Vestibular Schwannomas: History, Diagnosis, and Management

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OUTLINE

• History of Vestibular Schwannomas
• Relevant Anatomy Review
• Epidemiology and Tumor Biology
• Signs and Symptoms
• Diagnostic Approaches
• Differential of Cerebellopontine Angle Masses
• Management Options and Considerations
Acoustic Neuroma or Vestibular Schwannoma

- They are not technically neuromas
- Arise from the vestibular components of the nerve
Overview of Cerebellopontine Angle Masses

• 10% of intracranial tumors are found in the Cerebellopontine Angle (CPA)
• Roughly 80% are Vestibular Schwannomas (VS)
• Differential of CPA masses
  • Meningiomas
  • Epidermoids
  • Schwannomas of other nerves
  • Dermoid Tumors
  • Arachnoid Cysts
  • Lipomas
  • Metastatic Tumors
  • Vascular Tumors
Historical Perspective

• Acoustic Neuromas were first described in the late 1700s
• Sandifort
  • 1777 during an autopsy in a deaf patient “documented a small body adherent to the right auditory nerve”
• In the mid 1800s an understanding that unilateral deafness, facial numbness, and progressive blindness from “optic neuritis” (papilledema) likely due to a CPA mass
Historical Perspective

- **Sir Charles Ballance**
  - 1894 first removal of VS, but patient died from complications
  - 1907 first successful excision without patient mortality
  - Overall high surgical mortality
- **Horsley, von Eiselsberg, and Krause**
  - Largely performed unilateral suboccipital craniotomy with removal by finger dissection
  - 1913 Presented data on 63 patients, 78% mortality
- **Harvey Cushing**
  - 1917-1931 Developed bilateral suboccipital craniectomy for subtotal resection
  - Described CPA syndrome
  - Reduced mortality to 4%
  - Critical of translabyrinthine approach
"If the otologist has ambitions to treat these lesions there is no possible route more dangerous or difficult"

Cushing 1917
Historical Perspective

• Walter Dandy
  • 1916 Advocated for unilateral occipital flap with total tumor removal
  • Decreased mortality to 10%, lower recurrence rate

• William House
  • 1960s Resurrected translabyrinthine approach
  • Pioneered use of operating microscope and surgical drill
  • Also introduced middle cranial fossa approach
Relevant Anatomy

- Cerebellopontine Angle
  - Located in Posterior Fossa
  - Contains origin of many cranial nerves
  - Medial Border: brainstem
  - Lateral Border: petrous portion of temporal bone
  - Superior Border: middle cerebellar peduncle
  - Inferior Border: arachnoid of lower cranial nerves
  - Anterior Border: Clivus
  - Posterior Border: cerebellar tonsil
Epidemiology

- 80-90% of all CPA masses
- Incidence of 10 cases per million every year
- 2005 study suggested prevalence of incidental VS to be 2 in 10,000 people
- Autopsy studies have detected higher rates indicating VS may not become clinically apparent
- 95% sporadic, 5% associated with neurofibromatosis type 2
- Presents in Adulthood: mean age 50 (31 for NF2)
Tumor Biology

- Arise from vestibular division of 8th cranial nerve
- Likely equal distribution between superior and inferior divisions
- Historically thought to originate from Obersteiner-Redlich zone
  - Transitional area from central to peripheral myelin
- Now hypothesized to originate from Scarpa’s ganglion
  - Highest concentration of schwann cells
Genetics

- Neurofibromatosis type 2
  - Autosomal Dominant
  - NF 2 gene at 22q12.2: tumor suppressor gene normally prevents cell proliferation
  - Codes merlin protein

- Sporadic cases
  - “Double Hit” hypothesis: 2 hits to normal genes
Histology

Antoni A

Antoni B
Natural History

• Intracanalicular Phase
  • Hearing loss tinnitus, vertigo
• Cisternal Phase
  • Hearing loss, disequilibrium
• Compressive Phase
  • Trigeminal symptoms, Headaches
• Hydrocephalic Phase
  • 4th ventricle obstruction
  • Clinical deterioration
  • Facial weakness, vision loss
  • Respiratory Failure and death
## Symptoms

<table>
<thead>
<tr>
<th>Common Findings</th>
<th>Less Frequent Findings</th>
</tr>
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<tbody>
<tr>
<td>• Unilateral/asymmetric hearing loss</td>
<td>• Vertigo</td>
</tr>
<tr>
<td>• Up to 95%</td>
<td>• Roughly 20% predominantly with smaller tumors</td>
</tr>
<tr>
<td>• Tinnitus</td>
<td>• Headache</td>
</tr>
<tr>
<td>• Up to 70%</td>
<td>• Mid sized 1-3cm tumors: 20%</td>
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<tr>
<td>• Disequilibrium</td>
<td>• Tumors &gt;3cm: approaching 40%</td>
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<tr>
<td>• Up to 50%</td>
<td>• Facial Spasm</td>
</tr>
<tr>
<td>• Greater than 70% with tumors &gt;3cm</td>
<td>• 10% typically orbicularis oculi</td>
</tr>
<tr>
<td>• Facial Anesthesia</td>
<td>• Ophthalmic Manifestations</td>
</tr>
<tr>
<td>• Found in 50% in those with tumors &gt;2cm</td>
<td>• Nystagmus and Papilledema</td>
</tr>
<tr>
<td>• Typically midface hypoesthesia</td>
<td>• Lower Cranial Neuropathies</td>
</tr>
<tr>
<td></td>
<td>• &lt;3.5%</td>
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</table>
Signs

- Hitselberger’s Sign
  - Sensory component of CN 7
- Nystagmus
- Trigeminal dysfunction
  - Absent Corneal Reflex
  - Temporal Wasting and Masseter Weakness
    - Found rarely in advanced disease
- Cerebellar Signs
  - Gait ataxia
  - Poor finger to nose testing
Workup

• Audiologic Testing
  • Pure Tone and Speech Audiometry
  • Auditory Brainstem Responses
• Vestibular Testing
• Imaging
Hearing Loss

Audiometry

- Most common symptom
- Unilateral/Asymmetric
- 20% atypical pattern
- 5% present without hearing loss
- High frequencies typically affected
- Speech discrimination out of proportion to pure tone
- SSNHL
  - Occurs in 20% of patients
  - 1% of those with SSNHL will have a VS
Hearing Loss

Classification Systems

- Used in research to compare pre- and post-operative hearing
- Can aid in determining surgical approach
- AAO-HNS: Grade A and B considered serviceable hearing
- Gardner-Robertson: Grade 1 and 2 considered serviceable hearing

<table>
<thead>
<tr>
<th>Scheme</th>
<th>SDS</th>
<th>PTA</th>
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<tr>
<td>WRS</td>
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<td></td>
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<tr>
<td>I</td>
<td>70-100</td>
<td></td>
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<tr>
<td>II</td>
<td>50-69</td>
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<tr>
<td>III</td>
<td>1-49</td>
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<tr>
<td>IV</td>
<td>0</td>
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<tr>
<td>AAO-HNS</td>
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<tr>
<td>A</td>
<td>70-100</td>
<td>0-30</td>
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<tr>
<td>B</td>
<td>50-69</td>
<td>31-50</td>
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<tr>
<td>C</td>
<td>&gt;50</td>
<td>50+</td>
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<tr>
<td>D</td>
<td>&lt;50</td>
<td>50+</td>
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<tr>
<td>Gardner-Robertson</td>
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<tr>
<td>I</td>
<td>70-100</td>
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<td>II</td>
<td>50-69</td>
<td>31-50</td>
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<tr>
<td>III</td>
<td>5-49</td>
<td>51-90</td>
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<tr>
<td>IV</td>
<td>1-4</td>
<td>91+</td>
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<tr>
<td>V</td>
<td>0</td>
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Hearing Loss

Acoustic Reflexes

- Acoustic Reflex Decay
- Thresholds increased or absent if retrocochlear
Hearing Loss

Auditory Brainstem Responses

- Most common finding (40-60%) increased wave V latency
- Most specific finding only wave I present
  - Pattern present in only 10-20% of cases
- Normal ABR in 10-15% of cases
  - 33% of intracanalicular tumors
- High false positive rate >80%
Electronystagmography

Vestibular Testing

- Not very sensitive or specific for VS
- 70-90% will show an abnormality
- 50% of small tumors produce no abnormality
Imaging

- **CT**
  - Non-contrast: Poor Sensitivity
  - Contrasted: Ok for intracanalicular tumors
    - 90% of tumors enhance
    - Can miss tumors <2cm

- **MRI**
  - Gold standard
Imaging

MRI Findings

- Most Sensitive and Specific
- False negatives are rare
- Viral neuritis can produce false positives
- Sequences to obtain
  - Pre- and post-contrast T1 images
  - Fast spin echo T2 images

T1: mildly hypointense to brain, hyperintense to CSF

T1 Gd: contrast enhancement
Imaging

MRI Findings

- Most Sensitive and Specific
- False negatives are rare
- Viral neuritis can produce false positives
- Sequences to obtain
  - Pre- and post-contrast T1 images
  - Fast spin echo T2 images
Imaging

When to get an MRI?

• MRI Triggers
  • 15dB or greater asymmetry in 2 or more frequencies
  • Discrimination asymmetry of 16% or greater
  • Unilateral tinnitus
  • Asymmetric SNHL + trigeminal symptoms
  • Asymmetric SNHL + disequilibrium

• Goal of detection
  • Small tumors have great surgical outcomes
  • 98% cure, 99% intact facial nerve function, 80% preserved hearing

• Cost Analysis of ABR vs MRI: Study reviewed >300 cases of ASNHL
  • ABR screening: $125,000, with 29% of tumors unidentified
  • MRI screening: $156,000, with 0% lesions missed (at a cost of $99 more per case)
Reflexive MRI vs Serial Audiograms

- VS is not the only cause of asymmetric SNHL
- Tumor growth is generally slow
- Cost analysis of serial audiograms comparing MRI early vs after progression
  - Early MRI: $210,000
  - After audiogram progression: $186,000
- May risk:
  - Delayed diagnosis
  - Irreversible hearing loss
  - Inability to perform hearing preservative procedure
- Decision to obtain MRI should be based on global clinical picture
<table>
<thead>
<tr>
<th>Meningioma</th>
<th>Vestibular Schwannoma</th>
</tr>
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<tbody>
<tr>
<td>“Dural tail”</td>
<td>Ice cream cone appearance in IAC</td>
</tr>
<tr>
<td>Extend along petrous ridge</td>
<td>Bony erosion in IAC</td>
</tr>
<tr>
<td>Iso/hypointense on T1</td>
<td>Globular appearance</td>
</tr>
<tr>
<td>Hypo to hyperintense on T2</td>
<td>Can have cystic degeneration.</td>
</tr>
<tr>
<td>Sessile appearance</td>
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</tbody>
</table>
Imaging

Meningioma

Vestibular Schwannoma
Differential Diagnosis

- **Epidermoids:**
  - similar to cholesteatomas, from epithelial rest cells
  - Hypointense T1 and bright on T2
- **Arachnoid Cysts**
  - Treat with drainage
- **Schwanommas of other nerves**
- **Cholesterol Granulomas**
  - bright on T1 and T2
- **Embryonic Tumors**
  - excise when they cause dysfunction
- **Hemangiomas**
  - can form near geniculate producing progressive facial weakness
- **Glomus Tumors**
  - Jugular Foramen Syndrome
Management Options

- Observation with Serial Imaging
- Stereotactic Radiation
- Microsurgery
Management

Observation

- **Set Expectations**
  - 14-24% of patients being observed will go on to some form of treatment
  - Average growth 2mm/year, but can range from 0 to 2cm annually

- **Factors favoring observation**
  - Minimal symptoms
  - Long standing stable history, may indicate slow growth rate
  - Comorbid conditions and general health
  - Refusing other treatment

- **Protocol**
  - Follow up scan in 6 months, then serial imaging yearly
Management

Stereotactic Radiosurgery

• Introduced by Leskell in 1969
  • Damages vascular endothelium
  • Goal is to arrest tumor growth
• Gamma Knife uses multiple beams of ionizing radiation
• LINAC uses a linear accelerator as its source of radiation
• Benefits
  • Non-invasive
  • Early return to function
  • Avoids surgery
• Fractionated Radiotherapy
  • Multiple sessions
Management

Stereotactic Radiosurgery Outcomes

• Transient increase in tumor size due to central necrosis in roughly 20%
• Gamma Knife study by Boari et al
  • Tumor control in 97.1%
  • Tumor volume reduction in 82.7%, mean relative volume reduction 34.1%
• May require salvage surgery
• Increased trigeminal neuropathy
• Does not address disequilibrium
Management

Surgery

• Translabyrinthine Approach
• Middle Cranial Fossa Approach
• Retrosigmoid-Suboccipital Approach
Management: Translabyrinthine

Advantages
• Least cerebellar retraction
• Wide exposure of posterior fossa
• No size limitations for resection
• Easy identification of facial nerve
• Low recurrence rates
• Less headaches

Disadvantages
• Sacrifices residual hearing
• Fat graft donor site morbidity
Management

Translabyrinthine

• Postauricular incision
Management

Translabyrinthine

- Postauricular incision
- Mastoidectomy
- Dural Exposure
Management

Translabyrinthine

- Postauricular incision
- Mastoidectomy
- Dural Exposure
- Facial Nerve Identification
- Labyrinthectomy
- IAC Exposure
Management

Translabyrinthine

- Postauricular incision
- Mastoidectomy
- Dural Exposure
- Facial Nerve Identification
- Labyrinthectomy
- IAC Exposure
- Dura Incised
- Identify CN VIII components
Management

Translabyrinthine

- Postauricular incision
- Mastoidectomy
- Dural Exposure
- Facial Nerve Identification
- Labyrinthectomy
- IAC Exposure
- Dura Incised
- Identify CN VIII components
- Tumor Debulking
- Tumor Dissected from Facial Nerve
- Fat graft, dural repair, and closure
Advantages

• Best hearing preservation
• Adequate exposure of lateral IAC, CPA, and clivus
• Extradural drilling

Disadvantages

• Limited to smaller tumors <2cm
• Temporal lobe retraction
• Limited posterior fossa exposure
• Facial nerve dissection
Management

Middle Cranial Fossa

• Temporal skin incision
• Temporal craniotomy
Management

Middle Cranial Fossa

- Temporal skin incision
- Temporal craniotomy
- Temporal lobe retraction
- Dural elevation
- Geniculate identified and exposed
- Labyrinthine portion followed to IAC
- IAC exposed
- Tumor debulking and dissection
- IAC packed
- Craniotomy repaired
- Wound closure
Management: Retrosigmoid

Advantages

• Wide exposure of brainstem and lower cranial nerves
• Can preserve hearing
• Can resect large tumors
• Posterior approach to facial nerve

Disadvantages

• Cerebellar retraction
• Tumor must have <2cm IAC extension
Management

Retrosigmoid

• Post auricular Incision
• Suboccipital craniotomy
• Tumor debulking and dissection
• Dural and craniotomy closure
Management Considerations

• Factors
  • Serviceable Hearing
  • Tumor Size
  • Tumor Position
  • Anatomic Considerations
  • Surgeon Preference
  • Patient Preference
  • Debulking vs Total Resection
Complications

• Vascular Injury
• Facial Nerve Paralysis
• CSF Leak
• Headache
• Meningitis
• Tinnitus
• Balance Dysfunction
• Seizure, Hydrocephalus, and Stroke
• Mortality
Complications

Vascular Injury

- Arterial: AICA, labyrinthine artery, branches to facial nerve
- Venous: sigmoid sinus, petrosal vein of Dandy, vein of Labbé
  - Can produce cerebral edema
- Posterior Fossa Hemorrhage
  - Brainstem compression
  - Rapid neurologic deterioration
  - Potentially fatal
Facial Nerve Injury

Complications

- Preservation Rates (HB 1-2)
  - Retrosigmoid: 91%
  - Middle Cranial Fossa: 88%
  - Translabyrinthine: 77%

- Delayed Onset (>72hrs post op)
  - Occurs in 5% of cases
  - 79% regain function after 1 year
Complications

CSF Leak

• Can occur early (POD 2-3) or late (POD 10-14)
• Highest in retrosig approach: 11%
• Translab has 8% rate
• Middle cranial fossa has 6%

• Treatment
  • Conservative: bed rest, stool softeners, diuresis
  • May require lumbar drain, shunting, or operative repair
Complications

Headache

• Post op headache of >3 months duration occurs in roughly 10% of all cases
  • Retrosig: 21%
  • Middle Cranial Fossa: 8%
  • Translab: 3%

• Possible etiology
  • Bone dust contamination producing aseptic meningitis
  • Cranioplasty
  • Dural scarring
  • Migraines
Other Complications

- **Bacterial Meningitis**
  - *Staph aureus* most common organism
  - Standard presentation and treatment
- **Balance Issues**
  - Transient typically resolves in 6-9 months
- **Tinnitus**
  - 50% of patients with preop tinnitus have cessation of symptoms
- **Seizure/Hydrocephalus/Stroke**: rare <2% of cases
- **Mortality**: roughly 1%
Outcomes

• Recurrence Rates
  • Overall < 5%
  • Most occur after retrosig approach
  • Suspect with headaches, facial numbness, lower cranial neuropathies

• Hearing Results
  • Hearing preservation ranges from 30-80% depending on approach and patient selection
  • Better pre op hearing predicts better post op hearing
  • Larger tumors associated with lower rates of serviceable hearing
  • Gamma Knife and surgical treatment have similar hearing preservation rates
Conclusion

- Vestibular Schwannomas are the most common CPA tumor
- Unilateral SNHL or tinnitus requires further evaluation
- MRI is the confirmatory study of choice
- Some patients with few symptoms and small tumors can be observed
- Radiosurgery is appropriate for tumors <3cm in size
- In the setting of unserviceable hearing translab approach is best
- For hearing preservation use middle cranial fossa or retrosigmoid approach
Questions?