Acoustic Neuroma / Vestibular Schwannoma

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Introduction

- Benign tumour arising from abnormally proliferative schwann cells, which envelope the lateral portion of the vestibular nerve in the internal acoustic meatus

History

- First described by Sandifort in 1777
- William House introduced the translabyrinthine and middle fossa approach
- At 1992 consensus conference, name ‘vestibular schwannoma’ adopted

Epidemiology

- 8% of all intracranial tumors
- 90% of CP angle tumors
- > 95% sporadic (unilateral)
- 5% Neurofibromatosis type 2 (bilateral)
- Gender F:M= 3:2
- Age- fourth and sixth
- Incidence in general population 10 – 15 / million/ year
- Incidence in temporal bone collections 1-2%

Pathogenesis

- Usually arise from vestibular portion of eight nerve
- Commonly from IAC portion of the nerve, from its OBERSTEINER-REDLICH zone which is junction of central myelin produced by glial cells & and peripheral myelin produced by schwann cells. Also is the glial & neurilemmal junction.
- Superior division more common (some books mention equal frequency)

Histopathology

- Macroscopic
  - Benign
  - Yellowish to pinkish grey
  - Firm to rubbery consistency
  - Encapsulated
  - Nodular surface
  - Well defined plane of separation
  - Cysts formation within substance of tumor common → may form bulk of tumor
  - Cyst may have –CSF/hemorrhagic fluid
- Microscopic
  - Two patterns
    - Antoni A → closely packed cells with small spindle-shaped and densely stained nuclei. A whirled appearance of Antoni type A cells is called a Verocay body
    - Antoni B → looser cellular aggregation of vacuolated pleomorphic cells
Positive immunostaining with S100 \(\rightarrow\) differentiates from meningioma

Tumor spread
- From schwann cells of vestibular nerve in IAC
- Expands \(\rightarrow\) widens & erodes IAC \(\rightarrow\) cerebellopontine angle
- Involves cranial nerve V or IX, X, XI depending upon the direction of spread
- Later, brainstem, cerebellum \(\rightarrow\) increased intra cranial pressure

GROWTH RATE AND GROWTH PATTERN
- Mean growth rate \(\rightarrow\) 1.1 mm / yr (Scott Brown)
- Growth patterns
  1. Continuous growth
  2. No measurable growth
  3. No measurable growth followed by continuous growth
  4. Negative growth
  5. Various positive growth patterns

Classification
- Jackler system (adopted at Tokyo consensus conference 2001)

<table>
<thead>
<tr>
<th>Grade</th>
<th>Extrameatal Size (mm)</th>
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</thead>
<tbody>
<tr>
<td>Grade 1</td>
<td>Small 1-10</td>
</tr>
<tr>
<td>Grade 2</td>
<td>Medium 11-20</td>
</tr>
<tr>
<td>Grade 3</td>
<td>Moderately Large 21-30</td>
</tr>
<tr>
<td>Grade 4</td>
<td>Large 31-40</td>
</tr>
<tr>
<td>Grade 5</td>
<td>Giant &gt;40</td>
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</tbody>
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- Note should also be made if fundus empty or filled by tumor
- Also if VS sac was cystic

Clinical Features
- Stage 1 – Otological Stage
  - Progressive unilateral SNHL with tinnitus, more in higher frequency
  - In 5 – 10 % can be SSNHL
  - Very poor speech discrimination
  - Imbalance, unsteadiness
  - Nystagmus +/-
  - Facial nerve involvement \(\rightarrow\) hypoesthesia post EAC, taste loss, decreased lacrimation
• STAGE 2 – Trigeminal nerve involvement
  o Corneal reflex loss
  o Facial paraesthesia
• STAGE 3 – Brain stem & cerebellar compression
  o Ataxia, motor/sensory loss over extremities
  o Cerebellar ataxia, +ve romberg, dysdiadochokinesis, incoordination
• STAGE 4 – Increasing intracranial pressure
  o Headache, nystagmus, vertigo, diplopia, papilledema
• STAGE 5 – Terminal stage

INVESTIGATIONS

• PTA
  - Asymmetric unilateral SNHL
  - Slow progressive
  - More in higher frequencies
• Audiological tests
  - Speech discrimination poor; Roll over phenomenon present
  - SISI low → recruitment absent
  - ABLB → no recruitment
  - Tone decay → high → retrocochlear hearing loss
  - Stapedial reflex absent
• Diagnostic bekesy audiometry
  - Types III & IV → retrocochlear loss
• Electrocochleography
  - Broad waveform
• ABR
  - Very sensitive → 95% -100% detection rates
  - Interaural latency of >0.2 msec in wave V between 2 ears is significant
  - I-V interpeak latency > 4ms
• Caloric tests & electronystagmography
  - Canal paresis in 96 % of cases
  - Inferior vestibular nerve schwannoma may not show changes in caloric response
• Conventional x-ray
  - Views
    ▪ Stenver projection
    ▪ Chamberlain-towne projection
    ▪ Caldwell view
  - Look for
    ▪ Dilated IAC
    ▪ Funelling of IAC
    ▪ Erosion of posterior lip of IAC
• CT Scan
  - Can detect posterior fossa tumors upto 0.5 cm
  - Combined with intra thecal air (oxygen cisternography), even intra meatal tumors can be detected
• MRI with gadolinium contrast
  - Superior to CT Scan
  - Gold standard in imaging for vestibular schwannoma
  - Intracanalicular tumor of even a few mms can be detected.
  - Advantages
    ▪ High intrinsic contrast between tissues
    ▪ Absence of bone artefacts
    ▪ Multiplanar imaging
    ▪ No radiation

Differential diagnosis of CP angle tumor

1. Acoustic Neuroma 90%
2. Meningioma
   o Broad base on posterior surface of temporal bone or petrous bone
   o Imaging features
3. Primary Cholesteatoma
   o From congenital epithelial rest cell in temporal bone of PCF
   o Presents with progressive facial palsy or hemifacial spasm
   o Imaging
4. Arachnoid cyst
   o Congenital malformation
   o Infection-adhesive aracnoiditis
   o Trauma
   o Have CSF-so hypointense on T1
5. Schwannoma of other cranial nerves
   o Low in Jugular foramen(IX, X)
   o Facial paresis
6. Lipoma
7. Choroid plexus papilloma
   o Von Hippel Lindou disease
8. Hemangioma
9. Hemangiopericytoma – Bad prognosis
10. Glomus Jugulare(type IV)

Treatment decision factors

• Patient → age, general health, status of hearing in the contralateral ear, preference
• Tumor → size, location, extent, growth rate, bilateral, recurrence
• Surgeon skill & preference

• Treatment options
  o Observation
  o Stereotactic radiation therapy
  o Complete surgical excision
• Currently, no randomized, prospective clinical trial has compared the three treatment options and there are no clearly accepted, evidence-based, best practices for managing
acoustic neuroma

Conservative management  (Wait & Scan)

- **Indications**
  - Patients with small tumors
  - Advanced age
  - Poor general condition not fit for surgery
  - Unwilling for surgery
- **Yearly scanning advised (MRI)**
- **Disadvantage**
  - Risk of losing useful hearing (10-43%) in spite of no growth on MRI

Surgery

- **3 main approaches**
  - Middle fossa approach
  - Translabyrinthine approach
  - Retrosigmoid approach
- **Middle Fossa approach**
  - Young pt
  - Hearing to be preserved
  - Small tumour < 2.5 cms
  - Good exposure of lateral IAC, CPA, and clivus
  - Drilling is extradural decreasing morbidity
  - Disad
    - Temporal lobe retraction
    - Must dissect around facial nerve due to its superior position
    - Limited posterior fossa exposure
  - Steps
    - Position: Supine with head turned to affected side
    - Incision: Front of ear at level of zygomatic arch & curves upwards and backwards to sup temporal line expose the squamous temporal bone
    - 4X4 cm craniotomy, 2/3 ant to EAC, 1/3 post to it.
    - Middle fossa dura elevated off surface of petrous apex
    - Middle meningeal artery identified, followed till GSPN
    - Arcuate eminence identified
    - **House approach** – Follow GSPN to geniculate ganglion → trace back facial nerve to reach IAM
    - **Fisch approach** - angle between line of GSPN and plane of SCC (arcuate eminence) bissected → gives line of IAM
    - IAC identified, dura of IAC opened to long axis
    - Tumor dissected free, sup and inf vestibular nerve totally ablated
    - Internal auditory artery preserved
- **Translabyrinthine approach**
  - In cases with unserviceable hearing
  - Wide exposure of posterior fossa
  - No size limit for resection
  - Facial nerve easily identified throughout
  - Ease of facial nerve repair if damaged/resected during removal
  - Low recurrence
- Low headaches
- Disadv
  - Residual hearing is sacrificed
  - Requires abdominal fat graft
- Steps
  - Extension of Std postauricular incison upper end upto anterior wall of external meatus
  - Lower limit: 2cm behind mastoid tip
  - Superiorly based periosteal flap
  - Complete Mastoidectomy done, labyrinthectomy – IAC dissection
  - Dura of IAC opened, transverse crest & Bill’s bar identified
  - Debulking of tumor done & gradual complete dissection
  - Intracapsular removal of tumor using (House Urban rotatory dissector)
  - Meticulous closure middle ear with muscle, fascia graft for dura and antrum, mastoid defect with abdominal fat

- Retrosigmoid/Suboccipital Approach
  - Large size tumor
  - Hearing preservation possible
  - Wide exposure of brainstem and lower cranial nerves
  - Neurosurgeon familiarity
  - Consistent facial nerve identification
  - Disadv
    - Must be medially located
    - Lateral tumors risk injury to endolymphatic sac and vestibular labyrinth
    - Cerebellar retraction
  - Steps
    - Position: *Modified Park Bench* (supine with ipsilateral shoulder and hip bumped with rolls and padding, head flexed and rotated to opp shoulder)
    - ‘C’ shaped curvilinear incision from upper edge of pinna to spine of C2
    - 4x4 cm Craniotomy performed
      - **Anterior limit: Sigmoid sinus**
      - **Superior limit: transverse sinus**
    - Dura opened by triradiate incision
    - CSF run off or anaesthetist decreases CSF pressure
    - Cerebellum falls under own pressure plus retractor can be applied
    - Ant based U shaped opening made in dura
    - Intracranial segment of tumor debulked
    - Prevent injuy to V nerve, AICA, Lower Cr nerves
    - Another Laterally or medially based dural flap opened over IAC
    - Meatus drilled-exposing lesion till Bill’s bar & transverse crest
    - If hearing preservation desired **drill medial to lateral**
    - Seal with Bone wax, fibrin glue after closing dura
Conventional Radiotherapy – No role

Gamma knife radiosurgery

- **Principle** → To deliver a single precise, conformed dose of radiation tailored to margins of tumor
- **Indication**
  - No fixed guidelines
  - Smaller lesion to medium
  - Older individual
- **Method**
  - Rigid stereotactic frame attached to patient’s head
  - X, Y & Z co ordinates
  - Combined with radiological images, usually MRI
  - Targets pathological structures
  - Uses ionizing radiation (array of cobalt 201 sources or linear accelerator)
  - Median peripheral dose of 15 Gy
  - Directed through a variable array of collimators onto a single point
  - Delivers single high dose of radiation
- **Fate of tumor**
  - Tumor remains in situ but growth is stabilized
  - Modest degree of shrinkage
  - May swell in initial 6-18 months due to radiation oedema
  - Gadolium enhancement in centre is reduced
  - 5% regrowth
  - **Upto 92% control rate has been achieved**
- **Complications**
  - Cerebral oedema
  - Hydrocephalus
  - Vertigo
  - Seizures
  - Headache
  - Secondary oncogenesis - 5 cases reported

Fourth pathway

- Stereotactic RT after microsurgical debulking

Bilateral Vestibular Schwannoma (Neurofibromatosis-2)

- **Treatment options vary**
- **Bilateral-small tumor (less than 2 cm) & good hearing is a candidate for hearing preservation**
  - Surgery on one side (larger tumor) or with worse hearing & if hearing preserved → surgery other side in 6 months
- **Observation without surgery**
  - Small tumor
  - Unilateral hearing ear
Recent advances

- Ultrasonic aspiration
- Minimally invasive endoscopic neurosurgery
- KTP-532 CO2, Argon laser

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