Developing an Evidence Based Referral Protocol for Early Diagnosis of Vestibular Schwannomas

Jessica Scott, AuD
JDVAC Conference
February 23, 2010
The views expressed in this presentation are those of the author and do not reflect the policy of the Department of the Army, Department of Defense, or US Government.
Introduction

• Vestibular Schwannoma (VS)
  ▪ Slow growing benign tumor
  ▪ Develops on Vestibulocochlear Nerve (CN VIII)
  ▪ Overproduction of Schwann Cells
    • Wrap around nerve fibers
    • Help support and insulate for increased conduction
  ▪ Common Symptoms (Cummings et al., 2005)
    • Hearing loss, Tinnitus, Vestibular, Aural Fullness, Otalgia
    • CN VII (Facial) or CN V (Trigeminal)
    • Labyrinthine artery
  ▪ Incidence of 1 in 100,000 (NIDCD, 2004)
Introduction

• Tests that can potentially indicate the presence of a VS
  ▪ Audiologic (Pure tone thresholds, speech discrimination testing, acoustic reflex thresholds, acoustic reflex adaptation (decay))
  ▪ Vestibular (ENG/VNG, rotary chair testing, VEMP)
  ▪ Evoked Potentials (ABR, ECochG)
  ▪ Radiologic (MRI, CT Scan)

• Most definitive test for diagnosing VSs
  ▪ Contrast-enhanced MRI
  ▪ Contrast (typically gadolinium) - provides a greater contrast between normal and abnormal tissue (Cummings et al., 2005)
Literature Review

• Previous Referral Criteria
  - Obholzer et al. (2004)
    • Unilateral hearing loss: $\geq 15$ dB HL at two adjacent frequencies
    • Bilateral hearing loss: $\geq 20$ dB HL at two adjacent frequencies
  - Sheppard et al. (1996)
    • $\geq 15$ dB HL asymmetry
    • Unilateral tinnitus in the presence of normal hearing
Data Collection

• All data were collected retrospectively
  ▪ Age/Gender
  ▪ Previous Hx of Hazardous Noise Exposure
  ▪ Hx of hearing loss (newly identified, stable, progressive, none)
  ▪ Who referred them for a MRI (Audiology, ENT, Neurology, etc.)
  ▪ Patient’s presenting symptoms
  ▪ Audiologic test results
    • Pure tone thresholds (excluded if air-bone gaps)
    • Speech discrimination scores
    • Tympanometry* (excluded if abnormal)
    • Acoustic reflex thresholds
    • Acoustic reflex adaptation (decay)

• Only demographic exclusion criteria: No patients < 18 years of age
Participants

• Original chart review – 628 patients
• Final participant groups ($N = 328$)
  ▪ Positive MRI: 19 patients (age 31-73, $M = 53.37$)
    • Males ($n = 10$)
    • Females ($n = 9$)
  ▪ Negative MRI: 309 patients (age 20-85, $M = 48.28$)
    • Males ($n = 221$)
    • Females ($n = 88$)
Referral Breakdown

• Audiology
  ▪ Hearing Loss – 114
  ▪ Tinnitus Only – 4
  ▪ Dizziness/Vertigo Only – 0

• ENT
  ▪ Hearing Loss – 160
  ▪ Tinnitus Only – 16
  ▪ Dizziness/Vertigo Only – 12

• Neurology
  ▪ Hearing Loss – 111
  ▪ Tinnitus Only – 0
  ▪ Dizziness/Vertigo Only – 5

• PCM
  ▪ Hearing Loss – 110
  ▪ Tinnitus Only – 7
  ▪ Dizziness/Vertigo Only – 4
Results/Conclusions

• Evaluation
  ▪ Case Hx
  ▪ Diagnostic Testing
    • Pure tones (air and bone)
    • Word recognition measures (in quiet and/or noise and to include measure of PIPB Rollover)
    • Immittance evaluation
      – Tympanometry
      – Acoustic Reflex Thresholds (specifically contralateral acoustic reflexes)
      – Acoustic reflex decay (adaptation)
    • Vestibular Testing (as needed based on presenting symptoms)
Results/Conclusions

• Diagnosis
  – Presenting symptoms of unilateral/asymmetric hearing loss and unilateral/asymmetric tinnitus **AND** one or more of the following test outcomes
  – Pure tone threshold asymmetry of 10 dB or more at three adjacent frequencies
  – Asymmetric word recognition scores (not explained by hearing loss)
  – Positive (measured) rollover
  – Elevated or absent contralateral acoustic reflexes (not explained by degree of hearing loss, e.g., in the presence of a conductive component to the hearing loss).
Results/Conclusions

• If the patient has either unilateral/asymmetric hearing loss (asymmetry of 10 dB or more at three adjacent frequencies) or unilateral/asymmetric tinnitus with no other significant test results or symptoms the audiologist can choose to:
  
  – Based on clinical judgment of the specific case, refer the patient for an MRI, or
  – Refer for medical evaluation for evaluation of other non-retrocochlear (medical) etiology.
Case 1

- 32 y.o. M; Blackhawk Mechanic
- Hx of PE tubes as a child
- Seen in FEB 2008 for hearing screening
  - Identified as normal with no subsequent referral
  - Moderate loss noted at 6 kHz in the right ear only
- Seen by PCM in FEB 2008 for
  - Earache AD
  - Pressure/fullness with inability to pop AU
  - Otoscopy: Normal AD; Erythemia noted AS
  - Tx for OM
- Seen by PCM in NOV 2008
  - Sudden onset (2 days prior) of bifrontal headache
  - Sudden hearing loss AD (1 week prior), although the pt. reported suspected hearing loss AD for 1 year w/o evaluation
  - Earache AD
  - Sore Throat – 2 days
  - Congestion
  - Evaluation
    - Hemorrhagic discharge noted AU
    - Right Otoscopy – decreased mobility with signs of fluid
    - Left otoscopy – unremarkable
  - Tx for OM and referred to Audiology for chronic hearing loss AD
Case 1

- Hearing Screening in DEC 2008
  - Moderate to severe responses AD above 2 kHz
- Initially seen in DEC 2008 in ENT
  - Subjective right sided hearing loss (HL)
  - HL often accompanied by
    - Crackling sounds
    - Sharp pains lasting seconds with rapid, spontaneous resolution
  - No balance complaints
  - Intermittent right ear tinnitus
  - Refer to Audiology
Case 1

- OAEs
  - Present/robust AS
  - Largely absent above 1 kHz AD

- WR
  - AS: 100% @ 40 dBHL
  - AD: 98% @ 45 dBHL

- Normal tympanograms AU

- ARTs (I) – present AS (.5, 1, and 2 kHz) and AD (.5 and 1 kHz)
Case 1

- ENT Ordered MRI of the IACs with gadolinium enhancement
- Impression: 1.8 cm anterior-posterior x 2.5 cm transverse x 1.8 cm craniocaudal right cerebellar pontine angle mass. The lesion expands into the internal auditory canal. Some erosion of the petrous apex of the right temporal bone was also noted
Case 1

- **WR**
  - AS: 100% @ 50 dBHL
  - AD: 64% @ 50 dBHL; 96% @ 90 dBHL

- **Normal tympanograms AU**

- **ARTs**
  - Probe AS - Normal
  - Probe AD
    - Ipsi – Normal (.5, 1 kHz); Absent (2 kHz)
    - Contra – Elevated (.5 kHz); Absent (1, 2 kHz)

- **Negative Acoustic Reflex Decay**
Case 1

• VS was removed in AUG 2009
  – Post-occipital approach with total severance of auditory/vestibular nerve
• Complication of CSF seroma
• Migraines – successfully tx with Floricet and caffeine pills
• Pt. initially had facial nerve paresis which is almost resolved by DEC 2009
• Also c/o balance issues and blurry vision which has also improved
• Pt. discussed BAHA vs. CROS aid
• ENT noted only a small portion of bone to work with for BAHA considering his craniotomy
Case 2

• 60 y.o. M initially seen in Audiology in June 1987
  – Presented with
    • Dysequilibrium problems
    • Hearing loss AS>AD
    • Distortion AS
    • Intermittent pressure in his head
    • Tinnitus AS
Case 2

- **WR**
  - AD: 96% @ 55 dBHL; 92% @ 90 dBHL
  - AS: 50% @ 65 dBHL; 44% @ 90 dBHL

- **Normal tympanograms AU**

- **ARTs**
  - Probe AD - Normal
  - Probe AS
    - Ipsi – Normal/Elevated (.5, 1, 2 kHz)
    - Contra – Normal (.5 kHz); Elevated/Absent (1, 2 kHz)

- **Negative Acoustic Reflex Decay**
Case 2

- ABR – June 1987
  - AD
    - Absolute wave latencies were delayed for Waves III and V.
    - Morphology was considered fair
  - AS
    - Absolute latencies were delayed for Waves III and V.
    - Comparison between ears indicated significant delay for Wave V in the left compared to the right
    - Morphology was considered poor
Case 2

• Vestibular Evaluation in July 1987
  – Ocular Motor Evaluation
    • Significant for saccadic smooth pursuit at low frequencies
  – No spontaneous nystagmus
  – Dix Hallpike right – right beating, non-latent nystagmus with no symptoms. Not classic for BPPV
  – Dix Hallpike left – Normal
  – Direction fixed, right beating nystagmus of 7-9 deg/sec seen in head/body right positions
  – Bithermal calorics – 44% left weakness
Case 2

- Stable SNHL since JUN 1987
- WR
  - AD: 100% @ 50 dBHL
  - AS: 60% @ 60 dBHL
Case 2

- Significant decreased thresholds noted AS
- WR
  - AD: 96% @ 50 dBHL; 96% @ 90 dBHL
  - AS: 28% @ 55 dBHL; 8% @ 90 dBHL
- Normal tympanograms AU
- ARTs
  - Probe AD – Normal
  - Probe AS – Essentially Absent
- Negative Acoustic Reflex Decay – Probe Right Only
Case 2

• Referred for MRI by ENT
  – Noted a 1 x 1.5 cm acoustic neuroma on the left side
Case 2

• ENG evaluation in NOV 1990
  – Ocular Motor tests were normal
  – No spontaneous
  – No positional nystagmus
  – Negative Dix Hallpikes
  – Rotary Chair – abnormal phase leads and decreased gain
Case 2

- Removal of VS in NOV 1990.
- Audio was unchanged AD with an anacoustic AS.
- Pt. reports no significant hearing difficulties
Case 3

- 73 y.o. M
- Presented to Audiology in DEC 2008
- Previous Noise Exposure
- Fm hx: Father and brother have both had different brain tumors
- Presentation:
  - Hearing loss
  - Difficulty understanding speech
  - No earache
  - No tinnitus
  - No balance difficulties
Case 3

- WR
  - AS: 100% @ 90 dBHL
  - AD: 84% @ 75 dBHL; 48% @ 90 dBHL
- Normal tympanograms AU
- ARTs
  - Probe AS – Normal, except Absent at 2 kHz in contra condition
  - Probe AD – Absent
Case 3

- Negative MRI of the IACs
- Pt. was fit with hearing aids in both ears
Case 4

• 42 y.o. F

• Pt. initially presented to Audiology in OCT 2005 for a second opinion on a Meniere’s Dx

• Presentation
  – No subjective hearing loss
  – Intermittent tinnitus
  – Vertigo, noted since Spring 2005
  – No ear pain
Case 4

- **WR**
  - AS: 92% @ 80 dBHL
  - AD: 100% @ 80 dBHL
- Normal tympanograms AU
- **ARTs**
  - Contralateral right and left reflexes were WNL
- **Negative Acoustic Reflex Decay** in both ears

![Graph showing threshold vs frequency for right and left ears]
Case 4

• Next OTO-HNS note is from 5/2006

• Presentation
  – Vertigo following a sinus infection 4 weeks prior
  – Vertigo lasted 2.5 weeks. It began to resolve but not entirely
  – Today, she could say her symptoms occur looking to the right
  – Intermittent tinnitus (non-localized)

• Conclusion
  – Audiogram
  – Consult for VRT
  – If no improvement consider MRI
Case 4

• Seen in Audiology in May 2006
  – Presentation
    • Subjective hearing loss AD
    • Muffled sound AD
    • Pressured sensation AD
    • Tinnitus AD only
Case 4

- Stable hearing
- Excellent high level word recognition in both ears
- Normal tympanograms and ARTs
Case 4

• Seen again in ENT in AUG 2006
  – Positive Romberg to the right
• Suspected BPPV; however ordered MRI to rule out intracranial pathology considering
  – Unilateral tinnitus
  – Abnormal Romberg
Case 4

- MRI of the IACs
  - 3 x 4 mm lesion noted in the right IAC involving the right CN XIII
Case 4

• Seen in DEC 2007 in Audiology for monitoring
  – Severe Tinnitus AD
  – Difficulty understanding speech especially in background noise

  – Stable Normal hearing noted AU; Excellent WR with no evidence of rollover; Normal tympanograms and ARTs in both ears
Case 4

• FEB 2008 (Audio and Vestibular Testing)
  – Presentation
    • Constantly imbalanced
    • Positional vertigo
    • Vertigo to temperature changes
    • Uses CC on television
    • Right severe tinnitus
    • Has begun balance training
Case 4

- DHI 68/100
- Audiogram
  - Stable, normal thresholds
  - Excellent word recognition w/ no evidence of rollover
  - Normal tympanograms
  - Normal acoustic reflexes
  - Negative acoustic reflex decay in both ears
- ABR – Normal
- Normal OAEs
- No spontaneous
- Positional – not clinically significant
- Negative Dix Hallpike
- Left beating head shake nystagmus
- Calorics – 76% right unilateral weakness
- Normal VEMPs
- Slightly abnormal posturography. Normal sensory analysis
Case 4

• Removal of VS in APR 2008 via a middle fossa approach
  – Persistent tinnitus
  – Mild imbalance that is improving
  – Intermittent echo sound AD with occasional tinny quality
  – Some right ear pain behind the right pinna
Case 4

- MRIs in 2009 have indicated a 7mm lesion in the right IAC
  - Residual lesion vs. granulation tissue
- Newest Audio in SEPT 2009 was essentially normal
- ENT in SEPT 2009
  - Right eye is “slow”
  - Pain in the medial side of R eyebrow
  - Mildly improved tinnitus
  - Dry eyes
  - Difficulty with taste on the right side of her tongue
  - Mild preauricular numbness and pain AD
  - Twitch in right eye tx with botox that is more tolerable
Case 5

• 37 y.o. M
• Presented to Audiology Clinic in JAN 2010
• C/o
  – Intermittent ear pain AS
    • Episodic, sharp pain
    • Mostly AS, but rarely noted AD
    • Pain for 10-15 seconds
    • Randomly several times a week
  – No hearing loss
  – No tinnitus
  – No dizziness
  – Ears frequently feel like there is cotton in them
Case 5

- WR
  - AS and AD: 100% @ 50 and 90 dBHL
- Normal tympanograms AU
- ARTs - WNL
- Negative Acoustic Reflex Decay in both ears
Case 5

• Initially had CT scan which was normal in DEC 2009
• Sent for MRI of the IACs for chronic left ear pain in JAN 2010
• Impression: 1.5 mm right intracanicular VS
References


Contact Information

• Jessica.Scott1@amedd.army.mil
Thank You!!