Cerebellopontine Angle Tumors

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Overview

- Case presentation
- Historical perspective
- Acoustic Neuroma
- Other types of CP angle tumors
- Additional cases
Case Presentation

• 54 yo M with right-sided tinnitus for past year, mild dysequilibrium, headaches, and occasional facial twitching.

• Normal physical exam except:
  • Periodic spasms of right orbicularis oculi
  • Tuning fork lateralizes to left, with AC>BC AU

• Next step?
Case Presentation

Discrimination:
RIGHT: 40%
LEFT: 92%
CP Angle Tumors

- Acoustic Neuroma (60-90%)
- Meningioma (3-7%)
- Epidermoid (2-4%)
- Other schwannoma (1-4%) – CN V, VII, IX, X, XI
- Arachnoid cyst (1-2%)
- Others: hemangioma, lipoma, chondroma/chondrosarcoma, chordoma, metastases

A History Lesson

• Acoustic neuroma first described late 1700’s

• Mid-1800s: recognized that CP angle tumors cause syndrome of unilateral deafness, facial numbness, progressive blindness due to “optic neuritis” (papilledema)

• 1891: First attempt at removal – Charles McBurney
  • Suboccipital crani
  • Cerebellem swelled massively and had “to shave off the excess”
  • Tumor not removed
  • Patient expired 12 days later

A History Lesson

• 1913: Krause, Horsley, von Eiselsberg
  • 78% mortality
  • Survivors severely crippled

• 1917-1931: Cushing
  • Said attempts at total removal were “foolhardy in the extreme”
  • Debulking of central core of tumor
  • Used electrocautery and clips
  • Surgical mortality reduced to 4%
  • High mortality from recurrent tumor later

A History Lesson

• 1922-1941: Dandy
  • One of Cushing’s residents
  • Advocated total tumor removal (even while in training)
  • Mortality of 10%, but much lower recurrence

A History Lesson

• Translabyrinthine approach
  • Tried by multiple surgeons in early 1900s
  • Poor results
  • Cushing: “If the otologist has ambitions to treat these lesions there is no possible route more dangerous or difficult.” and “A proposal of this sort would never occur to an otologist who has general surgical training before he engaged in the particular surgery of his specialty.”
  • Technique resurrected by William house, who used surgical drill and microscope

Acoustic Neuroma: Epidemiology

- Incidence 10 per million per year
- 95% sporadic, 5% associated with NF-2
- Mean age at presentation 50 years
  - 31 years for NF-2

Acoustic Neuroma: Pathogenesis

- ANs are neither neuromas nor acoustic
- More accurate term: vestibular schwannoma
- 85% from inferior vestibular nerve in one study, other studies suggest equal frequency in superior vestibular nerve
- Most common site of origin: Scarpa’s ganglion
  - Highest concentration of Schwann cells
- NF2 gene on chromosome 22q12.2
  - Tumor suppressor gene – codes protein Merlin
  - 2-hit hypothesis

Question

Which of the following statements is false?

A) Vertigo is an uncommon symptom of acoustic neuroma.
B) Vertigo is more common with large acoustic neuromas than small tumors.
C) Sudden hearing loss may occur in more than 20% of acoustic neuroma cases.
D) Facial weakness is rare with acoustic neuromas.
Question

Which of the following statements is **false**?

- A) Vertigo is an uncommon symptom of acoustic neuroma.
- B) Vertigo is more common with large acoustic neuromas than small tumors.
- C) Sudden hearing loss may occur in more than 20% of acoustic neuroma cases.
- D) Facial weakness is rare with acoustic neuromas.
Acoustic Neuroma: Natural History

Intracanalicular → Cisternal → Brainstem compressive → Hydrocephalic

- Growth rates variable, but usually slow (average 1-2mm per year)
- Small proportion grow rapidly (1cm per year or more)

Acoustic Neuroma: Intracanalicular Phase

- Hearing loss
- Tinnitus
- Vertigo (uncommon)
  - Can indicate sudden growth, or vascular insult

Acoustic Neuroma: Cisternal Phase

- Hearing loss worsens
- Dysequilibrium
- Possible headaches
- Considerable growth can occur without much increase in symptoms

Acoustic Neuroma:
Brainstem Compression Phase

- Trigeminal symptoms may develop
- Headaches

Acoustic Neuroma: Hydrocephalus Phase

- Obstruction of 4th ventricle
- Rapid clinical deterioration
- Headaches
- Facial weakness
- Vision loss/diplopia
- Lower cranial nerve dysfunction
- Life threatening


Acoustic Neuroma:
Common Signs and Symptoms

- Unilateral/asymmetric hearing loss – up to 95%
- Tinnitus – up to 70%
- Dysequilibrium – up to 50%
  - More than 70% of large (>3cm) tumors
- Decreased facial sensation
  - In 50% of tumors larger than 2 cm
  - Most common in midface

Acoustic Neuroma: Less Common Signs and Symptoms

• Vertigo – uncommon (less than 20%)
  • More common in small tumors

• Facial weakness and spasm
  • Weakness rare
  • Minor twitching of orbicularis oculi in 10%

• Headache
  • 20% in medium-sized (1-3cm), 40% in large (>3cm)

• Ophthalmologic manifestations
  • Nystagmus and decreased corneal reflex

• Lower cranial nerves – rare (<3.5%)


Question

- Which of the following patients has an acoustic neuroma? (assume all sensorineural)

X = left
O = right
Which of the following patients has an acoustic neuroma? (assume all sensorineural)

X = left
O = right

A
B
C
D
Question

- Which additional finding(s) on audiometric evaluation would increase suspicion of an acoustic neuroma?
  - A) Rollover
  - B) Acoustic reflex decay
  - C) Present OAEs
  - D) A and B
  - E) All of the above
Question

Which additional finding(s) on audiometric evaluation would increase suspicion of an acoustic neuroma?

- A) Rollover
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<table>
<thead>
<tr>
<th>Acoustic Neuroma: Hearing Loss</th>
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<tr>
<td><strong>Most frequent symptom</strong></td>
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<td><strong>Unilateral/asymmetric, typically high frequencies first to be affected</strong></td>
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<td><strong>Speech discrim affected out of proportion with pure tones</strong></td>
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<td><strong>Can be atypical pattern (upsloping, trough-shaped) in 20%</strong></td>
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<td><strong>Can involve sudden SNHL +/- recovery in &gt;20%</strong></td>
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<td>• Although only 1-2% of those with sudden SNHL will have an acoustic neuroma</td>
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<td><strong>15% have subjectively normal hearing</strong></td>
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<td>• 4% normal on audiogram</td>
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<td>• 7% symmetric on audiogram</td>
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Question

The ABR finding most specific for an acoustic neuroma is:

- A) Presence of wave I and nothing thereafter
- B) All waves present with increased wave V latency
- C) No waves present
- D) None of the above
The ABR finding most specific for an acoustic neuroma is:

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- A) Presence of wave I and nothing thereafter
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- D) None of the above
The ABR finding most *common* with an acoustic neuroma is:

- A) Presence of wave I and nothing thereafter
- **B) All waves present with increased wave V latency**
- C) No waves present
- D) None of the above
Acoustic Neuroma: Histopathology

- Two morphologic patterns
- **Antoni A**: dense, packed cells
  - Verocay bodies: whirled appearance of Antoni A cells with intervening anuclear areas
- **Antoni B**: looser, vacuolated, pleomorphic cells
  - Seen more in larger tumors
- **Positive S-100 stain**


Acoustic Neuroma: Workup

• Pure tone and speech audiometry

• Auditory brainstem responses (ABR)
  • Increased wave V latency most common (40-60%)
  • No waves present (20-30%)
  • Wave I only (10-20%)
  • Normal (10-15%; 33% of intracanalicular tumors)
  • False positive rate of >80%
  • Stacked ABR – more sensitive and specific

• Vestibular testing
  • Insufficient sensitivity and specificity to help in diagnosis
  • Prognostication about speed of vestibular compensation after AN surgery

• Imaging (Gd-enhanced MRI gold standard)

Acoustic Neuroma: Imaging

- Non-contrast CT
  - Poor sensitivity: relies on dilation of IAC

- Contrast-enhanced CT
  - Reliable for tumors >2cm

- MRI
  - Most sensitive and specific
  - Thin cuts through IAC/CPA
  - Obtain pre- and post-contrast T1 images
  - Fast spin echo T2 high-res images
  - Fat suppression/fat saturation
  - False negatives rare
  - False positives due to viral mononeuritis of CN VII or VIII

Acoustic Neuroma: MRI Findings

- CPA and IAC components, early compression of pons
- Rounded CPA mass with cone-shaped IAC stem
- T1: mildly hypointense to brain, hyperintense to CSF
- Gd-T1: Marked contrast enhancement, small cystic areas more visible

Acoustic Neuroma: MRI Findings

- Fast spin echo T2
- Intracanalicular acoustic neuroma

Acoustic Neuroma MRI Findings

- **A)** T1 pre-contrast
- **B)** T1 post-contrast
- **C)** T2
- **D)** Constructive interference in steady-state (CISS)

MRI: A Note on Fat Suppression

• Should routinely include in assessment of IAC/CPA lesions

• Normally fat is bright on T1 and fast spin echo T2

• Fat suppression (a.k.a. fat saturation) reduces signal intensity of fat
  • Allows contrast with fluid-filled structures on T2
  • Allows contrast with Gad-enhancing areas on T1
Fat Suppression in Action

Acoustic Neuroma: Why to get the MRI

- **Goal is early detection**
  - Small tumors (<1cm) have excellent surgical outcome
    - 98% cure, 99% facial nerve preservation, 80% hearing preservation

- **Audiometric/clinical thresholds:**
  - 15 dB or higher asymmetry in 2 or more frequencies
  - And/or discrim asymmetry of 16% or more
  - Unilateral tinnitus
  - Asymmetric SNHL and trigeminal symptoms
  - Asymmetric SNHL and dysequilibrium

- **Cost analysis of 312 patients with ASNHL (ABR vs MRI)**
  - ABR as initial screen: $125,000, with 29% of tumors missed
  - MRI as initial screen: $156,000 ($99 more per patient), with 0% of lesions missed

Acoustic Neuroma: Why NOT to get the MRI

- Asymmetric SNHL highly prevalent in general population
- Tumors are typically slow growing
- Another cost analysis (early MRI vs serial audiograms with later MRI for progression)
  - For 100 patients:
    - serial audiogram with later MRI: $186,000
    - MRI first: $210,000
- Con argument: do not rely on strict audiometric criteria, instead need to look at whole clinical picture, patient preferences and individual situation, etc.

CP Angle Tumors

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Meningioma

- Arises from posterior surface of petrous bone or along sigmoid sinus
- Presentation similar to acoustic neuroma
- Usually large before causing CN VIII compression
- Radiation exposure is risk factor


Meningioma

- 98% sporadic
- 2% hereditary, especially NF2
  - 35% of NF2 patients have meningiomas
- Originate from cells lining arachnoid villi
- Firmly adherent to dura
- Displaces neural tissue
- May invade adjacent bone via haversion canals


Meningioma
Histopathology

• WHO Grade I – typical. 90%
  • Common Subtypes: Meningothelial, fibrous, transitional, psammomatous, angiomaticus
  • Low aggressiveness
• WHO Grade II – atypical. 7%
  • Subtypes: atypical, clear-cell, chordoid
  • Intermediate aggressiveness
• WHO Grade III – anaplastic. 3%
  • Subtypes: anaplastic, papillary, rhabdoid
  • Highly aggressive, potential for brain invasion

Meningioma
Histopathology

Meningioma Imaging

• Well-circumscribed, extra-axial, sessile, broad dural attachment
• May have calcifications
• CT: usually hyperdense to brain, may be isodense
  • Intense, mostly homogeneous contrast enhancement
  • May have hyperostotic bone
• MRI:
  • T1: isointense or mildly hypointense to gray matter
    • Most enhance brightly with Gad – can be heterogeneous
    • Dural tail may be present
  • T2: may be isointense (most common), hyperintense, or hypointense. Mottled/speckled heterogeneity.

Meningioma Imaging

- A) Post-Gd T1 MRI
- B) T2 MRI
- C) CISS MRI

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Epidermoid
a.k.a. Primary Cholesteatoma

- Originate from epithelial rests within temporal bone or CP angle
- Stratified squamous epithelium with keratin debris
- Slow-growing, symptoms not apparent until 2\textsuperscript{nd} to 4\textsuperscript{th} decade
- Facial nerve twitching and/or weakness more common than with acoustic neuromas

Epidermoid Imaging

- **CT:**
  - Density similar to CSF (hypodense to brain)
  - No contrast enhancement
  - Irregular borders, may erode bone

- **MRI:**
  - T1: hypointense
    - No contrast enhancement (differentiates from schwannoma, meningioma, chondroma)
  - T2: iso- or hyperintense
  - Diffusion-weighted: differentiates from arachnoid cyst
    - Epidermoid: bright
    - Arachnoid cyst: dark

Epidermoid Imaging

- T1 MRI
- T2 MRI

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Arachnoid Cyst

- Thin-walled cysts, contain entrapped, yellow CSF
- Derived from congenital developmental abnormalities
- Similar CT and MRI appearance to epidermoids, except:
  - Smooth surfaced (epidermoid is scalloped)
  - DWI: Dark (epidermoid is bright)
- Management:
  - Conservative: observation, diuretics
  - Surgical: drainage (not excision)

Hemangiomas

- Capillary – typically arise from perigeniculate area of facial nerve
  - Cause facial nerve symptoms much earlier than facial neuroma
  - Pulsatile tinnitus with exposure of basal turn of cochlea
  - Usually small lesion with contrast enhancement on imaging
  - Irregular bony erosion on CT

- Cavernous – IAC
  - Cause symptoms typical of acoustic neuroma
  - Similar appearance to AN on MRI, except slightly more hyperintense


Acoustic Neuroma


Name that Tumor

Epidermoid

Name that Tumor

T1 MRI without contrast

T1 MRI with contrast

T2 MRI

CT without contrast

Petrous Apex Mucocele

Arachnoid Cyst

Summary

- Acoustic neuroma is most common CP angle tumor
- Others to consider include meningioma, epidermoid, arachnoid cyst, other schwannomas, and many other more rare lesions
- Early diagnosis leads to higher chance of cure with lower complications and better chance of hearing preservation
- No consensus on when to obtain MRI. Audiometric data as well as overall clinical picture should be considered.
- Different CP angle tumors have distinguishing characteristics on MRI
- Various treatment strategies are beyond the scope of this grand rounds, and may make an excellent future topic!
Questions?
References


