Glomus Tumors of the Temporal Bone:
Synopsis of Glomus Tympanicum and Jugulare

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Grand Rounds Presentation
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Glomus Tumors of the Temporal Bone

Synopsis of Glomus Tympanicum and Jugulare

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Outline

- Paraganglia
- Paraganglioma
- Presentation
- Diagnostics
- Classification
- Treatment

(Swartz 2009)
Glomus bodies
- Clusters of chief cells
  - Identical to carotid body
  - Similar to adrenal autonomic ganglia
- “Zellballen” network with arterioles and venules

Chief cells
- Arise from neural crest cells, migrate with sympathetic ganglia (Gulya 1993)
- Neurosecretory modulators of vascular activity
  - Dopamine
  - Norepinephrine
Physiology

Temporal Bone Paraganglia

- Oxygen baroreceptors
- Cell clusters
  - Type I
    - Chief cells
    - Catecholamines
    - Dark cell type
    - Light cell type
  - Type II
    - Sustentacular cells
    - Modified Schwann cells

(Rao 1999)
Physiology
Temporal Bone Paraganglia

- **Third branchial arch** (Zak 1982)
  - Two or three present, maybe more in fifth decade
  - Nonchromaffin, lack chromium salt affinity
  - Distinction from adrenal neuroendocrine system

- **Location**
  - Two typical regions
    - Anterolateral jugular fossa
    - Within middle ear
  - Jugular bulb adventitia
  - Jacobson nerve (cranial nerve IX)
  - Arnold nerve (cranial nerve X)

(Swartz 2009)
Physiology
Temporal Bone Paraganglioma

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Extratemporal Paraganglioma
- Glomus jugulare
- Glomus tympanicum
- Carotid body tumor
- Glomus vagale
Temporal Paraganglioma

Epidemiology

- First described by Gould in 1941 (O’Leary 1991)
- Most common true neoplasm of middle ear
  - Most common pathologic condition of jugular foramen
  - Second most common tumor of the temporal bone
  - Rare overall, 0.012% in 600,000 (Bertrand 1976, Balatsouras 1992)
- More frequent in Caucasians
  - Usually on left side
  - Female:Male ratio 6:1
  - Peak incidence in fifth decade
- Aggressive in younger patients
  - Commonly multifocal
  - Likely to secrete vasoactive substances

(N Engl J Med 2010; 362:e66.)
Temporal Paraganglioma

Epidemiology

- Multiple tumors in 5-10%
- Familial autosomal dominant disorder
  - Less than 10% of cases
  - Multicentricity in 30-50% affected
  - Associated with defects on chromosome 11q23 (Petropoulous 2000)

- Metastasis in 3-4% (Pluta 1994, Motegi 2008)
  - Lymph nodes
  - Lung
  - Liver
  - Spleen
  - Bone
Temporal Paraganglioma

Characteristics

- Slow-growing, firm red mass
  - Bleeds profusely with manipulation
  - Neurological deficits in advanced cases
    - Dysphagia, dysarthria (hypoglossal canal)
    - Facial hypesthesia (petrous apex)
    - Ataxia, imbalance (posterior fossa, cerebellum)

- Chief cells
  - Clusters, “zellballen”
  - Nuclear pleomorphism and hyperchromatism
  - Store catecholamines
    - Actively secrete norepinephrine in 1-3%
    - More likely to be secreted by glomus jugulare

(Rao 1999)
Temporal Paraganglioma

Symptomatology

- Hearing loss (80%)
- Pulsatile tinnitus (60%)
- Aural fullness (18%)
- Rupture through tympanic membrane (7%)
  - Otalgia
  - Otorrhagia
- Vertigo/dizziness (9%)
- Headache (4%)
Temporal Paraganglioma

Symptomatology: Secreting Tumors

- Hearing loss (80%)
- Pulsatile tinnitus (60%)
- Aural fullness (18%)
- Rupture through tympanic membrane (7%)
- Vertigo/dizziness (9%)
- Headache (4%)

- Flushing
- Diarrhea
- Palpitations
- Headache
- Labile hypertension
- Orthostasis
- Diaphoresis
Temporal Paraganglioma

Physical Examination

- Reddish-blue mass on otoscopy
  - Medial to inferior tympanic membrane
  - Pulsatile
- Blanching with positive pressure (Brown sign)
- Decreased pulsations with carotid compression (Aquino sign)
- Audible bruit auscultated (objective tinnitus)
  - Mastoid
  - Infra-auricular region
Hearing loss
- Typically conductive (52%)
- Sensorineural if labyrinth invaded (5%)
- Mixed hearing loss (17%)

Neurological
- Facial nerve palsy
- Vernet syndrome
  - Jugular foramen syndrome
  - Cranial nerves IX, X, XI
- Villaret syndrome
  - Jugular foramen and Horner syndromes
  - Miosis, ptosis, anhydrosis
- Ataxia
Pulsatile Tinnitus

Differential Diagnosis

- Uncommon otologic symptom
- Vascular pathophysiology
  - Increased vascular blood flow
  - Vascular lumen stenosis
- Nonvascular
  - Sensorineural hearing loss
  - Superior semicircular canal dehiscence
  - Myoclonic contractions
    - Tensor veli palatini
    - Levator veli palatini
    - Salpingopharyngeus
    - Superior constrictor
Pulsatile Tinnitus

Arterial Etiologies

- Carotid atherosclerosis
- Intracranial vascular abnormalities
  - Dural arteriovenous fistula (AVF)
  - Arteriovenous malformation (AVM)
  - Aneurysm
    - Anterior inferior cerebellar artery
    - Internal carotid artery
    - Vertebral artery
- Tortuous internal carotid artery
- Fibromuscular dysplasia

(J Neurol Neurosurg Psychiatry 2004; 75:993)

(RadioGraphics 2004; 24:1637-1653)

(J NeuroIntervent Surg 2010; 2:202-207)
Pulsatile Tinnitus
Venous Etiologies

- Idiopathic intracranial hypertensive syndrome (IIH)
- Jugular bulb abnormalities
  - High jugular bulb
  - Jugular bulb dehiscence
  - Jugular diverticula
- Abnormal condylar and mastoid emissary veins
Pulsatile Tinnitus

Physical Examination

- Otoscopy
- Inspect oral cavity and pharynx for contractions
  - Wide oral opening may decrease contractions
  - Fiberoptic nasopharyngoscopy
- Venous pulsatile tinnitus
  - Decreases with internal jugular vein compression
  - Decreases with head rotation ipsilaterally
- Neurological
Pulsatile Tinnitus Diagnostics

- Audiological
- Carotid Duplex ultrasound
- Computed tomography angiography (CTA)
- Magnetic resonance venography
- Carotid angiography if suspect AVF/AVM
- Lumbar puncture if suspect IIH
- CTA temporal bone and neck if abnormal otoscopy

(Sismanis 2011)
Temporal Paraganglioma Diagnostics

- Computed tomography (CT)
- Magnetic resonance imaging (MRI)
- Angiography
- Serum catecholamines
- Urinary vanillylmandelic acid
- Urinary metanephrine
- Positron emission tomography (Hoegerle 2003)
- Octreotide scintigraphy (Bustillo 2004)
Diagnostics

CT

- Bony partition between jugular fossa and hypotympanum
  - Glomus jugulare erodes jugular fossa
  - Glomus tympanicum occupies middle ear

- Jugular foramen
  - Enlargement if length + width greater than 20mm
  - Bony erosion with decalcification

(Rao 1999)
(Swartz 2009)
(http://me.hawkelibrary.com)
Diagnostics

CT

- Caroticojugular spine
  - Separates jugular bulb from petrous carotid artery
  - Erosion with glomus jugulare
- Intracranial extension
- Invasion of fallopian canal
- Other vascular abnormalities
  - High jugular bulb
  - Aberrant carotid artery

(Rao 1999)
(Imaging 2007; 19:55-70)
(Radiology Case Reports 2009; 4(4))
(http://me.hawkelibrary.com)
Diagnostics
CT Differential: Schwannoma

- Smooth erosion of jugular foramen
- Glomus erosion typically irregular margins
Diagnostics
CT Differential: Meningioma

- May be difficult to distinguish from schwannomas
- Tend to infiltrate bone around jugular foramen

Diagnostics

MRI

- Vascular
  - “Salt and pepper” flow voids
  - Intraluminal involvement of petrous carotid artery
  - Occlusion of jugular vein and sigmoid sinus
- Intracranial extension
  - Intradural
  - Extradural
- Screen for multiple tumors
- Assess for glomus vagale tumor

(http://sumerdoc.blogspot.com)
Diagnostics

MRI Differential: Schwannoma

- Smooth contoured mass
- T1-weighted images
  - Iso-intense without contrast
  - Significant gadolinium enhancement
- High signal intensity on T2-weighted images

[Link to Radiopaedia.org](http://radiopaedia.org)
Diagnostics
MRI Differential: Meningioma

- T1- and T2-weighted imaging
  - Iso-intense to grey matter
  - Increased intensity with contrast
- “Dural tails”

(Iran J Radiol 2011; 8:176-81)  (Surgical Neurology 2001; 56:8-20)
Diagnostics

**CT versus MRI**

- **CT usually sufficient imaging alone**
  - Jugular bulb major preoperative consideration
  - Supplants venography to assess jugular bulb

- **MRI when diagnosis or extent is questioned**
  - Delineates neoplastic and native tissue
  - Good for intradural tumors
  - T1 images may overestimate tumor extent \(\text{(Brackmann 2010)}\)

- **Petrosus apex best imaged with CT and MRI** \(\text{(Arriaga 1991)}\)

- **Magnetic resonance angiography typically inadequate**
Diagnostics

Angiography

- Better assess larger tumors
- Identify multicentric disease
- Preoperative embolization
  - Performed 1-2 days before surgery
  - Polyvinyl alcohol or intravascular coils
  - Decrease intraoperative blood loss
- Balloon occlusion if anticipate carotid sacrifice
- Carotid sacrifice morbidity (Linskey 1994)
  - Cerebral infarction 26%
  - Fatality 46%

(http://www.aneurysm-stroke.com)
Better assess larger tumors

Identify multicentric disease

Preoperative embolization
  - Performed 1-2 days before surgery
  - Polyvinyl alcohol or intravascular coils
  - Decrease intraoperative blood loss
  - Balloon occlusion if anticipate carotid sacrifice
  - Carotid sacrifice morbidity: 26% cerebral infarction, 46% fatality (Linskey 1994)

Before Embolization

After Embolization
Primary supply
- Inferior tympanic artery (from ascending pharyngeal artery)
- Stylomastoid artery
  - Occipital artery (60%)
  - Posterior auricular artery (40%)

Other contributors (Hesselink 1981)
- Middle meningeal artery
- Internal carotid artery
- External carotid artery

Contributors
- Internal maxillary
- Middle meningeal
- Posterior auricular

(AJNR 2006; 27:1820-1822)
Vascular compartments
(Moret 1980, Moret 2982, Russel 1986, Young 1988)
- Inferomedial
- Posterolateral
- Anterior
- Superior

Each compartment hemodynamically independent

Multicompartamental in 85% glomus tumors

Contributors
- Occipital
- Posterior auricular
## Angiography

### Arterial Supply

<table>
<thead>
<tr>
<th>Compartment</th>
<th>Location</th>
<th>Blood Supply</th>
</tr>
</thead>
<tbody>
<tr>
<td>Inferomedial</td>
<td>Jugular bulb, Hypotympanum</td>
<td>Inferior tympanic branch of ascending pharyngeal</td>
</tr>
<tr>
<td>Posterolateral</td>
<td>Posterior tympanic cavity, Mastoid</td>
<td>Stylomastoid</td>
</tr>
<tr>
<td>Anterior</td>
<td>Protympanum, Pericarotid area</td>
<td>Anterior tympanic branch of internal maxillary, Caroticotympanic branch</td>
</tr>
<tr>
<td></td>
<td></td>
<td>of internal carotid</td>
</tr>
<tr>
<td>Superior</td>
<td>Epitympanum, Supralabyrinthine</td>
<td>Superior tympanic branch of middle meningeal artery</td>
</tr>
</tbody>
</table>

(Alerstone 1996)
Glomus Tumors

Pattern of Spread

- “Danger zone” around jugular bulb (Minor 1994)
  - Inferior petrosal sinus
  - Internal jugular vein
  - Sigmoid sinus
- Protympanum
- Hypotympanum
- Mesotympanum
- Intracranial extension
  - Radiographic diagnosis 14-20% (Rigby 1996)
  - Up to 50% with dural involvement intraoperatively (Spector 1976, Andrews 1989, Jackson 1990)
Patterns of Spread

Protympanum

- Peritubal cell tract
  - Petrous apex

- Carotid canal
  - Middle cranial fossa
  - Most common route for intracranial extension via protympanum

- Eustachian tube
  - Nasopharyngeal mass
  - Epistaxis

(Minor 1994)
Patterns of Spread
Hypotympanum

Sigmoid sinus lumen

Inferior petrosal sinus

Internal jugular lumen

Neural foramina at skull base

Facial Nerve
Jugular Vein
Internal Carotid

(Minor 1994)
Patterns of Spread
Mesotympanum

- Antrum and epitympanum
- Facial recess
- Sinus tympani
- Mastoid
- Round window
  - Erode cochlea
  - Internal auditory canal
- Laterally
  - External auditory canal

(Minor 1994)
## Classification
### Glasscock-Jackson

### GLOMUS TYMPANICUM

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<tr>
<td>I</td>
<td>Limited to the promontory</td>
</tr>
<tr>
<td>II</td>
<td>Completely fills the middle ear space</td>
</tr>
<tr>
<td>III</td>
<td>Fills the middle ear and extends to mastoid</td>
</tr>
<tr>
<td>IV</td>
<td>Extend into external auditory canal. May extend anterior to internal carotid artery</td>
</tr>
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**Glasscock-Jackson**

### GLOMUS JUGULARE

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<tr>
<td>I</td>
<td>Involves jugular bulb, middle ear, and mastoid</td>
</tr>
</tbody>
</table>
| II   | Extends underneath internal auditory canal  
      | May have intracranial extension |
| III  | Extends into petrous apex  
      | May have intracranial extension |
| IV   | Extends into clivus and infratemporal fossa  
      | May have intracranial extension |
GLOMUS TUMORS

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| B    | Limited to tympanomastoid complex  
  No infralabyrinthine involvement |
| C    | Involves labyrinthine compartment  
  Extends to petrous apex |
| D    | Intracranial involvement |
# Classification

**Fisch**

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(Fisch 1988)
## Classification

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(Fisch 1988)
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<td>Limited involvement of vertical portion of carotid canal</td>
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<tr>
<td>C2</td>
<td>Invades vertical portion of carotid canal</td>
</tr>
<tr>
<td>C3</td>
<td>Invades horizontal portion of carotid canal</td>
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<tr>
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- **C**: Involves labyrinthine compartment and extends to petrous apex
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(Fisch 1988)
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**Classification**

- **Fisch**

- **Type A**
  - Limited to middle ear cleft

- **Type D**
  - Intracranial involvement

- **Subtypes**
  - **D1**: Intracranial extension less than 2cm
  - **D2**: Intracranial extension greater than 2cm
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(Fisch 1988)
Temporal Paraganglioma

Treatment

- **Excision**
  - Definitive treatment in 90%
  - Staged for larger tumors
  - Laser or bipolar cautery for excision and hemostasis

- **Alpha- and beta- blockade secreting tumors**

- **Radiation**
  - Adjuvant therapy if incomplete resection
  - Poor or unwilling surgical candidates
Treatment

Surgery

- Transcanal approach
  - Small tumors on promontory
  - Drill canal for hypotympanum access
- Facial recess approach for middle ear and mastoid
- Transmastoid-transcervical for glomus jugulare
- Modified infratemporal approach for larger tumors
- Unresectability (Jackson 1982, Rufini 2006)
  - Foramen magnum
  - Cavernous sinus
Treatment

Transmastoid-Transcervical

Complete mastoidectomy
Open facial recess
Identify vasculature and nerves

(Minor 1994)
Treatment

Transmastoid-Transcervical

- Amputate mastoid tip and ligate internal jugular vein
- Pack sigmoid sinus and transpose facial nerve

(Minor 1994)
Treatment
Fisch Surgical Approach

- Postauricular approach to infratemporal fossa
- Type A
- Type B
- Type C

(Snow 2009)
Treatment

Fisch Surgical Approach

- Postauricular approach to infratemporal fossa
- Type A
- Type B
  - Radical mastoidectomy
  - Anterior transposition of facial nerve
  - Explore posterior infratemporal fossa
  - Cervical dissection
    - Jugular bulb
    - Vertical petrous carotid
    - Posterior infratemporal fossa
Treatment

Fisch Surgical Approach

- Postauricular approach to infratemporal fossa
  - Type A
  - Type B
    - Explore petrous apex
    - Mid-clivus
    - Horizontal internal carotid artery
    - Superior infratemporal fossa
  - Type C
Treatment

Fisch Surgical Approach

- Postauricular approach to infratemporal fossa
- Type A
- Type B
  - Nasopharynx
  - Peritubal space
  - Rostral clivus
  - Parasellar area
  - Cavernous sinus
  - Pterygopalatine fossa
  - Anterosuperior infratemporal fossa
  - Foramen rotundum
Treatment

Modified Infratemporal Approach

Similar postauricular incision

External auditory canal (EAC) is transected and oversewn

- EAC, tympanic membrane, middle ear contents resected
- Facial nerve mobilized

(Canalis 2000)
(Glasscock 2003)
Treatment

Modified Infratemporal Approach

- Resect eustachian tube
- Expose internal carotid artery
- Access
  - Middle and posterior cranial fossa
  - Nasopharynx
  - Foramen rotundum
  - Clivus
  - Cavernous sinus

Anterosuperior extension
- Resect zygoma and temporomandibular joint
- Reflect temporalis inferiorly
- Dislocate mandibular anteroinferiorly

(Canalis 2000)
(Glasscock 2003)
Treatment

Transcondylar/Suboccipital

- Intracranial tumors near foramen magnum
- Supplement intratemporal approach
  - Trans-sigmoid exposure
  - Trans-labyrinthine exposure
- Exposures cranial cervical junction

(Brackmann 2010)
Treatment

Transcondylar/Suboccipital

- Identify vertebral artery
  - Posterior and inferior to mastoid tip
  - On transverse process of C1 vertebra
- Expose occipital condyle and jugular tubercle
  - Access hypoglossal nerve
  - Access cranial cervical junction
- Cervical stabilization procedure if more than half of condyle resected

(Brackmann 2010)
Treatment

Surgical Considerations

- Leave tumor portion if adherent to internal carotid
- Cranial nerve preservation enhanced if medial wall of jugular bulb preserved
- Intradural involvement
  - Commonly at jugular bulb
  - En-bloc resection if blood loss less than 2000mL
  - Otherwise staged procedures
Treatment
Surgical Complications

- Bleeding
- Facial paresis/paralysis
- Cranial nerve palsy
  - Hoarseness
  - Dysphagia
  - Dysarthria
- Cerebrospinal fluid leak
- Tympanic membrane perforation (O’Leary 1989, Forest 2001)
- Cholesteatoma (O’Leary 1989, Forest 2001)
Treatment
Radiotherapy

- External beam
- Stereotactic
- Not ablative
  - Obliterative endarteritis in tumor vessels stops growth
  - Tumor control in over 90% (Maarouf 2003, Krych 2006)
  - Regrowth possible after 10-15 years (Brackmann 2010)
- Comparable to surgery
  - Tumor control
  - Recurrence
  - Morbidity
Treatment
Radiotherapy

- Indications
  - Elderly
  - Poor surgical candidates
  - Some multicentric lesions
  - Patient preference

- Dual-modality treatment with surgery
  - Limit cranial neuropathies
  - Adjuvant therapy after near-total resection

- No long-term data
Treatment

Radiotherapy

- Surveillance (Swartz 2009)
  - Residual mass
  - Stabile or decreased size
  - Decreased enhancement on CT
  - Reduced T2-weighted signal on MRI
  - Diminished flow voids

- Morbidity
  - Cranial nerve deficit
  - Osteoradionecrosis
  - Tumor regrowth
  - Radiation-induced malignancy (Lustig 1997)
Conclusion
Glomus Tumors of Temporal Bone

- Most common tumor of middle ear but rare
- Pulsatile tinnitus and hearing loss classic symptomatology
- Characteristic radiographical features
- Surgery most definitive treatment
  - Transcanal for small tumors
  - Transmastoid-transcervical for larger ones
- Radiation acceptable alternative for nonsurgical candidates
References


References


