Cerebro spinal fluid (CSF) leaks in ear: Revision of 5 cases

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Abstract: Cerebrospinal (CSF) leaks in the ear are the result of abnormal communication between the subarachnoid space and the tympanic-mastoid cavity, and the etiology in the majority of cases is of trauma (90%). They are of great clinical interest on account of the potential risk of meningitis they pose (4-50%), directly related to the etiology.

Our aim is to present the cases of CSF leaks seen in our hospital over the past 2 years and, at the same time, review and analyze the methods currently used in their management.

A multilayer technique with two or more support materials is essential for a successful surgical repair, producing a success rate of close to 100% when artificial materials are used in combination with multiple layers of autologous tissue.

Key words: Otoliquorrhea/otorrhea. Cerebrospinal fluid leaks in the ear. Mastoidectomy. Antrotomy. Lumbar drainage. Meningitis.

CLINICAL CASES

Clinical case 1

A 60-year-old woman with acetylsalicylic acid (ASA) intolerance, without a personal medical history of interest, attended the clinic after having suffered from hearing loss and tinnitus for a number of weeks. The otoscopy showed that the inner ear was filled with a transudate. The detection of beta-2 transferrin confirmed that this was CSF. In the CT scan a defect was observed at the level of the tegmen antri. The patient was consequently put forward for surgery, which included an antrotomy, and closure of the leak with temporal fascia, lyophilized dura, mastoid cortical bone and Tissucol®. The patient has been asymptomatic for 16 months since surgery.

Clinical case 2

A 67-year-old man with type 2 diabetes, a history of abundant, though not fetid, otorrhea in the right ear which did not improve with normal medical treatment, and hearing loss in the same ear. A posterior tympanic membrane perforation with clear otorrhea could be seen from the otoscopy. The audiometry showed sensory neural hearing loss (50dB) in the right ear. A CT scan determined the erosion of the tegmen tympani due to a cholesteatoma of the right middle ear, as well as the mesotympanum of the left ear being filled by a soft mass without apparent epitympanic or ossicular chain affection. The facial nerve canal was free in both ears. An antrotomy and atticotomy were performed and it was observed that the tegmen antri had eroded with leaking CSF and the attical tissue was inflamed. Surgical closure was achieved using the temporal fascia, Tissucol®, muscle and Surgicel®. The patient has so far remained asymptomatic throughout the year and a half of follow-up.

Clinical case 3

A 46-year-old woman, who had had two cholesteatoma operations on the right ear due to recurrence, had clear liquid leaking from her right ear suggestive of CSF. This was later confirmed by measuring beta-2 transferrin. A CT scan, followed later by a magnetic resonance imaging (MRI) scan, confirmed the presence of a cerebral hernia through the basicranial bone defect (right tempo-tympanic meningoencephalocele). A transmastoid approach was used together with a right temporal craniectomy. Currently the patient has remained asymptomatic for 18 months.

Clinical case 4

A 59-year-old man, whose most significant medical antecedents included snoring, hypertension, vocal cord polypectomy and a head injury 32 years ago. He had been referred by his county hospital having been suffering from hearing loss for 3 years and otoliquorrhea for 2 years, both in the right ear. A perforation of the posteroinferior tympanic membrane was found in the examination. A CT and MRI revealed fullness of the
mastoid air cells. A transmastoid approach was used and the defect closed using Surgicel®, Tissucol®, bone dust, bone and fascia plate. The patient has been asymptomatic for 8 months.

Clinical Case 5

A 71-year-old woman with chronic lymphatic leukemia (CLL) was admitted into the emergency department with symptoms of meningitis and acute otitis media (AOM) in the left ear. Blood cultures were taken and a lumbar puncture performed (both positive for pneumococcus) as well as a CT brain scan which came back as normal. A puncture drainage of the left ear was carried out, and antibiotic (cefuroxime and vancomycin) and corticoid treatment was started. The patient was disoriented, but conscious and apyretic. The emergency department contacted our department. A CT scan of the petrous apices revealed an irregularity in the posterior cortex of the petrous bone touching the posterior fossa on the left side. Following the stabilization of the patient, and with the infectious process under control, a mastoidectomy was performed during which a perforation was seen in the posterior wall of the petrous bone (1 mm) at 1 cm from the anterior wall of the sigmoid sinus. This was closed with muscle, bone wax and Tissucol®. The patient has been asymptomatic for 2 months.

DISCUSSION

The etiology of CSF leaks is predominantly trauma (90%), although in recent studies the surgical etiology has predominated (vestibular neurinoma) as opposed to pure trauma¹. Neurinoma surgery causes CSF leaks in 6-12% of cases, the majority are translabyrinthine, without any connection being found to the surgical technique used or to the size of the tumor, however a link has been found to the patient being over the age of 50². Mastoid surgery for chronic ear pathology (cholesteatoma) is also an etiology worth highlighting.

The longitudinal fractures mainly locate their point of exit in the tegmen, while the transversal fractures leave from the depth of the internal ear canal and internal wall of the tympanic cavity. 80% appear in the first 48 hours post-trauma, with those that appear between 48 hours and 3 months later considered to be of late presentation. There is a 3% risk of meningitis when the leak has lasted less than 7 days, reaching 23-55% in those of more than 7 days’ development³. Others talk of a risk of between 7-30%³. In general, it has not been possible to show that the longer the duration of the CSF leak the greater the percentage of risk of meningitis, but authors such as Leech, Mincy or Spetzler¹⁰-¹² have suggested as much. 85% of the CSF leaks resulting from trauma will close spontaneously in 7 days, 95% will do so in 21 days and only a minimum number of cases will persist for 3 months³.

Non-traumatic etiology constitutes 10% of cases. We can establish two groups depending on whether the intracranial pressure is normal (55%) where congenital etiology dominates (90%) and the group with high intracranial pressure (ICP) where hypophysis adenoma dominates (84%). Both can be perilabyrinthine (less frequent) and/or translabyrinthine. The former are mainly spontaneous and can be separated into two groups: children under the age of 5 with a surgical approach around and through the bony labyrinth who have symptoms of post-AOM meningitis or serous otitis media (SOM) resistant to medical treatment, and adults over the age of 50 in whom the pressure of CSF and the weight of the temporal lobe (arachnoid granulations) generate symptoms similar to those of recurrent, and later
# Table 1

<table>
<thead>
<tr>
<th>AGE</th>
<th>SEX</th>
<th>PERSONAL MEDICAL HISTORY</th>
<th>SYMPTOMS</th>
<th>OTOSCOPY</th>
<th>BETA-2 TRANSFERRIN</th>
<th>CT/MRI</th>
<th>SURGERY</th>
<th>NO. MONTHS ASYMPTOMATIC</th>
</tr>
</thead>
<tbody>
<tr>
<td>60</td>
<td>F</td>
<td>ASA intolerance</td>
<td>Hearing loss Tinnitus</td>
<td>Middle ear filled with transudate</td>
<td>-POSITIVE</td>
<td>CT: defect at the level of the tegmen antri</td>
<td>Antrotomy and closure with lyophilized fascia, mastoid cortical bone and Tissucol®</td>
<td>16 months</td>
</tr>
<tr>
<td>67</td>
<td>M</td>
<td>Type 2 Diabetes</td>
<td>Right ear otorrhea Sensory neural hearing loss (50dB)</td>
<td>Posterior tympanic membrane perforation with clear otorrhea</td>
<td>-POSITIVE</td>
<td>CT: erosion of the tegmen tympani by cholesteatoma, left ear</td>
<td>Antrotomy and atticotomy. Closure with temporal fascia, Tissucol®, muscle and Surgicel®.</td>
<td>18 months</td>
</tr>
<tr>
<td>46</td>
<td>F</td>
<td>2 previous operations for cholesteoma in the right ear</td>
<td>Right ear otorrhea</td>
<td>Clear liquid leaking from right ear</td>
<td>-POSITIVE</td>
<td>CT and MRI: right temporotympanic meningo-encephalocele</td>
<td>Transmastoid approach plus right temporal craniectomy and closure with a temporal Tissucol® fascia, bone dust, bone and Surgicel® plate.</td>
<td>18 months</td>
</tr>
<tr>
<td>59</td>
<td>M</td>
<td>Snoring Hypertension Vocal cord polypectomy Head injury 32 years ago</td>
<td>Hearing loss in right ear Otoliquorrhea</td>
<td>Posterior inferior tympanic membrane perforation</td>
<td>-POSITIVE</td>
<td>CT and MRI: occupation of the mastoid air cells</td>
<td>Transmastoid approach and closure with temporal bone fascia, Tissucol®, bone dust, and bone and Surgicel® plate.</td>
<td>8 months</td>
</tr>
<tr>
<td>71</td>
<td>F</td>
<td>Chronic lymphatic leukemia</td>
<td>Meningitis symptoms (lumbar puncture and blood cultures positive for pneumococcus) AOM of the left ear.</td>
<td>Purulent-looking liquid in left ear</td>
<td>-POSITIVE</td>
<td>CT: irregularity in the posterior cortex of the petrous bone that touches the posterior fossa</td>
<td>Presurgery cefuroxime + vancomycin + IV corticoids. Mastoidectomy with the leak in the posterior petrous apex wall closed 1cm from the anterior wall of the sigmoid sinus</td>
<td>2 months</td>
</tr>
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persistent, unilateral SOM directly related to microdehiscences in the tegmen at the level of the middle cranial fossa. Some 100 cases have been described in the field of spontaneous CSF leaks with children clearly predominating (72%) over adults\(^5\). Other causes of perilabyrinthine leaks are congenital meningoencephaloceles and encephaloceles (20% of meningoencephaloceles), Hyrtl’s fissure, fistulization of the Fallopian tube and the defect of the petromastoid fissure. Translabyrinthine leaks are the type most closely linked to congenital etiology (95%). There are two hypotheses about the genesis of this type of leak: the first talks of abnormal communication in the inner ear caused by a defect in the cribiform plate in the middle and lateral vestibule walls and the second defends abnormal mid-inner ear communication where a central defect at the level of the footplate of the stapes is the most common cause. Congenital leaks appear in patients of an average age of 4\(^4\). The most common symptoms are recurrent meningitis (92%) and sensory neural deafness (86%)\(^4\). They are associated with different malformation symptoms, predominated by Mondini’s deformity or they appear idiopathically\(^4\).

94% of patients have otorrhea and/or rhinorrhea\(^5\). The risk of meningitis is closely connected to the cause (non-traumatic 30%, surgical 21% and cranial trauma 3.5%), presenting greater risk and seriousness (50% mortality) to children than to adults\(^5\). The type of hearing loss can guide us towards discovering the etiology. Hearing loss occurs in 63\(^%-82\(^%\) of patients with a congenital defect; a figure which cannot be calculated for surgery patients.

The physical examination can provide the diagnosis in 90% of cases\(^1\). The halo sign and the Dandy maneuver can be useful\(^6\). A glucose level greater than 30% in CSF is suggestive of a CSF leak. Blood, saliva, tear or serum contamination cause false positives (45-75%), which is why we should give greater value to a negative result (it excludes). Beta-2 transferrin is present in CSF. It is also present in perilymph and vitreous humor, although in a
smaller quantity. It appears in serum in olivopontocerebellar atrophy and in the disialotransferrin deficit. The beta-2 transferrin test is considered to be the most sensitive and specific. The “Nephelometric Assay”, with a sensitivity and specificity comparable to those of the former test, reduces the waiting time to 15 minutes (electrophoretic study: 3-4 hours)⁷,⁸.

Noncontrast high-resolution CT is the standard test for inspecting the temporal bone. Noncontrast high-resolution CT (metrizamide cisternography) is prescribed when the leak is not identified in the coronal CT, it is clinically active, there are multiple bone defects or a bone defect in the CT that does not stand out from the brain parenchyma or if MRI is contraindicated. Up until not so long ago many considered noncontrast high-resolution CT to be the diagnostic procedure of choice for the localization of CSF leaks, especially for active

Figure 6. Diagnostic flowchart.

Figure 7. Flowchart for the handling of CSF leaks

- Lumbar drainage 3-5 days. (Neurological check every hour first 24 hours then every 2 hour. 10cc/hour.
  - If hydrocephalus suspected or ICP: LP or ventricular.
  - If it does not drain, close the drain and 24 hour observation prior to removing drain.
  - Stop drainage
  - Horizontal or slight Trendelenburg position
  - 100% O2
  - Rule out pneumoencephalus (CT, cranial X-ray)
puncture has been kept demonstrated. Surgery is indicated when a lumbar infection. MRI is useful for the localization of CSF (brilliant intermittent or inactive CSF leaks can be as low as 40%).

The prophylactic use of antibiotics has been approved in situations such as: immuno-suppression, drainages or in the presence of penetrating traumas. Lumbar punctures are contraindicated if the CSF leak is not localized, but once it has been localized and obstructive hydrocephalus ruled out its utility where conservative measures have failed, surgery is indicated. Lumbar puncture should be used to confirm or rule out infection. The obliteration of the mastoids, with or without the occlusion of the Eustachian tube, improves the results, especially in postsurgical CSF leaks. The transmastoid approach, together with that of the medial cranial fossa, has almost completely eliminated the posterior approach. The translabyrinthine approach can be used with Mondini deformity (stapedectomy and obliteration of the oval window with soft tissue conserving some level of hearing) and with Hyrtl's fissure. The transmastoid approach enables the bipolar cauterization or the excision of herniated tissues under direct vision and the obliteration with fat when the site of the puncture has not been precisely located. This is indicated if the defect is less than 1 cm in size and if the CSF leak is from acoustic neurinoma with an approach through the posterior cranial fossa (retrosigmoid).

The combined approach (transmastoid and medial cranial fossa) permits greater control over the lesion, especially in large defects, along with greater ease in inserting the packing material. The indications are mainly multiple defects or defects greater than 1-2 cm which may be secondary or not to surgery at the base of the cranium. Vascularized grafts of the temporalis muscle, galeal etc are used.

To conclude we will say that the most important factor for the success of surgery is the use of a multilayer technique with two or more support materials, producing a success rate of close to 100% when the artificial materials (gelita, Surgicel, bone dust) are used in combination with layers of autologous tissue. To prevent recurrences, it is useful to seal the mastoid air cells and the small dehiscences in the tegmen, as the handling of CSF leaks is often linked to the cause of recurrence. A transmastoid approach is generally used with the occasional use of the approach through the medial cranial fossa.

References
5. Hanson MB. Skull Base, CSF otorhea. Temple University Hospital.