Otology Seminar: Spontaneous Cerebrospinal Fluid Otorrhea

2010.09.24 Presented by R3 林彥翰
Introduction

- Cerebrospinal fluid (CSF) otorrhea
  - Presence of CSF within the confines of the temporal bone
  - Defect in the dura → abnormal communication between subarachnoid space and air-containing space of temporal bone
Tegmen tympani

Petromastoid or subarcuate canal

Large apical air cells

Facial nerve canal

Hyrtl’s fissure

Perilabyrinthine fistulas
Translabyrinthine fistulas

Deficiency in the lamina cribosa

Deficiency in the modiolus

Translabyrinthine fistulas
Hyrtl’s fissure (First described in 1936)

- Transient anatomic landmark in the developing fetal temporal bone
- Communication between round window niche (lateral) and pars nervosa of the jugular fossa (medial)
- Tympanomeningeal fissure or hiatus
- Closed by progression of ossification in the 24th week of gestation
External auditory canal

Jugular fossa and where the Hyrtl’s fissure emerges.
Introduction

• The causes of CSF otorrhea
  – Trauma (temporal bone fracture)
  – Iatrogenic (skull base surgery)
  – Neoplastic
  – Infectious
  – Congenital

• Spontaneous CSF otorrhea
  – Not related to the above-mentioned causes
Congenital causes of spontaneous CSF otorrhea

- Mondini dysplasia
- Dehiscent stapes footplate
- Defect in the lateral end of the IAC
- Enlarged cochlear aqueduct
- Defect in Hyrtl’s fissure
- Dehiscent fallopian canal or oval window
Two distinct populations

• Young children:
  – Meningitis after acute otitis media.
  – Congenital anomalies of the temporal bone

• Obese middle-aged or elderly women:
  – Decreased hearing or aural fullness with middle ear effusions
  – Persistent serous or clear discharge after myringotomy
Meningoencephalocele with spontaneous CSF otorrhea
Laboratory tests

• Traditional test: glucose $> 30 \text{mg/dL}$
• Beta-2-transferrin:
  – Only in CSF, perilymph and vitreous humor
  – Greater sensitivity and specificity than glucose test
Imaging study

- HRCT of temporal bone
  - Most common and helpful
  - Determine the location of leaks
  - CT cisternography (intrathecal injection)

- MRI
  - T2-weighted image-->enhanced CSF signal
  - Brain tissues herniation detection
  - Empty or partially empty sella
Pathophysiology of spontaneous CSF otorrhea

- Congenital defect theory (Rao A et al, 2005)
  - Defects of the middle fossa tegmen enlarged (constant CSF pressure)
  - Dural herniation
  - Bony and dural thinning
<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (yr)</th>
<th>Sex</th>
<th>Side</th>
<th>Clinical history</th>
<th>Defect on HRCT</th>
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<tbody>
<tr>
<td>1</td>
<td>46</td>
<td>F</td>
<td>R</td>
<td>SOM/ventilation tube/otorrhea</td>
<td>None</td>
</tr>
<tr>
<td>2</td>
<td>79</td>
<td>F</td>
<td>R</td>
<td>Clear otorrhea through intact TM</td>
<td>Tegmen tympani</td>
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<tr>
<td>3</td>
<td>75</td>
<td>F</td>
<td>R</td>
<td>Meningitis/SOM</td>
<td>PCF bony plate</td>
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<tr>
<td>4</td>
<td>66</td>
<td>F</td>
<td>R</td>
<td>SOM/ventilation tube/otorrhea</td>
<td>Tegmen tympani</td>
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<tr>
<td>5</td>
<td>71</td>
<td>F</td>
<td>L</td>
<td>Tinnitus/SOM/ventilation tube/otorrhea</td>
<td>Tegmen mastoideum</td>
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<td>6</td>
<td>34</td>
<td>F</td>
<td>L</td>
<td>Meningitis/SOM/ventilation tube/otorrhea</td>
<td>Tegmen tympani</td>
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<tr>
<td>7</td>
<td>59</td>
<td>F</td>
<td>Bilateral</td>
<td>Pseudotumor/SOM/BVT/otorhinorrhea</td>
<td>Bilateral tegmen tympani</td>
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<td>8</td>
<td>43</td>
<td>F</td>
<td>L</td>
<td>SOM/ventilation tube/otorrhea</td>
<td>None</td>
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<tr>
<td>9</td>
<td>51</td>
<td>F</td>
<td>L</td>
<td>Rhinorrhea/SOM</td>
<td>Tegmen tympani and tissue mass</td>
</tr>
<tr>
<td>10</td>
<td>55</td>
<td>M</td>
<td>L</td>
<td>SOM/ventilation tube/otorrhea</td>
<td>Tegmen tympani</td>
</tr>
</tbody>
</table>

SOM, serous otitis media; R, right; L, left; TM, tympanic membrane; PCF, posterior cranial fossa; BVT, bilateral ventilation tubes; HRCT, high-resolution computed tomography.
Pathophysiology of spontaneous CSF otorrhea

- Arachnoid granulation theory (Gacek, et al. 1999)
  - Abnormally located arachnoid granulations
  - Autopsy: middle fossa tegmen (21%)
    Posterior fossa (9%)
  - Minor CSF reservoirs. Abnormal locations → decreased return to the venous systems
  - Thinning and erosion of bone
Additional factors involved

A majority of patients

- Obese, middle-aged females.

- A variant of idiopathic intracranial hypertension (IIH)
  (Schlosser et al, 2006)

- Empty or partially empty sella turcica: 63% to 100% of cases
  (5 to 6% in normal population)
  (Friedman et al, 2002 & John et al, 2010)
<table>
<thead>
<tr>
<th>No.</th>
<th>Age, yr</th>
<th>Sex</th>
<th>OSA</th>
<th>BMI (kg/m²)</th>
<th>Side</th>
<th>Findings</th>
<th>Repair</th>
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<tr>
<td>1</td>
<td>46</td>
<td>M</td>
<td>No</td>
<td>24</td>
<td>L</td>
<td>Petrous apex defect</td>
<td>HAC</td>
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<tr>
<td>2</td>
<td>58</td>
<td>M</td>
<td>No</td>
<td>27</td>
<td>R</td>
<td>MC</td>
<td>HAC</td>
</tr>
<tr>
<td>3</td>
<td>71</td>
<td>M</td>
<td>No</td>
<td>27</td>
<td>R/L*</td>
<td>MEC</td>
<td>HAC</td>
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<tr>
<td>4</td>
<td>58</td>
<td>M</td>
<td>No</td>
<td>29</td>
<td>R</td>
<td>MEC</td>
<td>HAC</td>
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<tr>
<td>5</td>
<td>74</td>
<td>M</td>
<td>No</td>
<td>31</td>
<td>R</td>
<td>MEC</td>
<td>HAC</td>
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<tr>
<td>6</td>
<td>70</td>
<td>F</td>
<td>No</td>
<td>33</td>
<td>L</td>
<td>Posterior fossa defect</td>
<td>Fat graft</td>
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<td>7</td>
<td>68</td>
<td>M</td>
<td>No</td>
<td>34</td>
<td>L</td>
<td>Tegmen tympani defect, no MEC</td>
<td>HAC</td>
</tr>
<tr>
<td>8</td>
<td>41</td>
<td>M</td>
<td>Yes</td>
<td>35</td>
<td>R</td>
<td>MEC</td>
<td>HAC</td>
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<tr>
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<td>48</td>
<td>F</td>
<td>No</td>
<td>36</td>
<td>L</td>
<td>MEC</td>
<td>HAC</td>
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<tr>
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<td>54</td>
<td>M</td>
<td>Yes</td>
<td>37</td>
<td>L</td>
<td>MEC</td>
<td>HAC</td>
</tr>
<tr>
<td>11</td>
<td>59</td>
<td>M</td>
<td>No</td>
<td>39</td>
<td>L</td>
<td>Tegmen tympani defect, no MEC</td>
<td>HAC</td>
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<tr>
<td>12</td>
<td>36</td>
<td>M</td>
<td>Yes</td>
<td>50</td>
<td>L</td>
<td>MEC</td>
<td>HAC</td>
</tr>
<tr>
<td>13</td>
<td>69</td>
<td>M</td>
<td>Yes</td>
<td>35</td>
<td>R</td>
<td>Tegmen tympani defect, no MEC</td>
<td>HAC</td>
</tr>
<tr>
<td>14</td>
<td>43</td>
<td>F</td>
<td>No</td>
<td>55</td>
<td>R</td>
<td>MEC</td>
<td>HAC</td>
</tr>
</tbody>
</table>
Demographic data

• Mean ages: 60 years
• Mean BMI: 36.3 kg/m² (Average BMI at US: 28 kg/m²)
• Female predominance (F:M=2:1)
• The same findings noted at CSF rhinorrhea (Schlosser et al, 2006)
• The association with idiopathic intracranial hypertension (IIH) (Schlosser et al, 2008)
# BMI v.s. CSF otorrhea

## TABLE II.

**Body Mass Index (BMI) in Patients With Spontaneous Versus Nonspontaneous Cerebrospinal Fluid Otorrhea.**

<table>
<thead>
<tr>
<th></th>
<th>Spontaneous</th>
<th>Nonspontaneous</th>
<th>$P$</th>
</tr>
</thead>
<tbody>
<tr>
<td>BMI (kg/m$^2$)</td>
<td>35.2 ± 8</td>
<td>28.5 ± 5</td>
<td>.01</td>
</tr>
<tr>
<td><strong>Table 1</strong> Modified Dandy criteria$^2$</td>
<td></td>
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<td></td>
</tr>
<tr>
<td>1. Signs and symptoms of increased intracranial pressure (headaches, nausea, vomiting, transient obscurations of vision, papilledema).</td>
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<td></td>
<td></td>
</tr>
<tr>
<td>2. No localizing neurologic signs otherwise, with the single exception being unilateral or bilateral VI nerve paresis.</td>
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<td></td>
<td></td>
</tr>
<tr>
<td>3. CSF can show increased pressure, but no cytologic or chemical abnormalities otherwise.</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>4. Normal to small symmetric ventricles must be demonstrated (originally required ventriculography, but now demonstrated by CT).</td>
<td></td>
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</tbody>
</table>
Table 2 Criteria for diagnosing idiopathic intracranial hypertension

1. If symptoms present, they may only reflect those of generalized intracranial hypertension or papilledema.

2. If signs present, they may only reflect those of generalized intracranial hypertension or papilledema.

3. Documented elevated intracranial pressure measured in the lateral decubitus position.


5. No evidence of hydrocephalus, mass, structural, or vascular lesion on MRI or contrast-enhanced CT for typical patients, and MRI and MR venography for all others.

6. No other cause of intracranial hypertension identified.
Table 3 Conditions that may produce intracranial hypertension and masquerade as idiopathic intracranial hypertension

Medical disorders
- Addison’s disease
- Hypoparathyroidism
- Chronic obstructive pulmonary disease
- Right heart failure with pulmonary hypertension
- Sleep apnea
- Renal failure
- Severe iron deficiency anemia

Medications
- Tetracycline and related compounds
- Vitamin A and related compounds
- Anabolic steroids
- Corticosteroid withdrawal following prolonged administration
- Growth hormone administration in deficient patients
- Chordecone
- Nalidixic acid
- Lithium
- Norplant® levonorgestral implant system

Obstruction to venous drainage
- Cerebral venous sinus thrombosis
- Jugular vein thrombosis
Pathophysiology of IIH

- Still unknown and multifactorial in nature (Goddard et al, 2010)
  - Central obesity
  - Narrowed cerebral venous sinuses
  - Hormonal dysfunction: association with polycystic ovarian disease (Connor et al, 2008)
- In spontaneous CSF otorrhea:
  - Ophthalmologic and neurologic evaluations
Progression of morbid obesity to encephalocele (Scurry et al, 2007)

- Morbid obesity
  - Increased abdominal pressure
    - Increased thoracic pressure
      - Decreased central venous return
      - Decreased ICP

- Encephalocele
  - Tegmen defect
    - Enlargement
  - Symptomatic benign intracranial hypertension
TABLE 1. *Body mass index as a function of preoperative MRI findings of the sella turcica* 

<table>
<thead>
<tr>
<th>Sex</th>
<th>No. patients</th>
<th>Age, mean ± SD, yr</th>
<th>F/M ratio</th>
<th>BMI, mean ± SD, kg/m²</th>
<th>p (age; BMI)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Female</td>
<td>13</td>
<td>60.8 ± 8.1</td>
<td>N/A</td>
<td>36.5 ± 5.6</td>
<td>0.356; 0.873</td>
</tr>
<tr>
<td>Male</td>
<td>10</td>
<td>59.4 ± 10.0</td>
<td></td>
<td>36.0 ± 8.4</td>
<td></td>
</tr>
<tr>
<td>Race</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Caucasian</td>
<td>14</td>
<td>61.2 ± 8.0</td>
<td>7:7</td>
<td>35.4 ± 8.2</td>
<td>0.216; 0.428</td>
</tr>
<tr>
<td>African American</td>
<td>9</td>
<td>58.7 ± 10.2</td>
<td>6:3</td>
<td>37.7 ± 3.4</td>
<td></td>
</tr>
<tr>
<td>Status of sella</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Normal sella turcica</td>
<td>3 (20%)</td>
<td>61.3 ± 10.7</td>
<td>1:2</td>
<td>28.5 ± 1.4</td>
<td>0.198; 0.034</td>
</tr>
<tr>
<td>Partially empty or empty sella turcica</td>
<td>12 (80%)</td>
<td>62.7 ± 9.0</td>
<td>9:3</td>
<td>38.0 ± 6.7</td>
<td></td>
</tr>
</tbody>
</table>
Conservative treatment

- Indicated for traumatic CSF otorrhea/otorhinorrhea ➔ fracture lines are small in caliber and tortuous
- Successful rates: 82.8% in traumatic CSF leak (Savva, 2003)
- Measures:
  - Restricted nose blowing
  - Avoidance of straining
  - Bed rest and head elevation of 30 degrees
  - Use of antiemetics, antitussives and stool softeners
  - Diuretics and fluid restriction
  - Lumbar drain
Surgical intervention

• Surgery-related or nontraumatic (spontaneous) leaks

• Various approaches
  – Transmastoid & translabyrinthine
  – Middle fossa craniotomy
  – Mini-craniotomy
  – Keyhole craniotomy

• Materials used to correct bony defect:
  – Bone, cartilage, fascia, abdominal fat, silastic and various combinations of autologous tissues.
Surgical intervention

• Transmastoid
  – Preferred approach for most patients (Rao AK, et al. 2005)
  – Extracranial visualization of middle and posterior fossa without damage of intracranial tissues.

• Multilayered Closure:
  – highest rate of definitive closure
  – lowest rate of recurrence
Fig. 3. Multilayered closure for tegmen defects. (By permission of Mayo Foundation.)
Surgical intervention

- Middle fossa craniotomy
  - Tegmen defect larger than 2 cm
  - Multiple defects in the tegmen with extending toward petrous apex

- Translabyrinthine approach
  - For patients with no hearing
  - Remove all middle ear structure
  - Occult Eustachian tube with bone wax, muscle and fascia
  - Obliteration of middle ear with muscle or fat and close EAC
Translabyrinthine approach
Middle fossa craniotomy approach
Fig. 3. Intraoperative view of multiple tegmen defects (case 3). The auricular cartilage of the left ear can be seen at the top, with retracted temporal lobe at bottom of image.
Middle fossa craniotomy approach
Middle fossa craniotomy approach
Keyhole craniotomy
Conclusions

• The diagnosis of spontaneous CSF otorrhea:
  – persistent, unilateral serous otitis media w/ previous normal otologic history.

• High-resolution CT
  – Confirmation of a tegmen defect in 80% of cases
  – Assist in surgical planning

• The transmastoid approach effective if
  – bony dehiscence and/or meningocele can be widely visualized
Conclusions

- Multilayered closure: preferred for most patients
- Patients with spontaneous cerebrospinal fluid otorrhea
  - middle-aged and obese
  - females being affected nearly twice as often as males.
- Empty or partially empty sella
  - 80% of patients with spontaneous cerebrospinal fluid otorrhea on MRI
Conclusions

• An underlying pathophysiologic process w/ anatomic defects (congenital bony dehiscence or aberrant arachnoid granulation)
  – Responsible of most “spontaneous” CSF otorrhea

• Neurology and ophthalmology consultations
  – Necessary to rule out the possibility of idiopathic intracranial hypertension


Thank you for your attention!!