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Imaging of Cerebrospinal Fluid Leak

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A cerebrospinal fluid leak from the cranial cavity requires presence of a fistulous communication between the subarachnoid and extracranial space through the skull base. Imaging plays a crucial role in identifying and characterizing the skull base defect and evaluating coexisting pathologies that may alter surgical approach. The authors discuss the epidemiology, imaging, and management of cerebrospinal fluid leak, with particular reference to the imaging modalities and findings.
Semin Ultrasound CT MRI 37:143-149

Introduction

A cerebrospinal fluid leak (CSFL) by definition implies egress of the CSF from the intracranial compartment through the skull base.[1] It is also referred to as a CSF fistula and requires presence of a concordant osseous and a meningeal defect as a prerequisite for the fistulous communication to exist.[1-3] The leaked CSF may manifest clinically as clear nasal (rhinorrhea) or ear (otorrhea) discharge. This is different from spontaneous spinal CSF leaks that often present with headache and symptoms of intracranial hypotension and are beyond the scope of current article.

Initially described by Galen in 200 B.C. as a normal, periodic release of CSF into the nose through the sella and ethmoids, CSF rhinorrhea was considered physiological until Thompson described it as a pathologic entity in 1899.[2,4,5] The most widely accepted classification system, proposed by Ommaya[6] in 1960s primarily divides CSF rhinorrhea into traumatic and nontraumatic categories, where the former may be accidental or iatrogenic, whereas the latter may occur in the setting of normal or elevated intracranial pressures. More recently, a third category of spontaneous CSFL has been added to include patients with no history of trauma or other predisposing factors.[1]

Posttraumatic CSFLs are the most common, accounting for approximately 80%-90% of cases.[3,5,7] These occur as a complication of 2% of all head traumas, are more common with basilar skull fractures and in men in the third to fifth decades.[3,5] CSFLs are also more common with temporal bone fractures, especially when the otic capsule is involved.[8] Approximately 80% of these patients present with rhinorrhea, whereas the remaining 20% have otorrhea.[1] Very rarely, CSFL may present with oculorrhea.[9] Most cases (70%-80%) present in the first week and almost all cases manifest clinically within the first 3 months.[5,10,11] Delayed presentation secondary to wound contraction, scarring or bone necrosis is uncommon but may occur.[3,12]

Iatrogenic trauma, especially during skull base and paranasal sinus surgeries, accounts for approximately 10%-16% of CSFLs.[1,3,13] Functional endoscopic sinus surgery is the most common otolaryngologic culprit, even though the risk of CSFL varies from 0.5%-3%, owing to the sheer frequency with which it is performed.[7,14] From a neurosurgical perspective, transsphenoidal pituitary tumor resection is the most common underlying etiology and carries a risk of up to 15%.[14] CSFL may also complicate septoplasty and otologic or skull base surgeries.[1]

Nontraumatic CSFL associated with normal intracranial pressure (55%) may either have a congenital predisposition such as a Sternberg canal, meningocele, or inner ear anomaly or occur secondary to skull base erosion from tumors or inflammatory processes.[1,8,5] High-pressure leaks (45%) are frequently precipitated by slow growing tumors, often a pituitary adenoma.[5]

Spontaneous CSFLs, by definition are nontraumatic and occur in the absence of any predisposing congenital or neoplastic substrate.[2,15] These most commonly affect obese middle-aged women and share striking epidemiologic and clinical similarities with patients who have idiopathic intracranial hypertension (IIH).[2,3,15,16] Isolated case reports describing evolution of CSFL in the setting of poorly controlled IIH exist, possibly suggesting a causative role of chronically elevated intracranial pressure.[17-19] In fact, some authors argue

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http://dx.doi.org/10.1053/j.sult.2015.12.002
0887-2171/

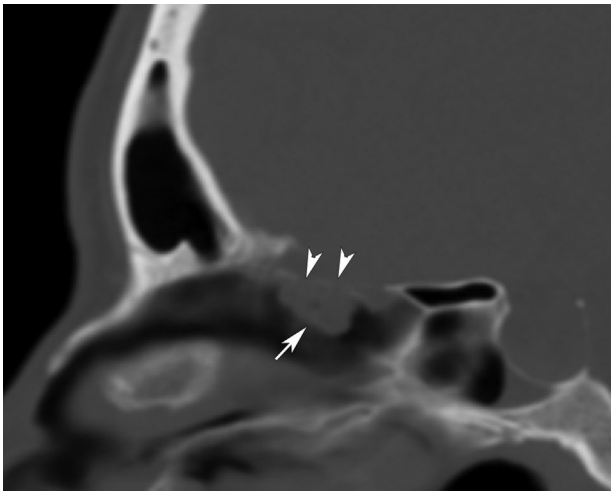

Figure 1 Sagittal multiplanar reconstruction CT in a patient with posttraumatic rhinorrhea shows presence of a defect involving the cribriform plate (arrowheads) along with lobulated, herniating soft tissue (arrow), consistent with a meningocele.

that spontaneous CSFL is part of the spectrum of IIH, as up to 72% of patients with CSFL meet IIH criteria.[15,19,20]

Clinically, the most common presentation is unilateral watery rhinorrhea in the setting of recent head trauma. The rhinorrhea is classically positional, often aggravated with standing or leaning forward and may have a salty or sweet taste.[3,14] Patients may develop pneumococcal meningitis, with overall reported incidence between 4% and 50% and mortality rate approaching 9%.[1,4,5,13,14,21] Patients with temporal bone CSFL may present with rhinorrhea if the fluid travels down the Eustachian tubes or have otorrhea if the tympanic membrane is perforated.[8] Patients with spontaneous CSFL may have headache and visual symptoms secondary to coexisting IIH.[22]

The next step involves verifying the fluid as CSF and is achieved by testing the fluid for β_2-transferrin, a protein produced by desialation of the β_1-transferrin in CSF.[13-15,23] This protein is not present in blood, tears, and ear secretions and has a sensitivity approaching 100% and specificity of 95% for confirming CSF.[13,24] However, the test result may be false positive in patients with coexisting orbital injuries, given its presence in the vitreous humor.[22] Other potential pitfalls are its presence in the serum of patients with alcoholic liver disease or existence of genetic variants of transferrin.[3,14]

Another useful CSF marker is β-trace protein (βTP), normally produced by the choroid plexus and meninges.[8] This protein also has sensitivity and specificity approaching 100% for detecting CSF but may be false positive with chronic renal insufficiency and false negative in meningitis.[3,25,26]

Once a CSFL has been confirmed, the next step involves localizing the underlying defect.[3] Nasoendoscopic localization is often unsuccessful as findings are nonspecific.[3,27] Imaging is therefore critical in localizing CSFL.

Imaging techniques and findings

The primary goal of imaging is to localize the underlying defect.[1,4,14] Other important objectives include characterizing the defect site, evaluating underlying etiology, and assessing coexisting meningoceles.[1,2,7] In patients with more than 1 bony defect, imaging may help localize the site of active CSFL.[1]

High-Resolution Computed Tomography

Even though there is no imaging "gold standard" to evaluate CSFL, high-resolution computed tomography (HRCT) of the basicranium is the most preferred screening modality and is often used initially.[1,3,14,15,27] Advances in imaging techniques, especially the advent of multidetector CT, have allowed faster isotropic volumetric acquisitions at submillimeter thickness.[1,28] This has enabled excellent depiction of multiplanar bony anatomy, obviating the need for acquiring separate coronal images, as with older helical scanners.[1]

Other advantages of HRCT include its noninvasive nature, wider availability, and absence of any major contraindication, unlike magnetic resonance imaging (MRI).[4,16] Additionally, the images may be used for intraoperative guidance during endoscopic repair.[1,3] The primary disadvantage is the use of ionizing radiation, more so in the pediatric age group, although it is lesser with multidetector CT.[14,16] Additionally, the study cannot differentiate actively leaking fistulas from inactive skull defects and is limited in characterizing meningoceles, given the poor soft tissue contrast.[3]

The primary CT finding in CSFL is a skull base bony defect.[1] Posttrauma, this often corresponds to a cribriform plate fracture, given the transition in bone thickness in this region and a tightly adherent dura (Fig. 1).[7,13,14] Defects commonly occur adjacent to the vertical insertion of the middle turbinate or involve the lateral lamella.[1] Fractures of the frontal or sphenoid sinus, sella, or temporal bone may also occur.[1-3,13] These patients have a significantly greater incidence of periorbital hematoma, presumably reflecting a higher risk of occult dural tears.[13] Additional findings may include deviation of crista galli or presence of asymmetric, nondependent soft tissue underneath a rarified anterior skull base.[1] In patients with iatrogenic injury, most common sites of leak are the roof of posterior ethmoid or lateral lamella following functional endoscopic sinus surgery and sphenoid sinus following pituitary surgery.[3,16] Patients with spontaneous CSFL most commonly show bony defects along the anterior ethmoid artery followed less frequently by the inferolateral recess and midline sphenoid sinus posteriorly.[1,2,19,29] Of these, the inferolateral recess defects tend to be the largest and may be associated with arachnoid pits, seen on CT as rounded, scalloped bony defects.[2,15] Patients with spontaneous CSFL frequently show multiple bony defects and generalized skull base attenuation, the latter being statistically significant when compared with general population.[15,30]

Occasionally, presence of focal pneumocephalus may localize underlying fistulous defect (Fig. 2). An air-fluid level within the contiguous sinus or middle ear cavity may be seen.[1] Occasionally, the entire sinus cavity may be opacified. Similarly, unilateral mastoid or middle ear opacification subjacent to a bony defect is sufficient imaging evidence of CSFL in the appropriate clinical context.[1]