Spontaneous bilateral intrasphenoidal lateral encephaloceles: CT and MRI findings

Ahmet Kemal Firat, MD; Yezdan Firat, MD

Abstract
Lateral intrasphenoidal encephaloceles are rare anomalies. We describe a unique case in which spontaneous bilateral intrasphenoidal lateral encephaloceles were discovered in a 53-year-old man during a work-up for cerebrospinal fluid leak. We discuss our clinical findings and the results of preoperative computed tomography and magnetic resonance imaging.

Introduction
The development of encephaloceles of the skull base can be congenital, spontaneous, or a result of trauma. Most intrasphenoidal encephaloceles are believed to arise in the central aspect of the sinus. Intrasphenoidal encephaloceles—especially those in the lateral aspect of the sphenoid sinus when this sinus has pneumatized extensively into the pterygoid recess—are exceedingly rare. In this article, we describe a particularly unusual case of bilateral temporal meningoencephaloceles that had projected through both lateral walls of the sphenoid sinus. We also review our preoperative findings on computed tomography (CT) and magnetic resonance imaging (MRI).

Case report
A 53-year-old man was referred to our institution for evaluation of intermittent cerebrospinal fluid (CSF) rhinorrhea and frontal headache. He had undergone an endoscopic endonasal procedure and bilateral frontal craniotomy for correction of a spontaneous CSF fistula at another hospital. When he experienced a residual watery nasal discharge postoperatively, he was admitted to our tertiary care unit.

Other than the copious amount of clear rhinorrhea that exuded from both nostrils, no abnormalities were noted on our physical and neurologic examination. Likewise, findings on laboratory testing were within normal ranges.

Noncontrast 3-mm axial and coronal CT of the paranasal sinuses revealed extensive postsurgical changes that were consistent with a bifrontal craniotomy and bilateral ethmoidectomy (figure 1). The multiple defects involved the anterior and posterior walls of the frontal sinuses, the fovea ethmoidalis bilaterally, the cribriform plate, the far-right lateral floor of the sphenoid sinus, and both lateral sphenoid walls. The two defects that involved the right sphenoid wall were both 7 mm long; two other bony defects in the left lateral wall measured 6 and 1 mm. Adjacent to these defects was a significant amount of soft tissue in both lateral recesses of the sphenoid sinus. The soft-tissue lesion was suggestive of an intrasphenoidal mass, although an encephalocele could produce the same findings.

Thin-section high-resolution MRI was obtained to delineate the location of the CSF fistula (figure 2). High T2-weighted (5,200/100/1) axial and coronal spin-echo imaging identified a mass lesion that was surrounded by a CSF-filled sac in the right lateral recess of the sphenoid sinus; the sac projected through the defects in the lateral wall. The lesion was isointense and contiguous with the temporal lobe. Another soft-tissue mass within a CSF-filled sac was seen in the left lateral recess, but there was no evidence of significant temporal lobe herniation. A small amount of brain tissue extended through the defect. These findings were consistent with the presence of bilateral intrasphenoidal lateral encephaloceles with prominent temporal lobe herniation into the lateral recess on the right side.

Bilateral frontotemporal transcranial exploration and successful multilayered dura and bone repair were performed. At the 2-month follow-up, no residual CSF leak was noted.

Discussion
A meningoencephalocele is a gap in the skull that results in a herniation of the adjacent meningeal and brain substances. Meningoencephaloceles are classified according to their location as either frontal, occipital, parietal, or basal. Basal meningoencephaloceles are further subclassi-
Figure 1. A: Noncontrast 3-mm axial CT demonstrates the bony defect (arrow) in the right lateral wall of the sphenoid sinus. The encephalocele largely fills the right lateral recess. B: Another axial image shows the bony defect (arrow) with well-defined margins in the left lateral wall of the sphenoid sinus and the soft-tissue density in the left lateral recess. C: Coronal CT demonstrates the bilateral bony defects (arrows) and the projecting encephaloceles that fill both lateral recesses.

Figure 2. A: Coronal, fat-saturated, T2-weighted (5,200/100/1) cisternographic MRI shows the defect in the lateral wall of the left sphenoid sinus (arrowheads) that contains CSF and projecting cerebral tissue (arrow). B: A mass (arrow) is seen extending through the bone defect in the right lateral wall of the sphenoid sinus and into the lateral recess, including the temporal lobe and surrounding CSF cavity. C: An irregular soft-tissue mass (arrow) that is hyperintense to brain is seen in the encephalocele.

ified on the basis of the location of the bone defect as either transsphenoidal, transethmoidal, sphenoethmoidal, or sphenoorbital.1

Abiko et al further distinguished two types of transsphenoidal meningoencephalocele: intrasphenoidal and true transsphenoidal.2 Intrasphenoidal encephaloceles extend into the sphenoid sinus and are confined by its floor. True transsphenoidal encephaloceles traverse the floor of the sphenoid sinus and protrude into the nasal cavity or nasopharynx.

Intrasphenoidal encephalocele is a rare entity. It is classified according to its location as either a perisellar type or a lateral-recess type.3,4 The perisellar type extends into the sphenoid sinus through the central roof, while the lateral-recess type extends through the lateral wall of the sphenoid sinus. The lateral-recess type is extremely rare and is associated with herniations of the temporal lobe. Only 3 cases of bilateral lateral-recess intrasphenoidal encephaloceles have been reported in the literature.5

The clinical presentation of intrasphenoidal encephalocele varies. According to Lai et al, intrasphenoidal meningoencephaloceles usually present with CSF rhinorrhea. The clinical history may also include headache, seizures, vertigo, meningitis, and brain abscess.3

Several imaging features aid in the preoperative characterization of intrasphenoidal encephaloceles and subsequent management. Fine-cut CT of the anterior and middle fossa may reveal the location of the bony defect. Although CT can clearly identify bony defects with the aid of a bone algorithm, it cannot as easily distinguish herniating brain tissue from mucosal thickening, a retention cyst, or a potential aneurysm of a dehiscent cavernous carotid artery.

MRI provides valuable information on detecting brain tissue and dural herniation. The most important feature is its ability to identify a stalk of soft tissue traversing the bone defect from the temporal lobe to the sac of the encephalocele. Multiplanar imaging allows for the detection of soft-tissue continuity and provides the ability to view the gyral pattern; the characteristic signal intensity of the herniated brain makes MRI the ideal imaging method for diagnosis and surgical planning. MRI is also the best method for identifying intracranial anomalies associated with meningoencephalocele: holoprosencephaly, Dandy-Walker malformation, aqueductal stenosis, agenesis of the corpus callosum, and other midline abnormalities.7 MRI is superior to CT in defining brain herniation. On the other hand, the appearance of gliotic herniated brain accompanied by other pathology may limit the accuracy of MRI.

Treatment of intrasphenoidal encephaloceles must be
directed toward two primary goals: the prevention of CSF leakage and the repair of the middle fossa defect. The goal of middle fossa repair is to diminish the risk of ascending meningitis by separating the nasal and cranial contents. Without repair, patients with CSF rhinorrhea are at an increased risk of ascending meningitis or brain abscess.8 According to the literature, most lateral-recess intrasphenoidal encephaloceles have been repaired endoscopically via a transsphenoidal approach.3,9,10 The preferred means of access to encephaloceles located in the lateral recess of the sphenoid sinus is via the endoscopic transpterygoid approach. In this approach, the most lateral portion of the sphenoid sinus is accessed through the posterior wall of the maxillary sinus and the pterygopalatine fossa.11 Bolger and Osenbach reported that this technique provided good control of CSF rhinorrhea.11 However, Landreneau et al argued that the endoscopic approach to repair is not safe because it fails to provide access to the lateral recess of the sphenoid sinus. They reported that the CSF fistulae of the lateral sphenoid sinus associated with temporosphenoidal encephaloceles are different from other types of sphenoid fistulae and that they can be repaired safely and reliably via a direct middle cranial fossa approach. Landreneau et al recommended exploration of the anterior floor of the middle cranial fossa in order to safely disconnect the temporosphenoidal encephalocele and to achieve adequate multilayered repair of dural and bony dehiscence under direct visualization. They recommended that this procedure be used until endoscopic technology proves to be consistently reliable in allowing for direct visualization and repair of lateral encephaloceles and dural and bone defects. Radiologic imaging is often undertaken in such cases, primarily to assess the location of a CSF leak and to further evaluate the pathology. We recommend fine-cut coronal and axial CT to evaluate bony defects and to provide a road map for focusing heavily T2-weighted coronal MRI at the possible leakage site on the base of the skull.

As occurred in our patient, an encephalocele can be the site of a CSF leak. MRI was able to provide three-dimensional images of the encephalocele that were useful for both diagnosis and surgical planning. As is the case with any meningoencephalocele, our diagnosis was based on the identification of a sac that was directly contiguous with the subarachnoid space and that contained a component of herniated cerebrum. We conclude that to plan the safest possible repair, preoperative evaluation of intrasphenoidal meningoencephalocele by MRI is essential to confirming the extension of the lesion and any associated abnormalities.

References