

Cerebrospinal Fluid Rhinorrhea and Otorrhea

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Introduction

Cerebrospinal fluid (CSF) rhinorrhea or otorrhea implies an abnormal communication between the subarachnoid space and the nasal cavity or tympanomastoid space. Such drainage can be a dangerous and potentially life threatening occurrence. If suspected, this diagnosis must be either confirmed or excluded as the risk of meningitis is high, with reported rates varying between 2-88%.¹ Unfortunately, such leakage of CSF from its intracranial location may present a significant challenge in the diagnosis, localization, and ultimate management. Thus, otolaryngologists should be aware of the various diagnostic and treatment options available in order to best manage such patients.

CSF Rhinorrhea

CSF rhinorrhea involves a breakdown of all barriers that separate the subarachnoid space from the nasal cavity or paranasal sinuses. Similar to CSF otorrhea, the etiology of CSF rhinorrhea is diverse. CSF rhinorrhea from the anterior cranial fossa may occur after head trauma, ablative tumor surgery, or surgery for paranasal sinus inflammatory disease. Although the incidence of a CSF fistula after endoscopic sinus surgery (ESS) is less than 1%, ESS is a common cause of CSF fistulae.² Blunt trauma to the head is another frequent cause of CSF leaks, which are diagnosed in 3% of all patients with a closed head injury and in up to 30% of patients who have skull base fractures.² Conditions that increase the ventricular pressure, such as intracranial tumors and post-traumatic and post-infectious hydrocephalus are also important causes of CSF leaks. In addition, arachnoid granulations present along the cribriform plate may also lead to spontaneous CSF rhinorrhea.²

The most important factor in its detection is a low threshold of suspicion and this often arises from the history. Any case of unilateral watery rhinorrhea, particularly if increased by posture, should not empirically treated with intranasal corticosteroids but requires further investigation. It is difficult to identify a leak on routine endoscopic exam, but allows the clinician to formulate a differential diagnosis and may identify an intranasal encephalocele.

Testing of nasal secretions

If the patient is able to produce rhinorrhea, often by leaning the head forward, a small sample may be sent for beta-2-transferrin. A small amount is required (0.4 mL) and the patient may supply the sample in a sterile container. This protein is highly specific to CSF and significantly aids in diagnosis. In the past, the diagnosis was aided by the use of glucose and protein determination. The guidelines used in the past was that a definitive CSF leak exists when the glucose content exceeds 0.4 g/l and the protein content is from less than 1g/l up to a maximum of 2 g/l.³

A new modality that may help identify the etiology of intranasal or otologic secretions is the electronic nose. Recent articles have demonstrated promising results in differencing CSF from serum in a small number of cases.⁴

Imaging

Imaging plays a pivotal role in the management of CSF rhinorrhea. The most important step in the management of CSF rhinorrhea is the identification of the site of the leak. A fine detail (1 mm) coronal computed tomography scan (CT) may show small defects or fractures in the region of the anterior skull base or sphenoid

sinus. However, volume averaging across the thin bone in this area may produce both false negatives or false positives. Adding the axial scan to the coronal aids in the diagnosis of posterior frontal sinus wall defects. Congenital dehiscences can occur at any point on the skull base, however the two most common areas for defects producing CSF rhinorrhea are the cribriform niche adjacent to the vertical attachment of the middle turbinate (fovea ethmoidalis) and the superior and lateral walls of the sphenoid sinus.⁵

Administration of intrathecal contrast prior to a high resolution coronal CT scan can provide proof of the true site of the leak especially in cases of active leaking. The use of such CT cisternography is currently the optimal imaging modality for demonstration of the site of a CSF leak in the absence of an obvious skull base defect.⁵

MRI cisternography using highly T2 weighted images is advantageous as the patient does not require any intrathecal contrast. However, the MRI lacks the fine bony detail along the skull base which severely limits its accuracy in localizing CSF leaks. A recent study that utilized a small dose of intrathecal gadolinium improved the ability of MRI to localize the site of CSF egress and appears to be a promising new imaging modality.⁶

Radioisotope cisternography was a popular method of CSF leak identification prior to the development of CT and MRI. Technetium DPTA scans have now been largely abandoned due to both false positive and false negative results and the inability of the technique to show the fine anatomical detail required to locate the site of CSF egress. It is still used in selected cases when the site of the leak has not been clearly demonstrated or when the leak is less active or intermittent. Endoscopically placed intranasal cottonoids are placed in the middle meatus and sphenoidal recess and are removed and measured for radioactivity within 24 hours of injection to help localize the leak. If a leak is detected, most surgeons will administer intrathecal fluorescein and endoscopically examine the area to help identify the exact anatomic locations of the leak.⁷ Identification of the site of the leak is aided by the use of a blue optical filter system introduced into the light source for the endoscopic equipment.

Should the leak not be identified after careful imaging and there remains a strong clinical suspicion, intrathecal fluorescein may be utilized. This is usually combined with an endoscopic approach for repair of the skull base. If a leak is not identified, no surgery is performed.⁷ Care must be taken in administering intrathecal fluorescein as potential complications may result. Complications such as lower extremity weakness, numbness, seizures, and cranial nerve defects have been reported.⁸ Topical fluorescein dye has also been utilized with successful identification of the site of leak as well.⁸

Treatment of CSF Rhinorrhea

Most cases of CSF leaks occurring after blunt trauma or skull base surgery resolve with conservative measures alone. Bed rest, elevation of the head, stool softeners, avoidance of straining, and decreasing CSF pressure with the use of a lumbar drain or daily spinal taps have been utilized as effective conservative options. Prophylactic antibiotics are recommended as the incidence of meningitis is statistically lower in patients receiving antibiotics as compared to those who do not.⁹ Surgical repair is indicated for patients who do not respond to these measures, patients who have traumatic CSF leaks associated with extensive intracranial injury requiring a craniotomy, and patients whose CSF leak is identified intraoperatively.²

Currently there are three types of techniques use to repair CSF leaks: intracranial, extracranial, and transnasal endoscopic repair.¹⁰ The intracranial approach has the advantage of direct visualization of a leak from above and allows treatment of coexisting intracranial pathology. However, it affords poor visualization of communicating fistulas from the sphenoid sinus to the anterior cranial fossa. Success rates with this technique vary from 50 to 73%.¹⁰ This approach has significant morbidity including anosmia, intracerebral hemorrhage, cerebral edema, seizures, frontal lobe dysfunction with memory loss, osteomyelitis of the frontal bone, and possible death. Repair from this route requires 5-7 days in the hospital, a long incision in the hairline, and a prolonged at home recovery period.¹⁰

The extracranial approach utilizes facial incisions to gain access to the site of CSF leak. The main disadvantage with this approach is facial scarring however success rates have been very good with upwards of 80% achieving success in closure of the leak.¹⁰

It is currently accepted that the endoscopic intranasal management of CSF rhinorrhea is the preferred method of surgical repair with higher success rates and less morbidity than the previously described techniques. Most authors agree that the type of endoscopic repair is dependent on the size of the bony versus dural defect. Regardless of bony defect size, small dural defects less than 3 mm may be repaired with a free mucoperichondral graft in an overlay type fashion. Success rates from 83-94% may be expected.⁷ Large dural defects (>3mm) or large bony defects (>2 cm) typically require composite grafts using cartilage or bone in an underlay type fashion and covered with a local flap or free graft.⁷

There is controversy in regards to the use of an overlay versus an underlay type of graft. In a recently published meta-analysis of endoscopic repair of CSF rhinorrhea, both techniques yielded statistically similar results.² Most surgeons utilize gelfoam or gelfilm packing over the repair site to prevent avulsion of the graft during packing removal.² Fibrin glue was found to be used in over half of the cases, which may enhance adhesion of the graft.² Nasal packing was shown to be used in all cases and was usually kept in place for 3-7 days.² Some authors recommend a spinal drain for 3-5 days to reduce CSF pressure, however drains are not necessary in all cases.² In selected cases that do not require a lumbar drain, patients may be discharged home the same day as surgery with strict bed rest, stool softeners, and antibiotics.⁷

A fistula of the sphenoid sinus may be repaired with a free graft technique or with an obliterative technique using abdominal fat.¹¹ Some authors have also used hydroxyapatite cement for repair within the sphenoid sinus as well. Success rates over 85% may be expected in either case in experienced hands.¹¹

Overall success rates with the endoscopic approach is 90% after the first attempt.² A second endoscopic approach may be used to close persistent fistulae with success rates in this setting of around 60% giving an overall success rate of 96%.² Failures with the endoscopic technique may require an open procedure usually performed by experienced neurosurgeons. Thus, close communication with a neurosurgeon is important prior to any proposed repair of CSF rhinorrhea.

CSF Otorrhea

Temporal bone CSF leak is an indication of an abnormal communication or series of communications between the subarachnoid space and the temporal bone. Such leaks may be categorized as either acquired or congenital. Acquired etiologies are by far the most common and include: trauma (temporal bone fractures), postoperative (delayed or immediate), temporal bone infections, and benign or malignant neoplasms. Congenital CSF leak of the temporal bone is most often secondary to Mondini deformities of the osseous and membranous labyrinth. Mondini dysplasia may be associated with dehiscence of the stapedial footplate with abnormal communication between the subarachnoid and perilymphatic spaces. Other rare causes reflect enlarged preexisting bony pathways such as a widened cochlear aqueduct, a petromastoid canal fistula, a patent Hyrtl's fissure (a bony cleft inferior to the round window niche and running towards the posterior fossa), or a widened fallopian canal. The most common type of congenital defect leading to a CSF leak is a deficiency of the bony tegmen of the middle cranial fossa caused by the weight of the temporal lobe and the constant pulsatile nature of the overlying arachnoid granulations in this area. It is with these congenital etiologies that most "spontaneous" CSF leaks occur.¹²

Temporal bone fractures

The most common cause of CSF otorrhea is fractures of the temporal bone. Blunt trauma to the skull may produce fractures in the temporal bone with tearing of dura and foramina causing acute leakage. Fractures may also produce defects in the bony tegmen plate, predisposing one to encephaloceles or meningoceles with resultant delayed CSF leakage. Temporal bone fractures have been traditionally divided into transverse or longitudinal, based on the relationship of the fracture line to the otic capsule and axis of the petrous ridge. In reality, however, most fractures are actually oblique in nature. An important factor in temporal bone fracture classification is whether the fracture passes through the otic capsule. Indications for surgical repair of and the approach for CSF fistula is significantly influenced by otic capsule violation. Tympanic membrane or EAC lacerations are frequently seen in longitudinal fractures which allows for the egress of CSF from the ear. However, with transverse fractures, the tympanic membrane is typically intact and the fluid may build within the

middle ear and mastoid and eventually drain through the eustachian tube producing CSF rhinorrhea. CSF otorrhea in temporal bone fractures usually occurs within minutes of the accident but may be delayed in its presentation if it is draining through the nasopharynx. A high resolution CT scan can demonstrate the course of the fracture line and give information as to the likely site of CSF fistula. Accurate identification of CSF is important. After trauma, CSF otorrhea is typically serosanguinous and can be mistaken for blood byproducts. The fluid should be sent for beta-2-transferrin, as this protein is highly specific to the CSF. As discussed earlier, measurements of glucose and protein in the fluid have fallen out of favor for CSF identification. Bed rest with head elevation, stool softeners, and occasionally the use of lumbar drains is indicated. Sterile cotton should be used to prevent contamination of the ear. Antimicrobial ear drops are unnecessary and may actually confuse the clinician in regards to cessation of CSF flow.¹

In a study by Brodie and Thompson¹, 820 temporal bone fractures were treated over a 5 year period. There were 122 patients with CSF fistulae (97 with otorrhea, 16 with rhinorrhea, and 8 with both). Ninety-five of the patients had the fistulae close spontaneously within 7 days, 21 closed within 2 weeks, and only 5 had persistent drainage over 14 days. Only seven patients underwent surgery for repair of the CSF leak (middle cranial fossa, transmastoid, or combined). Nine of the 121 developed meningitis (7%). The use of prophylactic antibiotics was not statistically correlated with the development or prevention of meningitis in this study. A later meta analysis by the same author, however, did reveal a statistically significant reduction in the incidence of meningitis with the use of prophylactic antibiotics.⁹

Spontaneous CSF Otorrhea (Congenital)

While CSF otorrhea secondary to head trauma or surgery is expected and obvious in most cases, spontaneous CSF otorrhea is often overlooked as it may be subtle and intermittent in nature. Most cases of spontaneous CSF otorrhea are caused by congenital dural defects and are typically divided into two groups. In one type, a preformed bony pathway around and through the bony labyrinth allows the higher subarachnoid pressure to communicate to the middle ear or mastoid. This form of spontaneous CSF leak usually presents early in life, from the ages of one to five years. The clinical presentation is usually meningitis after acute otitis media or as a serous otitis media which is resistant to medical treatment. Often the CSF in the middle ear is first recognized after myringotomy. The second type of congenital defect manifests itself later in life (usually over the age of 50). This is because congenital structures (arachnoid villi) carrying CSF enlarge with age and physical activity as a result of intermittent changes in subarachnoid pressure. This pulsatile pressure and the weight of the temporal lobe is capable of bony erosion over the course of many years. The clinical presentation is usually a unilateral serous otitis media, which at first is recurrent but eventually is persistent. It is hypothesized that the responsible congenital structures are arachnoid granulations which are aberrantly located over a pneumatized part of the skull rather than invaginated in the intracranial venous system enclosed in dura. Undetected arachnoid granulations may be responsible for the increased incidence of meningitis in the adult population older than 60 years.¹³

After elimination of neoplastic causes of unilateral serous otitis media, a search for cerebrospinal fluid otorrhea should be made. A cost efficient approach is to send a sample for beta-2-transferrin (if available) and to proceed with imaging of the temporal bone. High resolution CT is the most useful study in locating the lesion responsible for the spontaneous CSF leak. Because arachnoid granulations are most commonly located in the middle fossa, examination of the tegmen tympani and mastoid with a coronal CT is necessary. The presence of a soft tissue mass near a dehiscence in the bony tegmen is strong evidence for the site of CSF egress. If there is no radiographic evidence of arachnoid granulations or bony dehiscences, the possibility of CSF otorrhea through a preformed pathway should be entertained. Examination for an enlarged internal fallopian canal, patent Hyrtl's fissure, or communication of the IAC with the vestibule (Mondini's) should be evaluated. A contrasted CT (CT cisternogram) is useful in many of these cases.¹³ Recently, magnetic resonance cisternography has gained acceptance as a more noninvasive diagnostic study in determining the site of CSF egress.⁶

Surgical repair is recommended for those cases that do not resolve in order to prevent the morbidity and mortality associated with meningitis. The approach differs for defects in the middle vs. posterior fossa. Middle fossa craniotomy with extradural elevation of the temporal lobe avoids potential damage to the ossicular chain as can occur with a transmastoid approach. Additional arachnoid granulations if present can also be excised. This

approach is useful particularly for large tegmen defects. Smaller tegmen defects may be repaired via a transmastoid approach, however. Posterior fossa arachnoid granulations are best approached with an intact canal wall mastoidectomy approach and fat obliteration of the mastoid. Mondini's malformations can be treated with transmastoid or transcanal obliteration of the vestibule with soft tissue such as muscle or fascia.¹³ Recent articles describing the successful use of hydroxyapatite cement have also been described.¹⁴

Conclusions

The successful management of a patient with a history suggestive of a CSF leak involves ensuring that it is a true leak by testing the fluid for beta-2-transferrin. Imaging studies should be performed in order to anatomically localize the site. Surgery, if necessary, should minimize morbidity while maximizing the chances of a successful outcome. This may be achieved by meticulous preoperative assessment and meticulous intraoperative techniques. Success rates of over 90% can be expected with proper patient and surgical selection.

DISCUSSION

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- Most cases of CNS complications following intrathecal fluorescein injections have occurred because the dose was too high or because the patient had an underlying irritative lesion such as meningitis.
- Pseudomonas is known to survive in fluorescein.
- It is unlikely that intravenous fluorescein gets into the CSF or the perilymph in amounts that would make it clinically useful, but it does stain mucous membranes and skin quite readily and middle ear transudates. If it needs to be used the intrathecal route is perhaps the best route to use.
- If one is attempting to look at fluorescence the ideal system requires an excitation filter, which has a bandpass of 460-500 nm, which makes white light appear blue in conjunction with a barrier filter with a bandpass of 515-620 nm. These however significantly cut the light and it is hard to see through the microscope. We have used an ultraviolet woods lamp instead of the barrier filters in a darkened OR quite effectively.
- A weak solution of fluorescein appears lime green and fluorescent in this light.
- Higher concentrations of fluorescein will not fluoresce but rather appear orange-red due to a phenomenon called quenching.
- When the decision is made to operate on temporal bone fractures due to CSF leaks, one needs to perform a mastoidectomy and our tissue of preference is abdominal free fat graft along with fibrin glue. "One needs to exenterate in order to obliterate." Care is taken to prevent the fat from prolapsing into the additus ad antrum as they may cause a conductive hearing loss if one did not exist before.
- When all else fails for temporal bone fractures, we close off the external auditory canal and the Eustachian tubal orifice in combination with free fat grafts to achieve closure of the CSF leak.
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