Diagnosis and Management of Spontaneous Cerebrospinal Fluid Fistula

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SPONTANEOUS CEREBROSPINAL FLUID OTORRHEA

- Presence of CSF within the confines of the temporal bone
  - Absence of traumatic, neoplastic, infectious or iatrogenic cause

- Theories of Etiology
  - Erosion into middle ear/mastoid by arachnoid granulation
  - Congenitally thin skull base
    - Erosion by constant pulsation of temporal lobe
INCIDENCE

• True incidence is unknown
• Increased reporting in past 2 decades
  • Likely due to increased recognition of the disorder
    • Not an increased incidence
    • However, obesity is on the rise
• May be underdiagnosed

RISK FACTORS

• Obesity
• Idiopathic Intracranial Hypertension (IIH)
  • Formerly Benign Intracranial Hypertension (BIH)
    • Disease is NOT benign
• Female Gender
• Increasing age
ROLE OF OBESITY

- UTSW experience (n=34)
  - Spontaneous CSF fistula of temporal bone
  - January 2007-December 2011
  - Mean age 60.3 years (46-75 years)
  - Mean BMI 36.7 kg/m² (24.2-57.8 kg/m²)

IDIOPATHIC INTRACRANIAL HYPERTENSION

- Elevated intracranial pressure with an unknown cause
  - Absence of mass or hydrocephalus
  - Classically overweight women of childbearing age
  - Frequently associated with papilledema
  - 25% experience loss of visual function
  - Upper limit of normal CSF Opening pressure
    - 18-20 cm/H20
    - Opening pressure of 25 cm/H20 for required for diagnosis
EMPTY SELLA

- Normal Subjects
  - 5-6%

- Idiopathic Intracranial Hypertension
  - 10-93%

- Spontaneous CSF Otorrhea
  - 71-80%

COMPLICATION OF CSF OTORRHEA

- Meningitis
  - May be cause for initial presentation
  - Absolute risk has not been defined
CLINICAL PRESENTATION

• Complaint of hearing loss and aural fullness
• Middle ear effusion (typically unilateral)
  • Recalcitrant to treatment
  • Atypical population for otitis media
  • Persistent, watery otorrhea after myringotomy
• Persistent, clear rhinorrhea
• Meningitis
  • Possibly recurrent
CONFIRMATION OF CSF

- Middle ear fluid analysis
  - Tympanocentesis
  - Myringotomy
- Beta-2 Transferrin
  - Present in:
    - CSF
    - Vitreous humor
    - Perilymph
  - More specific than halo sign or glucose strip testing

RADIONUCLIDE CISTERNOGRAPHY

- Cases with a high index of suspicion and a negative Beta-2 Transferrin
- Procedure:
  - Myringotomy with tube placement
  - Wick placed in external auditory canal
  - Lumbar puncture with radionuclide administration
  - 24 hours later, presence of radionuclide on wick confirms CSF leak
RADIOGRAPHIC STUDIES

- High-Resolution Computed Tomography
  - Visualized defect and Beta-2 transferrin is positive
    - No additional radiographic studies necessary
- Coronal T2 Weighted MRI
  - Will help to determine presence of encephaloceles

HRCT
MRI

SURGICAL REPAIR

• Transmastoid Repair
  • Advantages
    • No craniotomy required
    • Access to posterior fossa defects
  • Disadvantages
    • Ossicles impede access to tegmen tympani
    • Difficult to repair large defects
SURGICAL REPAIR

• Middle Fossa Craniotomy
  • Advantages
    • Able to access entire middle fossa floor
    • No ossicular removal required
    • Large defects can be repaired
  • Disadvantages
    • Increased morbidity of craniotomy
    • Unable to repair posterior fossa defects

SURGICAL REPAIR

• Combined Transmastoid/Middle Fossa Craniotomy
  • For defects of middle fossa and posterior fossa
MIDDLE FOSSA CRANIOTOMY

- Incision
  - Straight vertical
  - Question Mark
- Harvest Fascia
  - Temporoparietal / Temporalis
- Divide Temporalis Muscle
  - Leave a 1 cm cuff for closure

MIDDLE FOSSA CRANIOTOMY

- Craniotomy
  - 4x4 cm
  - Centered over zygomatic root
- Elevate dura from middle fossa floor
- Resect encephalocele if present
- Pack epitympanum with gelfoam
MIDDLE FOSSA CRANIOTOMY

- Resurface middle fossa floor
  - Bone graft
  - Fascia
  - Bone cement
    - Calcium Phosphate (Hydroset)
- Dural repair
  - Direct suturing
  - Collagen matrix (Duragen)
MEDICAL AND SURGICAL TREATMENT OF IIH

- Acetazolamide
  - Can decrease CSF production by 48%
  - Reduction of intracranial pressure by 10 cm/H₂O
- Ventriculo-peritoneal shunt
- Referral to ophthalmologist

UTSW EXPERIENCE

- 36 ears in 34 patients since January 1, 2007

<table>
<thead>
<tr>
<th></th>
<th>Number</th>
<th>%</th>
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<tbody>
<tr>
<td>Female</td>
<td>27</td>
<td>79.4</td>
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<tr>
<td>Male</td>
<td>7</td>
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<table>
<thead>
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<th></th>
<th>Mean</th>
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<tr>
<td>Age (years)</td>
<td>60.3</td>
<td>46-75</td>
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<tr>
<td>BMI (kg/m²)</td>
<td>36.7</td>
<td>24.2-57.8</td>
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UTSW EXPERIENCE

- History of Meningitis
  - 14.7% (5 of 34 patients)

OPERATIVE APPROACH

- Transmastoid
  - 2 of 36 ears
- Middle fossa
  - 34 of 36 ears
- Combined procedure
  - 1 for a revision of a middle fossa approach
RECURRENTS

- 2 of 36 ears
- 5.5%
- Both middle fossa approach
- Pt 1: required revision operation
  - Combine middle fossa/transmastoid
  - Has redeveloped a leak
- Pt 2: Recurrent effusion has resolved with VP shunting

COMPLICATIONS

<table>
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<tr>
<th>Complication</th>
<th>Percentage</th>
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<tbody>
<tr>
<td>Recurrence</td>
<td>5.5%</td>
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<tr>
<td>Subdural hematoma</td>
<td>5.6%</td>
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<tr>
<td>Temporal lobe abscess/meningitis</td>
<td>2.8%</td>
</tr>
<tr>
<td>Tympanic Membrane Atelectasis</td>
<td>2.8%</td>
</tr>
<tr>
<td>Seizure</td>
<td>0%</td>
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UTSW EXPERIENCE

• Lumbar Puncture opening Pressure (19 patients)
  • Median: 19 cm/H2O
  • Range: 10.5-55 cm/H2O
  • Elevated (>20 cm/H2O) in 41%
• Five patients underwent ICP reduction techniques
  • Three placed on acetazolamide
  • Two under VP shunting

MRI EVIDENCE OF ELEVATED ICP

• MRI obtained in 24 patients
  • 45.8% with evidence of elevated ICP
    • Empty sella – 33.3%
    • Optic nerve dilation – 12.5%
    • Meckel’s Cave Diverticula – 37.5%
ASSOCIATION WITH SUPERIOR CANAL DEHISCENCE

- Superior Canal Dehiscence
  - Communication of superior semicircular canal and middle fossa
    - Symptoms
      - Conductive hearing loss on audiometry
      - Increased bone conduction
        - Ability to hear bodily noise (i.e. eyes moving)
      - Sound and pressure induced vertigo
      - Aural fullness
      - Autophony

PREVALENCE OF SCD

- Temporal bone study
  - Prevalence of 0.5% of ears

- UTSW Spontaneous CSF Otorrhea Patients
  - 5 or 33 ears undergoing middle fossa repair
    - 15.2% prevalence
  - Possible common etiology of spontaneous CSF otorrhea and SCD
    - Congenitally thin skull base with erosion over time
HRCT

INTRAOPERATIVE VIEW OF SCD
MANAGEMENT OF SCD

- Canal plugging in four patients
  - Patients report post-operative vertigo
  - Require vestibular rehab
- Canal resurfaced in one patient
  - No vertigo

CONCLUSIONS

- Spontaneous CSF Otorrhea
  - Most common in:
    - Female patients
    - Sixth and seventh decade
    - Obese patients
  - Increased prevalence of SCD
- Middle fossa approach provides best exposure of middle fossa floor
- Surgical repair successful in the vast majority of cases