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Nasal cerebrospinal fluid leaks and encephaloceles

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The diverse etiologies of cerebrospinal fluid leaks and encephaloceles make it essential to have a thorough understanding of the underlying pathophysiology, principles of treatment, and treatment options to achieve an excellent outcome. Our current diagnostic studies and preoperative imaging for localization of skull base defects permit accurate identification with minimal operative morbidity. The site of the skull base defect ultimately determines the specific surgical approach. Almost all skull base defects in the anterior skull base are amenable to endoscopic repair. However, external approaches are still mandatory for posterior table defects in the frontal sinus located beyond the reach of frontal sinus instruments. Reconstruction of the skull base defect is dependent on the etiology of the leak and other factors, including the underlying intracranial pressure. This article will highlight the surgical techniques and perioperative care relevant to sinonasal cerebrospinal fluid leaks and encephaloceles.

Most cerebrospinal fluid (CSF) leaks can be broadly classified into traumatic, including accidental and iatrogenic trauma, tumor-related, spontaneous, and congenital. These etiologies influence the size and structure of the bony defect, degree and nature of the dural disruption, intracranial pressure, and meningoencephalocele formation.

Trauma (accidental and iatrogenic)

Traumatic CSF leaks may result from blunt or penetrating trauma. Traumatic disruption of the skull base can create an obvious CSF leak or present years later with meningitis, delayed leak, or encephaloceles. Although conservative treatment is usually attempted with small CSF leaks, there is a reported 29% incidence of meningitis with long-term follow-up of CSF leaks that are treated nonsurgically.[1]

Iatrogenic trauma deserves special mention. Otolaryngologists are the only surgical subspecialty trained to perform sinus surgery. In the age of endoscopic sinus surgery and powered instrumentation, iatrogenic skull base injuries are all too frequent. As otolaryngologists, the ability to handle our complications, such as repairing iatrogenic skull base defects, should be a routine part of our armamentarium.

Idiopathic/spontaneous

Patients with spontaneous CSF leaks frequently have increased CSF pressure; this increases hydrostatic force at the weakest sites of the anterior and central skull base. Spontaneous leaks rarely occur in the frontal sinus but are more likely to occur immediately adjacent to the frontal recess in the ethmoid roof or anterior cribriform plate. Another common area for spontaneous CSF leaks and encephaloceles is in the lateral recess of the sphenoid sinus. Until recently, these latter areas were relatively undocumented.[2-4] These lesions evolve from the herniation of temporal lobe tissue through a middle cranial fossa defect lateral to the foramen rotundum and vidian canal. These patients have excessive pneumatization of the pterygoid process with an attenuated sphenoid sinus recess roof and skull base, which increases the likelihood of defects developing in the floor of the middle fossa.[5]

Increased CSF pressures may contribute to the development of these CSF leaks. The increased CSF pressures seen

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in this subset of patients lead to the highest rate (50% to 100%) of encephalocele formation and the highest recurrence rate after surgical repair of the leak (25% to 87%), compared with less than 10% for most other etiologies.[6-8] Underlay bone grafts in these patients, and those with large skull base defects, are particularly important because these help prevent encephalocele herniation and disruption of the repair. In addition, we recommend lumbar drains and acetazolamide to lower increased intracranial pressure.

Neoplasms

Sinonasal tumors and skull base neoplasms can create CSF leaks directly through erosion of the anterior cranial fossa or middle cranial fossa, or indirectly secondary to therapeutic treatments for the tumor. Persistent malignant tumor after resection and repair will continue to erode the skull base and create a CSF leak. Prior chemotherapy or radiation creates significant difficulties with healing because of poor vascularity of the wound bed.

Congenital

Congenital encephaloceles were initially divided into sincipital (also referred to as anterior or frontoethmoidal) and basal encephaloceles. The basal-type encephaloceles are intranasal in location, and have been variously described as transethmoidal, sphenoethmoidal, sphenomaxillary, sphenorbital, transphenoidal, and transtemporal.[9] In reality, congenital dehiscences can likely occur through any point in the skull base. Although these defects are present since birth, they may not be diagnosed clinically until the patient presents with CSF leak, meningitis, facial deformity, or nasal obstruction.

Diagnosis and preoperative tests

Before any surgical intervention, it is essential to establish firmly the diagnosis of CSF leak to differentiate CSF rhinorrhea from other causes of rhinorrhea. The most commonly accepted method of differentiating CSF from nasal secretions is testing for beta-2 transferrin. This is a very reliable, accurate, noninvasive method to establish the diagnosis of an active CSF leak with a low incidence of false-positives and false-negatives.[10,11] Beta-trace protein is another noninvasive marker that is very specific for CSF and is used most commonly in Europe.[12]

Imaging studies for CSF leaks and encephaloceles generally consist of both coronal and axial computerized tomography to identify any dehiscences in the skull base. These also provide important bony detail that is useful in the surgical approach. The walls of the frontal and sphenoid sinuses have contributions from the skull base in multiple planes. In particular, the unique orientation of the skull base in the sphenoid sinus with both anterior and middle cranial skull base contributing to the superior and posterolateral walls, respectively, absolutely necessitates coronal and axial imaging to evaluate these areas. Unfortunately, the inability to distinguish CSF from other soft tissue limits its diagnostic accuracy. In addition, bony dehiscences may be present without a leak. On the other hand, magnetic resonance imaging or magnetic resonance cisternography identifies brain parenchyma and CSF that have herniated into the sinus very well but is particularly poor at visualizing bony detail.

Although invasive, the injection of contrast medium or a radioactive tracer intrathecally can provide more information. In particular, a computerized tomography cisternogram can be diagnostic and assist in localization of the defect. Radioactive cisternograms are less useful for localizing defects but can localize the side of the leak and identify low volume or intermittent leaks. However, this study is used infrequently because of its invasiveness and low utility.

One of the most useful advancement in diagnosing and localizing CSF leaks has been the use of intrathecal fluorescein with a thorough endoscopic examination. For those patients with an unclear diagnosis, a thorough endoscopic examination after administration of intrathecal fluorescein can be particularly helpful in establishing the preoperative diagnosis. This test is more useful in individuals who have undergone prior sinus surgery and have an exposed, skeletonized skull base. For this reason, fluorescein is more commonly administered intraoperatively, so that complete skeletonization of the skull base may permit accurate diagnosis and localization. We typically use this intraoperatively in all cases because it poses little risk, is useful in localizing the defect, and ensures a watertight closure. If it is not readily apparent where the fluorescein is originating from, thorough skull base exposure must be obtained intraoperatively for evaluation. The fluorescein may be significantly diluted or excreted by the time skull base exposure is attained, depending on the rate of the leak, rate of CSF turnover, and timing of the intrathecal injection. The addition of a blue light filter can improve detection of dilute fluorescein.

Fluorescein is not Food and Drug Administration approved for intrathecal injection because seizures and neurotoxicity have been reported when using higher concentrations or more rapid injections. We have had no complications using a mixture of 0.1 mL of preservative-free 10% fluorescein diluted in 10 mL of the patient's CSF slowly injected over 10-15 minutes. We obtain informed written consent regarding the risks and benefits of intrathecal fluorescein and its lack of Food and Drug Administration approval in all patients. Preoperative tests should be individualized for each patient, and based on the clinical picture and information that is needed. The invasiveness of the diagnostic test and risks to the patient should be considered.

Operative technique

We generally begin each case with rapid sequence intubation to minimize the risk of pneumocephalus from bag mask ventilation. For almost all cases, our neurosurgical service then places a lumbar drain, which can be useful in cases that