Chapter

Cerebrospinal Fluid Leaks and Encephaloceles

Henry P. Barham, Harry E. Zylicz and Christian A. Hall

Abstract

Encephaloceles and cereberospinal fluid (CSF) leaks of the ventral skull base resulting from trauma (surgical and non-surgical), neoplasm, congenital, and spontaneous are a complex problem typically managed by rhinologists/skull base surgeons. Conservative management is often the first step in managing these complex problems. Endoscopic repair of CSF leaks and encephaloceles has greatly evolved with the evolution of endoscopic visualization and instrumentation. Endoscopic repairs of CSF leaks are effective and offer decreased morbidity compared to open approaches with comparative success rates. Meticulous technique is key to success in repair of skull base defects. Materials used are often less important than quality of repair.

Keywords: CSF leaks, Encephaloceles

1. Introduction

Cerebrospinal fluid (CSF) is produced by the choroid plexus in the lateral ventricles, third, and fourth ventricles at a rate of 0.35 mL/min (20 mL/hour or 350–500 mL/day) in the normal physiologic states and is reabsorbed into the dural venous sinuses through the arachnoid villi. The total volume of circulating CSF is 90–150 mL. The entire volume of CSF turns over three to five times per day. Typical intracranial pressure is 5–15 cm H2O and is considered elevated when it is greater than 15 cm H2O. The three layers of the meninges are the dura mater, arachnoid, and pia mater. The dura mater is separated into the superficial layer and the meningeal layers. Common causes of CSF leaks can be divided into trauma, both surgical and nonsurgical, neoplasm, congenital, and spontaneous [1].

An encephalocele is herniation of neural tissue through a defect in the skull base and is defined by the type of tissue that herniates through the defect. A meningocele contains herniated meninges, a meningoencephalocele contains herniated brain matter and meninges, and a meningoencephalocystocele is made up of herniated brain matter and meninges that communicate with a cerebral ventricle.

2. Etiology

2.1 Cerebrospinal fluid leak

The most common cause is nonsurgical (70–80%). 1–3% of acute head injuries result in a CSF leak. Conservative management is often the first step in managing CSF leaks resulting from acute trauma. Seventy percent of leaks close

spontaneously with observation and conservative management which may include bed rest, head of bed elevation, and lumbar drainage. Overall, there is a 30–40% risk of meningitis with conservative treatment [2].

Surgical causes (planned and unplanned) make up a large portion of leaks requiring intervention. Functional endoscopic sinus surgery (FESS) carries <1% incidence of CSF leak. The most common site of skull base injury is the lateral lamella of the cribriform plate. The posterior ethmoid skull base is at greater risk when the maxillary sinus is highly pneumatized in the superior–inferior dimension, which creates a relatively decreased posterior ethmoid height. Neurologic Surgery caries an increased risk albeit typically include planned CSF leak with expected violation of the meninges. Transsphenoidal approach for sellar and suprasellar lesions carry a reported 0.5–15% incidence of CSF leak [3].

Neoplasms can result in CSF leak via direct tumor invasion and/or mass effect leading to intracranial hypertension. Congenital causes result from failure of closure of developmental spaces with resultant herniation of intracranial contents. Foramen cecum is the most common location. Spontaneous leaks are often the result of idiopathic intracranial hypertension (IIH) resulting from decreased CSF reabsorption. Patient characteristics and symptoms often include middle-age, obesity, female, pressure-type headaches, pulsatile tinnitus, and balance dysfunction.

Empty sella syndrome is a radiographic appearance of CSF-filled sella due to flattening of the pituitary gland which is an endocrine gland that resides in the sella turcica and functions to control other endocrine glands by secretion of controlling hormones. Empty sella syndrome can be seen in IIH, which typically affects obese women. Patients typically will present with headaches, pulsatile tinnitus, and diplopia. A hallmark physical exam finding is bilateral optic disc edema secondary to increased intracranial pressure (ICP). Treatment is focused on decreasing ICP with pharmacologic therapy consisting of acetazolamide and furosemide to lower ICP, and headache management, which may include amitriptyline and propranolol. In severe cases with vision problems, surgical intervention may be required, including optic nerve decompression or CSF shunting. Empty sella syndrome can be seen in conjunction with spontaneous CSF leaks.

2.2 Encephalocele

Encephaloceles can occur in both the skull and spinal column. Twenty percent occur within the cranium and 15% of these are associated with the nasal cavity. Nasal encephaloceles are divided into two types: sincipital and basal. Sincipital (anterior and superior) encephaloceles make up approximately 60% of nasal encephaloceles and typically present as a soft compressible mass over the glabella. Basal encephaloceles occur through the skull base more posteriorly and make up approximately 40% of nasal encephaloceles. They may remain hidden for many years because they are located more posteriorly than the sincipital type.

3. Clinical presentation

Clear rhinorrhea that is unilateral, watery, and salty to taste is the most common complaint in CSF leaks. It may run out of the nose in more anterior leaks, or down the back of the throat in more posterior leaks. The drainage can be exacerbated by the Dandy maneuver, which entails tilting the head forward into a chin-tuck position and straining.

Patients with an encephalocele will often present with rhinorrhea or recurrent meningitis and may have a broad nasal dorsum or hypertelorism. Encephaloceles may

characteristically transilluminate, expand with the Valsalva maneuver, and demonstrate a positive Furstenberg sign (enlargement with compression of internal jugular veins). Radiologic imaging including computed tomography (CT) and magnetic resonance imaging (MRI) may be used to evaluate the size and location of encephaloceles.

4. Investigations

The most sensitive and specific test is qualitative β2-transferrin evaluation of the nasal drainage. β2-transferrin is detected in few fluids in the body including CSF, perilymph, and aqueous humor. Only 0.2 mL is needed for an adequate specimen. β2-transferrin has a sensitivity of 97% and specificity of 93%. False positive results can occur with abnormal transferrin metabolism from chronic liver disease, glycogen metabolic disease, and carcinomas; therefore, results should be verified with a negative serum β2-transferrin. β-trace protein is a newer laboratory test with higher sensitivity and specificity which offers faster results than β2-transferrin.

The radiologic evaluation of a CSF leak can be extensive and often begins with a fine cut maxillofacial CT scan to demonstrate bony abnormalities such as defects and fractures. CT is the mainstay for radiologic workup of CSF rhinorrhea with a sensitivity of 92% and a specificity of 92–96%. If the initial imaging does not show an obvious abnormality but suspicion is still high, a CT cisternogram may be useful. This study entails injection of radiopaque material through a lumbar drain into the intrathecal space to help delineate the CSF leak. Presence of contrast within the nasal space or paranasal sinuses indicates a CSF leak. CT cisternography has a sensitivity of 92% with an active leak to 40% with an intermittent leak. MRI cisternography (T2 weighted fast-spin protocol) can be helpful in cases of neoplasm, meningoencephalocele, encephalocele, and in patients with an iodine allergy.

5. Management

In patients who have a traumatic leak and normal CSF pressure, conservative treatment consists of bed rest with head of bed elevation and lumbar drainage of CSF for 5–10 days. With conservative management, there is a reported risk ranging from 7 to 30% of ascending meningitis. The incidence of spontaneous resolution with conservative management is reported to be 70%.

The general consensus among practicing otolaryngologist is that antibiotics should not be used for conservative management unless there is a very large defect with comminuted bone of the skull base as a simple CSF leak carries a 7% infection rate (meningitis, intracranial abscess, cellulitis abscess, and osteomyelitis) and prophylactic antibiotics have not been shown to decrease the risk of infection. After endoscopic repair, antibiotics are generally recommended for 24–48 hours including Cefazolin (1 gm q8), Vancomycin (1 gm q12), or Clindamycin (600 mg q8). This is done to cover possible contamination at the time of surgery in a non-sterile field with concomitant sealing of the sterile to non-sterile flushing of an active leak [4, 5].

6. Surgical Intervention

Endoscopic repair of CSF leaks is effective and offers decreased morbidity compared to open approaches. Meticulous technique is key to success in repair of skull base defects. Materials used and procedures employed are less important than the quality of the repair [6].

Advancements in the endoscopic surgical repair of CSF leaks and encephaloceles have resulted from improvements in instrumentation, visualization, access, and technique. Improved diagnostic imaging and surgical navigation have also improved management. Advancements in endoscopic reconstructive techniques of the skull base including utilization of local vascularized flaps have improved success rates with endoscopic approaches [7].

Compared to open surgery, the endoscopic approach allows for more direct visualization with less manipulation of the surrounding soft tissues. This may allow for a more precise reconstruction of the skull base due to better visualization, and minimal manipulation of nearby neurovascular structures. Compared to the traditional microscopic view, endoscopes give a dynamic operative view with the added ability to see around corners using angled endoscopes. ESBS can avoid scars, decrease hospital stays, and cause less postoperative pain [8].

Not all areas of the skull base can be visualized and safely instrumented via a transnasal endoscopic route. As a general rule, the endoscopic approach should not compromise the ability to achieve the appropriate reconstruction, and crossing major neurovascular structures is not suggested [9].

Common complications from skull base surgery include anosmia and associated taste dysfunction, epistaxis and neurologic complications such as cranial nerve injury. Major skull base surgery complications include CSF rhinorrhea, meningitis, intracranial hemorrhage, orbital complications such as diplopia or vision loss, vascular injury, stroke, and death [8–11].

Intrathecal fluorescein is often used in the surgical repair of CSF leaks. Its advantages include the ability to stain defects that may be more difficult to identify clinically, through the visible dye of CSF to a light green color. The surgeon can also use it to confirm a water-tight repair. It carries a 0% false positive rate. Its disadvantages include a moderate false negative result. It requires a lumbar puncture, and the use of fluorescein intrathecally is not FDA-approved. Rare complications including seizures (0.3%) and death have been reported; however, these have more commonly been associated with administration through a suboccipital puncture. If used to help localize a CSF leak it should be used with caution and should be dosed as 0.05–0.1 mL per 10 kg body weight up to maximum 0.1 mL 10% fluorescein. This is mixed in 10 mL of preservative-free normal saline or CSF. The surgeon should inject slowly (over 5–10 min) without paralytics in the anesthetic regimen to assess for seizure activity. Fluorescein should be avoided in patients with abnormal renal function [12].

The primary goal in endoscopic repair of CSF leaks and skull base reconstruction is to definitively identify all leaks in order to completely reconstruct all defects. After identifying the leak or leaks, the goals of reconstruction are creation of a safe barrier with separation of intracranial and sinonasal spaces and elimination of any dead space. As with any surgical intervention, meticulous surgical technique is paramount for success.

A reconstructive ladder should be used to help determine the type of repair performed. For simple, small (less than 1 cm) defects, a fat plug harvested from the earlobe or abdomen can be used to plug the defect. The next option includes a simple overlay graft harvested from the nasal floor mucosa, turbinate mucosa, or nasal septum. If a more complex, larger reconstruction is in order, a composite (underlay and overlay) graft can be used consisting of an intracranial underlay of bone or cartilage from nasal septum, auricular cartilage or turbinate bone, and an overlay graft of mucosa (free or pedicled) as above. Local pedicled flaps should include the nasoseptal flap, which is supplied by the posterior nasal septal artery, a terminal branch of the sphenopalatine artery. Additional grafts that can be useful in larger defects include temporal fascia or tensor fascia lata grafts. These grafts are

often bolstered in the sinonasal cavity with abdominal fat, a nasoseptal flap or both. In complex situations of extensive defects or poor local tissue, such as in chemoradiated patients, a craniotomy with pericranial flap or free flap reconstruction of the skull base may be necessary.

Lumbar drains are often used to decrease intracranial pressure and thereby reduce the pressure applied to the skull base reconstruction; however, they may be associated with significant morbidity and potential for complications. The use of lumbar drain primarily following a repair varies from different surgeons, and should not be universally utilized in a routine fashion. When used, the duration of drainage is also up to surgeon discretion.

Most reconstructive methods appear to have similar efficacy, and therefore there is no universal "best type of reconstruction." In general, small defects (<1 cm) can be closed in a single layer, and multilayer repair is preferred for larger defects. Some surgeons prefer to use a rigid layer of bone or cartilage to reconstruct the skull base, although this is not required [13]. Vascularized mucosal tissue (e.g., nasoseptal flap) has been demonstrated to improve repair results for larger defects; however, single layer nonvascularized tissue can also be successful in this setting.

Postoperative antibiotics are an important consideration for skull base surgery because of the temporary connection between the intracranial space and external world. Rates of postoperative wound infection following ESBS are approximately 2%, and appear to be higher in open skull base surgery. Broad coverage with IV cephalosporins with or without vancomycin (or oral amoxicillin/clavulanate) is most often recommended. Studies are lacking to support the use of prolonged postoperative antibiotics, although most surgeons prefer to use systemic or topical antibiotics in some form after surgery.

7. Conclusion

A multitude of studies over the past 25 years have shown high success rates of primary repair around 90%, and secondary repair around 97%. These success rates compare favorably to traditional craniotomy approaches with reported success rates between 70 and 80% that carry a higher morbidity profile. Symptoms of failure in reconstruction include clear rhinorrhea and constant postnasal drip. Other signs may include meningitis, severe headaches, seizures, and worsening pneumocephalus.

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