Spontaneous encephaloceles of the temporal lobe

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En cephaloceles are pathological herniations of brain parenchyma through congenital or acquired osseous-dural defects of the skull base or cranial vault. Although encephaloceles are known as rare conditions, several surgical reports and clinical series focusing on spontaneous encephaloceles of the temporal lobe may be found in the otological, maxillofacial, radiological, and neurosurgical literature. A variety of symptoms such as occult or symptomatic CSF fistulas, recurrent meningitis, middle ear effusions or infections, conductive hearing loss, and medically intractable epilepsy have been described in patients harboring spontaneous encephaloceles of middle cranial fossa origin. Both open procedures and endoscopic techniques have been advocated for the treatment of such conditions. The authors discuss the pathogenesis, diagnostic assessment, and therapeutic management of spontaneous temporal lobe encephaloceles. Although diagnosis and treatment may differ on a case-by-case basis, review of the available literature suggests that spontaneous encephaloceles of middle cranial fossa origin are a more common pathology than previously believed. In particular, spontaneous cases of posteroinferior encephaloceles involving the tegmen tympani and the middle ear have been very well described in the medical literature. (DOI: 10.3171/FOC.2008.25.12.E11)

K ey W ord s  •  CSF fistula  •  middle cranial fossa  •  neurofibromatosis Type 1  •  temporal lobe encephalocele

C ranial encephaloceles are rare conditions, with a reported overall incidence of approximately 1 in 35,000 people. Although the majority of cases are associated with an initiating process (such as a trauma, neoplasm, inflammatory condition, or previous surgical procedure), these circumstances may also be absent, thus leading to the less common diagnosis of spontaneous encephaloceles of idiopathic origin. Encephaloceles are reportedly most common in the anterior cranial fossa, but they occur in the middle cranial fossa as well. Temporal lobe encephaloceles are defined as pathological herniations of brain parenchyma through dura mater and bone and involve the middle cranial fossa or the lateral temporal cranial vault. Sphenoidal, temporal, and parietal bones form the osseous boundaries of the middle cranial fossa. An anatomical classification of spontaneous temporal encephaloceles based on site of origin has been previously proposed by Wilkins and Radtke in 1993. Several operative reports and surgical case series regarding the treatment of these conditions may be found in the otological, maxillofacial, radiological, and neurosurgical literature. We present a multidisciplinary review on the topic and discuss the diagnosis and management of temporal lobe encephaloceles (Table 1).

Spontaneous temporal lobe encephaloceles are defined as brain herniations of middle cranial fossa origin not associated with traumatic, neoplastic, inflammatory, or iatrogenic conditions. These lesions may become clinically manifest during either childhood or adulthood. Depending on the anatomical location, patient age at presentation, and associated pathological conditions of these lesions, they may present along with signs of occult or symptomatic CSF fistulas, recurrent meningitis, middle ear effusions, otitis, conductive hearing loss, or medically intractable epilepsy.

Classification

Sphenoidal encephaloceles may involve either the body or the greater wing of the sphenoid bone. Defects involving the vertical portion of the greater sphenoid wing may allow protrusion of cerebral matter into the posterior orbital region, leading to the onset of slowly progressive, pulsating proptosis (sphenoorbital or posterior orbital encephalocele). This condition has been described to more commonly affect young women (female to male ratio of 5:3) and patients suffering from NF1. Orbital osseous dysplasia with hypoplasia of both greater and lesser sphenoid wings, deformity of the lateral aspect of the sphenoid body, and/or enlargement of the superior orbital fissure may occur in patients with NF1. In this uncommon clinical subtype of NF1 (cranioorbital-temporal neurofibromatosis), orbital neurofibromas are associated with sphenoorbital encephaloceles and pulsating exophthalmos. Encephaloceles protruding into the infratemporal fossa or pterygopalatine fossa, or toward the lateral wall of the nasopharynx (anteroinferior or transalar encephaloceles) are commonly associated with deficits involving the horizontal portion of the greater sphenoidal wing, laterally to the cranial base foramina of the sphenoidal bone (foramen ovale and rotundum). Such en-
encephaloceles may either remain clinically occult until adulthood, and then present with nonspecific symptoms, or constitute the pathological substrate of a cohort of patients without mesial sclerosis who suffer from medically refractory temporal lobe epilepsy.\textsuperscript{11,15,28,42,44} Osseous defects located in the anteromedial portion of the middle cranial fossa, or more medially in the region of the sphenoidal wing foramina, could lead to the genesis of encephaloceles protruding into the sphenoid sinus or its lateral recess (anteromedial or lateral sphenoidal encephaloceles).\textsuperscript{3,5,6,9,10,11,12,24–27,33,37,40,41,52,54,58} Patients harboring this type of lesion classically present during adulthood with signs and symptoms of CSF rhinorrhea, the subclinical courses of which may lead to recurrent episodes of meningitis and delayed diagnosis.\textsuperscript{13,23,24} Herniations of temporal lobe parenchyma and/or meninges may also involve the posterior temporalis bone at the level of the tegmen tympani (posteroinferior or aural encephaloceles). Although the tegmen tympani remains the more common location for temporal bone encephaloceles, lesions involving the mastoid cavity as well as the petrous apex have been described in the literature.\textsuperscript{34,35,45} Although middle ear effusions, recurrent ear infections, and progressive hearing loss are the most common findings at presentation, cases of medically refractory temporal lobe epilepsy have also been reported in association with posteroinferior encephaloceles.\textsuperscript{31,46,57}

### Pathogenesis

Poorly understood congenital and embryological factors are believed to be involved in the pathogenesis of spontaneous encephaloceles of the temporal lobe, and few theories have been postulated to explain the location and idiopathic nature of such lesions.\textsuperscript{11,15,28} With the exception of its base and the medial portion centered on the foramen rotundum (which ossifies following a cartilaginous stage beginning 8 weeks after conception), the great ala of the sphenoid ossifies in membranes, fusing with the body and the small ala at the end of the 1st year of life.\textsuperscript{43} Up to 19 ossification centers appear to be involved in this ossification process during a normal embryological life.\textsuperscript{11} A developmental problem occurring at an early embryological stage could therefore lead to the formation of a nonossified membranous ala, thus favoring the formation of spontaneous encephaloceles at this level.\textsuperscript{11,28} The incomplete development and fusion of the medial aspect of the sphenoid bone may also lead to the formation of a persistent lateral cranial nasopharyngeal canal, known as the Sternberg canal. This rare cranial base abnormality may then act as the site of origin for congenital encephaloceles protruding into the lateral recess of the sphenoid sinus.\textsuperscript{8} As for the temporal bone, the presence of small areas of dehiscence or bone thinning on and around the tegmen tympani is not an uncommon anatomical finding. This occurrence is, in fact, very well known by neurootologists and skull base surgeons who are familiar with the surgical anatomy of the middle cranial fossa.\textsuperscript{16} Multiple areas of bone dehiscence of the tegmen tympani and petromastoid region have been found in up to 34\% of dissected temporal bones in postmortem anatomical studies, with bilateral deficits described in 20\% of the specimens examined by Kapur et al.\textsuperscript{1,22} Idiopathic meningoencephaloceles of the middle ear occur most frequently around the seventh decade of life,\textsuperscript{4} and Moreano and associates\textsuperscript{32} in an anatomical study involving 1000 human temporal bones observed an inverse relationship between thinning of the temporal bone around the carotid artery and age of the dissected anatomical specimen. A thin bone covering was observed in 17.3\% of temporal bones in specimens 40 years old or older, whereas such thinning was present in only 8.3\% of younger specimens.

Small bone perforations involving the floor of the middle cranial fossa (the so-called “pit-holes”) also constitute known natural variations of the temporal bone anatomy.\textsuperscript{18} Nevertheless, the occurrence of these osseous defects alone does not appear to be sufficient to promote the development of a pathological herniation, as the incidence of meningoceles is far less than the reported occurrence of such bone dehiscences.\textsuperscript{77} Progressive dural and osseous thinning secondary to changes in CSF pressure could also be involved in the genesis of spontaneous encephaloceles and low-normal pressure fistulas in adults, as previously described in the literature.\textsuperscript{23,24} Many factors such as early developmental disorders, osseous anatomical variations, aging and bone remodeling, erosive arachnoid granulations, and low-grade inflammation have all been proposed as factors in the pathogenesis of spontaneous temporal encephaloceles, but the mechanisms involved in the formation of these meningoencephalocele herniations remain controversial.\textsuperscript{19}

### Diagnosis

Prompt clinical diagnosis and correct neuroradiological localization of the site of the fistula/herniation are keys for successful treatment of symptomatic temporal lobe encephaloceles. In the presence of a CSF leak any diagnostic delay may reasonably increase the risk of bacterial meningitis, with a reported incidence of CSF infection that varies from 9 to 50\%.\textsuperscript{18} Recurrent episodes of clinical or subclinical meningitis may also occur, provided that the natural course of a reactive inflammatory response does not always lead to the spontaneous healing of a fistulous tract. Cases that manifest with CSF rhinorrhea may be confirmed by assessing both the levels of glucose and contents of asialo-transferrin (beta-2 transferrin) in the nasal discharge. In patients with normal serum glucose levels,
a concentration of glucose > 30 mg/dl in the collected sample is considered suspicious for a CSF leak. Dextrose sticks, although easy to use and readily available, have a low specificity for detecting CSF in the nasal discharge. The presence of substances that reduce lacrimal and nasal secretions may lead to a false positive result of this test even in the presence of low concentrations of glucose. Therefore, in cases in which nasal discharges can be collected, the measurement of beta-2 transferrin remains the first-line test for the diagnosis of a CSF fistula. This polypeptide involved in ferrous Fe transport may be detected only in the CSF, aqueous humor, and perilymph, and its discovery in a nasal discharge has high specificity and sensitivity for the confirmation of a clinically suspected CSF leak.

Once the diagnosis of a CSF leak has been made, an accurate radiological localization of the site of the fistula should be obtained. Localization of such areas may sometimes be challenging, and this is particularly true in cases of nontraumatic and/or intermittently active CSF leaks. High-resolution CT scans have a low sensitivity for the diagnosis of osseous-dural defects and associated rhinorrhea. Nevertheless, we found the indirect and 3D information provided by this noninvasive modality to be very useful in both preoperative planning and in the selection of a tailored surgical approach for definitive repair. Better accuracy may be obtained using cisternal injection of water-soluble contrast medium and its subsequent radiological visualization at the level of the osseous-dural defect (CT cisternography). Although this modality has a good sensitivity for the diagnosis of active leaks (80–85%), intermittent or inactive fistulas are usually associated with a high incidence of false negative results and low sensitivity. Magnetic resonance imaging with cisternography or constructive interference in steady-state protocols is commonly used in the diagnosis of CSF fistulas, and preliminary results suggest that these flow-sensitive MR imaging studies may have an important role in the evaluation of CSF leaks involving the skull base. Such examinations provide the advantages of noninvasiveness, excellent anatomical details, and the absence of ionizing radiation, and their sensitivity in the identification of the site of fistulas has been reported to be as high as 100%, even in patients with inactive fistulas. We believe that an MR imaging evaluation should also be performed in all cases of spontaneous CSF leaks to rule out associated or predisposing conditions such as hydrocephalus or destructive skull-base neoplasms. At our institution we routinely obtain flow-sensitive MR imaging sequences in conjunction with 3D CT in all patients with a clinical or suspected diagnosis of a CSF fistula. Radionuclide cisternography with the use of intranasal pledgets has been used in the past to confirm the diagnosis of intermittent or slow CSF leaks. Although the information provided by this invasive study may be useful in cases in which a discharge cannot be collected (such as low volume or drainage through the rhinopharynx), its sensitivity for the identification of sites of fistulas remains low, and especially in cases of uncooperative patients, the information provided can be misleading.

### Surgical Treatment

Definitive surgical repair of spontaneous encephaloceles of the temporal lobe is warranted in cases of persistent or recurrent symptoms. In general the appropriate surgical treatment should be selected and tailored according to presenting symptoms, location of the lesion, and presence and type of the CSF leak, as well as the general medical condition and expectations of the patient. In cases of active CSF fistulas, all of the appropriate medical management should be started while completing the neuroradiological workup. Bed rest with head elevation, as well as general precautions to avoid increases in intracranial pressure, is commonly recommended in such cases. Also, placement of an indwelling lumbar catheter for continuous or intermittent CSF drainage may serve the purposes of minimizing the leak at the site of herniation, allowing for the preoperative or intraoperative injection of fluorescein dye, and decreasing the CSF pressure after definitive surgical repair.

The ideal operative approach should allow full exposure and visualization of the encephalocele and its surgical reduction/amputation, as well as definitive repair of the associated osseous-dural defect. Open surgical repairs has been the classic approach to treatment of symptomatic encephaloceles in the past. Anteromedial, anteroinferior, and posteroinferior encephaloceles can be treated using either intracranial or extracranial routes. For open intracranial access, frontotemporal or temporal craniotomies (with the optional addition of a zygomatic osteotomy or zygoma-plasty) can be performed to reach and repair lesions involving either the temporal or sphenoidal component of the middle cranial fossa as well as the lateral extension of the sphenoid sinus (lateral sphenoidal recess). Once the area of herniation has been located, usually by means of an extradural dissection, the final repair may be performed intradurally, extradurally, or by combining the two routes. When possible, we prefer to reduce/amputate the encephalocele extradurally, repairing any osseous defect before considering the addition of an intradural exposure, so as to confine any non-sterile sinus cavity before entering the arachnoidal plane. Although performing a duraplasty with a directly sutured fascial autograft (or any allograft of choice) would be ideal in these situations, such a possibility is hardly allowed, if not precluded, by the anatomical location of the vast majority of temporal encephaloceles.

After the herniated parenchyma has been amputated and hemostasis achieved, the osseous defect may be repaired using fat, local muscle, fascia, calvarial graft, bone chips, or some combination of these. When dealing with anteroinferior or posteroinferior lesions, a vascularized flap of temporoparietal pericranium can also be harvested to cover the osseous repair and act as an onlay graft over the area of dural deficiency (see Illustrative Case). We prefer to avoid splitting the temporal muscle during the repair to minimize the risks of postoperative muscular atrophy and related cosmetic issues. Extracranial procedures such as transfacial approaches, LeFort osteotomies, and midfacial degloving techniques have also been used for the repair of anteromedial and anteroin-
inferior encephaloceles. Although these procedures offer the benefit of avoiding consequences directly related to brain manipulation (such as seizure, contusion, or postoperative brain edema), the possibility of a thorough exploration of the middle cranial fossa may be limited when using these routes, thus restricting their usefulness in cases of unclear preoperative localization of the area involved by the dural defect.

Frontotemporal extradural approaches, with or without posterior orbital osteotomy and/or orbital cranioplasty, may be used to repair sphenoorbital encephaloceles, whereas lateral temporal encephaloceles are usually treated with a standard frontotemporal or temporal craniotomy centered over the osseous-dural defect.

Posteroinferior encephaloceles involving the tegmen tympani can also be treated using a partial mastoidectomy, with or without the addition of a subtemporal approach for a combined intracranial-extracranial repair. Such extradural transpetrosal route allows the combination of both encephalocele repair and surgical toilette of the mastoid, also avoiding the need for brain manipulation. Failure to seal both the dural defect and open air cells during this procedure may lead to recurrent episodes of indolent CSF leaks through the posterior pharynx (via the middle ear and eustachian tube); therefore, an endoscopic examination of the oral cavity should be performed in the early postoperative period in dubious cases.

With the advent of endoscopic approaches to the skull base, the surgical armamentarium for treating basal encephaloceles has greatly expanded. These procedures carry the benefit of minimizing brain manipulation while also avoiding the need for cutaneous incisions. On the other hand, definitive dural reconstruction during endoscopic approaches (using onlay/inlay techniques and/or vascularized flaps) may sometimes be challenging. Although often described as minimally invasive techniques, the extent of anatomical dissection that is sometimes required to expose and repair certain osseous-dural defects by means of endoscopic techniques should not be minimized. Therefore, before considering such routes, surgeons should become familiar with sinus endoscopic anatomy, equipment, surgical techniques, and management of related complications. Specific approaches are dictated by presenting symptoms and the exact location of the encephalocele, with the addition of the careful preoperative evaluation of the patient’s individual sinus anatomy. Within the middle cranial fossa, the anteromedial encephaloceles have been the lesions most commonly approached using endoscopic techniques, mainly due to their proximity to easily accessible sinus routes. These encephaloceles may be repaired via either an endoscopic transnasal transphenoidal or transethmoidal route. Although the surgical anatomy for a direct transsphenoidal approach is familiar to the majority of surgeons, this approach may not always provide complete visualization of the lateral recess of the sphenoid sinus, often the site of the defect. For endoscopic access to this area, a transmaxillary transpterygoid approach has also been advocated. In this case, following a standard endonasal sphenoethmoidectomy, the posterior wall of the maxillary sinus is removed, allowing entrance into the pterygopalatine fossa. The lateral recess of the sphenoid sinus is then entered via direct drilling of the pterygoid process.

In conclusion, a wide array of surgical techniques, such as a combination of open intracranial and extracranial as well as endoscopic approaches, is now available to otolaryngologists and neurosurgeons for the treatment of symptomatic temporal lobe encephaloceles. A thorough understanding of the anatomy of any individual bony defect as well as a meticulous preoperative workup may allow surgeons to provide successful individualized surgical planning, tailoring the appropriate management on a case-by-case basis.

Case Illustration

This 61-year-old woman presented to our institution with clinical symptoms consistent with meningitis. Her medical history showed recurrent episodes of spontaneous rhinorrhea and her neuroradiological workup revealed the presence of a middle fossa osseous dehiscence associated with a lateral temporosphenoidal encephalo-
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Fig. 2. Intraoperative photograph showing the bone-dural dehiscence lateral to V2 after amputation of the encephalocele.

locele (anteroinferior variant; Fig. 1). An ipsilateral temporal mass consistent with an intraparenchymal abscess was also identified. Computed tomography cisternography confirmed the presence of a CSF fistula in the region of the encephalocele. The patient underwent an elective frontotemporal craniotomy with stereotactic aspiration of the abscess, extradural middle fossa exploration, amputation of the encephalocele, and repair of the osseous dehiscence via the transposition of a vascularized temporoparietal pericranial flap (Fig. 2). The rhinorrhea resolved and the patient remains asymptomatic (Fig. 3).

Disclaimer

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

References
