilson et al $\rightarrow$surgery is complicated by previous radiation therapy

**Gamma-knife irradiation**
- Poor candidates for surgery
  - Age
  - Disease state
  - Unacceptable morbidity
- Expensive
- Clear remission is not reported
- Can control the tumor and prevent it from growing larger

**Surgery + radiotherapy**
- Large tumor (class 4) with multiple extensions, which cannot be resected completely

**Outcome and Prognosis**
- Grow slowly $\rightarrow$ cranial nerve palsies, long-term reduced quality of life
- Mortality rate
  - Overall mortality rate 8.7%
  - 2.5% among those treated surgically
  - 6.2% among patients treated with radiation
- Twenty years after treatment
  - Survival rate is 94%
  - 77% of patients remain symptom free

**Future and Controversies**
- Intraoperative guiding and imaging systems
- Definitive optimal treatment of type d glomus jugulare tumor is still controversial
- Increased use of gamma knife stereotactic radiosurgery
- Recent genetic research on familial glomus jugulare tumors $\rightarrow$ future $\rightarrow$ gene manipulation

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