

Middle Fossa Approach to a Temporosphenoidal Encephalocele

—Technical Note—

Jae Eun KWON and Ealmaan KIM

*Division of Skull Base Surgery, Department of Neurosurgery, Dongsan Medical Center,
Keimyung University School of Medicine, Daegu, Republic of Korea*

Abstract

Temporosphenoidal encephalocele (TSE) is a rare entity caused by herniation of the anteromedial temporal lobe into the sphenoid sinus (SS) through a middle fossa (MF) defect. A 45-year-old woman presented with a spontaneous TSE manifesting as a 4-year history of recurrent cerebrospinal fluid rhinorrhea and meningitis. Coronal computed tomography showed a skull defect in the superior wall of the right lateral recess of the SS. This homogeneous intrasphenoidal lesion appeared hypointense on T₁-weighted magnetic resonance (MR) imaging and hyperintense on T₂-weighted MR imaging. The patient underwent a frontotemporal craniotomy and extradural MF exploration. The encephalocele was amputated and the temporal base dura primarily sutured and reinforced with fat graft. The MF hole was plugged with temporalis fascia and a calvarial graft layered over the bone defect. Histological examination confirmed meningoencephalocele. Rhinorrhea resolved and the patient remained asymptomatic. Resection of an anteromedial TSE and closure of the craniodural defect in the MF floor via a less invasive extradural skull base approach is effective.

Key words: intrasphenoidal encephalocele, cerebrospinal fluid rhinorrhea, meningitis, skull base surgery

Introduction

Sphenoidal encephaloceles caused by temporal lobe herniation through a middle fossa (MF) defect into the sphenoid sinus (SS) are rare clinical entities.⁵⁾ Sphenoidal encephaloceles can be subdivided on the basis of the defect origin into the median type, arising from the superior or posterior wall of the SS, and the lateral type, arising from the lateral wall of the sinus.²²⁾ This anomaly can extend to the SS (intrasphenoidal encephalocele), and traverse the sinus floor and reach the epipharynx (transsphenoidal encephalocele). The so-called temporosphenoidal encephalocele (TSE) is considered to occur lateral with an osseous defect located in the anteromedial portion of the MF.²⁴⁾ Surgical treatment of sphenoidal encephaloceles is not simple in adults, and surgeons can select either intracranial or extracranial routes.^{12,25)}

We report a case of a spontaneously occurring TSE manifesting as cerebrospinal fluid (CSF) rhinor-

rhea and meningitis and treated through a small pterional window and MF extradural dissection. This modified technique provides safe access to deep-seated basal lesions and simple repair of dural-bony dehiscence of the temporal fossa.

Case Report

A 45-year-old female with a history of spontaneous CSF leak presented with persistent headache located around the orbit, forehead, and cheek. She also noted some ringing sensation in her right ear. Her history revealed an episode of meningitis 3 years previously. At that time, CSF was actively leaking from her nose. She described episodes of clear fluid draining from the right nasal passage, especially on arising in the morning, for 2 weeks prior to admission. Evaluation of organ systems found no other abnormalities, and endocrine tests were normal. Rhinoscopy was utilized to evaluate the sinonasal cavities, mucosa, sinus ostia, and turbinates. No intranasal masses were observed. Computed tomography (CT) of the sinuses revealed a circumscribed soft tissue mass within the well pneumatized

Received July 8, 2009; Accepted September 16, 2009

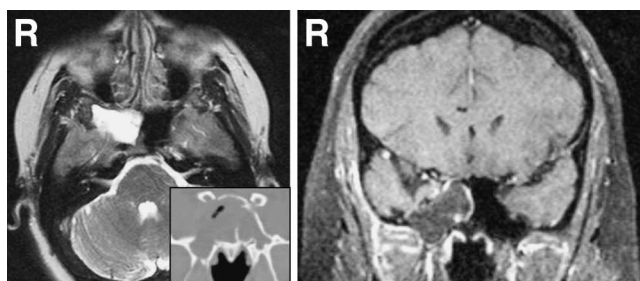


Fig. 1 Preoperative axial and coronal magnetic resonance images revealing a localized well-defined cystic lesion in the temporal fossa protruding into the right sphenoid sinus, which appears hyperintense on the T₂-weighted image (left) and hypointense with rim enhancement on the T₁-weighted image (right). The inset shows bone erosion of the lateral wall of the sphenoid sinus and an air-fluid interface.

SS and a bony dehiscence in the lateral aspect of this sinus. No other evidence of skull base defect was seen. Magnetic resonance (MR) imaging of the brain clearly showed herniation of brain tissue through the bony defect in the MF floor, with moderate wall enhancement after gadolinium injection. Empty sella was also identified (Fig. 1). Treatment options included endoscopic transnasal repair versus minimal craniotomy and repair from above. We determined that this TSE could be repaired via an extradural technique with repair of the associated dura and bone.

A lumbar drain was inserted and the patient secured in the supine position with the head tilted by 45 degrees. A curvilinear scalp frontotemporal area incision was made. The scalp flap was elevated interfascially to protect the frontalis nerve, thus exposing the MF base. After reflecting the temporalis muscle flap, a small frontotemporal craniotomy was fashioned in the standard manner. Inferiorly, the craniotomy was drilled down to the MF floor. Dural tack-up sutures were applied along the craniotomy superior edge. At this point, the lumbar drain was opened to reduce the required retraction of the temporal dura. The medial sphenoid ridge and the posterior orbital roof were resected and the superior orbital fissure was uncovered. Under the operating microscope, epidural dissection was undertaken in anterior to posterior manner starting at the superior orbital fissure. The middle cranial base was extradurally exposed anteriorly and posteriorly. The middle meningeal artery was cauterized and divided above the foramen spinosum. The procedure then moved inward with dural elevation and dissection proceeding anteriorly and medially until the foramina of ovale and rotundum were exposed. The lateral

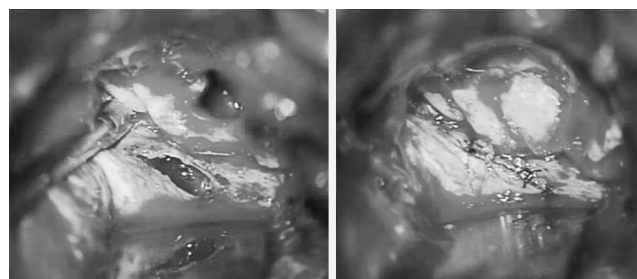


Fig. 2 Operative photographs of the right-sided middle fossa approach to the temporosphenoidal encephalocele. The dural-bone dehiscence is located in the anteromedial portion of the middle cranial fossa, lateral to the cavernous sinus (left). After amputation of the encephalocele, the temporal dura is sutured primarily and the calvarial bone graft is layered over the basal defect (right).

wall of the cavernous sinus was partially elevated free from the trigeminal roots and ganglion. A self-retaining retractor was then applied. The second division of the trigeminal nerve was then identified. A thin-walled, CSF-filled cyst was observed medial to the foramen rotundum. Bone around the cyst was drilled to fully expose the cyst, and the lesion was removed to the fullest extent possible. The herniating tissue was loosely attached to the surrounding basal dura and cranium, and appeared partly firm and lobulated, indicating the presence of glial tissue. The encephalomeningocele extended extracranially between the ophthalmic and maxillary divisions of the trigeminal complex and entered the SS. A bony dehiscence of approximately 10 mm in diameter was found in the anteromedial portion of the MF. After dissecting and amputating the stalk completely, the anterior temporal dura was resected out of the encephalocele. The dural defect of the temporal lobe was closed with sutures and reinforced with fat tissue. The hole in the MF was packed with muscle and temporal fascia, and the osseous defect was repaired using a bone chip harvested from the calvarium (Fig. 2). Tissue glue was applied to seal and fix the newly-developed barrier in the MF. Hemostasis was ensured and the bone flap was replaced in standard fashion. The temporalis muscle and scalp were then closed, and the lumbar drain removed at completion.

Histological examination of the resected tissue revealed encephalomeningocele. Neuroglial tissue was identified in the subepithelial tissue in several fragments. Chronic inflammatory infiltration was sparse. The patient fully recovered from the pterional craniotomy and MF dissection to repair the TSE. No recurrence of CSF leakage or meningi-

tis had occurred 3 years after surgery.

Discussion

Sphenoidal encephaloceles are rare forms of congenital basal encephaloceles and can be subdivided into the intra- and trans-sphenoidal types. According to the current classification and depending on the defect location,⁷⁾ these developmental anomalies seem to represent a specific clinical entity, so should be differentiated from transtethmoidal, sphenothmoidal, frontosphenoidal, and sphenoorbital encephaloceles. Furthermore, intrasphenoidal encephalocele, in which the herniating brain and meninges are enclosed within the SS, has different morphological and clinical characteristics and thus appropriate surgical treatment compared to true transsphenoidal encephaloceles.²¹⁾ TSEs are a rare subgroup of lateral sphenoidal encephaloceles, probably developmental in origin, and have been described as a separate entity.²⁾

Few cases of intrasphenoidal encephalocele have been reported.^{1,4,5)} Therefore, diagnosis and treatment is a challenge for the rhinologist and skull base neurosurgeon. The more common trans-sphenoidal encephalocele can occur through a larger defect in the sellar floor, planum sphenoidale or posterior ethmoid.^{5,21)} Intrasphenoidal encephalocele may in general be immediately apparent in infants who present with cranial-facial midline defects and endocrine dysfunctions. In contrast, all eight previous cases of intrasphenoidal encephalocele occurred in middle-aged patients presenting with CSF rhinorrhea.^{14,16)} Although the pathogenesis remains uncertain, basal temporal encephaloceles are thought to arise either from defective embryological development of the skull base or from postnatal enlargement of small bony dehiscences on the MF floor caused by CSF and brain pulsations.³⁾ Many adult cases were associated with empty sella turcica, and association with other intracranial anomalies was uncommon.^{24,25)} In the present case, the evidence of empty sella supported our opinion that our patient most likely experienced benign intracranial hypertension, which put her at risk for the development and recurrence of spontaneous CSF leaks from TSE.

The initial clinical presentation of CSF rhinorrhea was consistent with intrasphenoidal encephalocele. The clinical picture is then often complicated by recurrent meningitis, chronic headache, orbital pain, cranial neuropathy, and subdural hematomas.^{11,19)} Preoperative CT cisternography often reveals that all fistulae involve lateral extension of the SS into the floor of the MF. Modern MR imaging

can aid the clinician in discriminating between intrasphenoidal encephalocele and more common causes of isolated SS mass such as polyp, mucocele, fungal ball, and localized sinusitis.^{7,8)} MR imaging may also provide valuable information about the location of vital structures within the herniated tissue of the basal encephaloceles.^{9,10)}

The goals of surgery for basal encephaloceles include: removal of the herniated sac, preservation of functional neural tissue, closure and reconstruction of the defect with healthy tissue, and restoration of the CSF pathway via shunting. The need for surgical repair of intra- and trans-sphenoidal types of encephaloceles is indicated by the presence of persistent CSF rhinorrhea, epipharyngeal respiratory obstruction, or progression of neurological deficits.²²⁾ To select the optimal surgical approach for repair of the skull base defect in patients with sphenoidal encephaloceles, careful preoperative evaluation and localization of the sphenoid defect are essential. Transcranial reduction and repair has previously been regarded as the therapy of choice for transtethmoidal encephaloceles, but the feasibility for transsphenoidal encephaloceles remains controversial, particularly in pediatric patients, because of the high morbidity and mortality rates associated with cranial surgery.^{15,23)}

Adult patients with true transsphenoidal meningoencephalocele should not undergo surgery, in view of the complex anatomy and the slow progression of symptoms.¹⁴⁾ Adult TSEs can be repaired via extradural MF surgery from above or via an endoscopic transnasal approach from below, but the surgical route has to be tailored and selected according to the size and location of base defect, status of SS pneumatization, CSF pressure, recurrence of CSF leaks, type and failure of prior treatment, and the experience and preference of the surgeon. Some cases of TSEs have been successfully treated via transsphenoidal or transtethmoidal routes. However, surgical failures related to this midline approach could be due to lack of control of the lateral recess of the SS.^{13,17,18)} Therefore, the frontotemporal basal approach with extradural and/or intradural exploration seems more effective, but a transnasal endoscopic approach will most likely be used in the future. A new surgical procedure, extended endoscopic transpterygoid approach performed by an expert otolaryngologist may be useful in selected instances within the lateral SS.^{6,20)} Transcranial and transnasal repair of sphenoidal encephaloceles followed by a shunting procedure for high pressure can abolish CSF leakage.¹⁶⁾

The present case shows that surgical treatment of CSF rhinorrhea secondary to MF encephalocele as

sociated with lateral extension of the SS differs from the surgical strategy for more medial sphenoidal fistulae. Fistulae involving the lateral recess of a widely pneumatized SS require a transcranial approach for direct visualization and obliteration of the defect, whereas fistulae involving the central portion of the sinus may be successfully obliterated transsphenoidally.

Anteromedial meningoencephaloceles of the MF are rare. A middle-aged woman who presented with CSF leaks underwent MF epidural surgery through a miniature frontotemporal craniotomy to expose and amputate the TSE involving the lateral SS. This simple and safe skull base approach is the most suitable approach for patients with TSE to resect the herniated sac and reconstruct a water-tight barrier in the temporal fossa.

References

- 1) Abe T, Lüdecke DK, Wada A, Matsumoto K: Transsphenoidal cephaloceles in adults. A report of two cases and review of the literature. *Acta Neurochir (Wien)* 142: 397–400, 2000
- 2) Albernaz MS, Horton WD, Adkins WY, Garden PD: Intrasphenoidal cephalocele. *Otolaryngol Head Neck Surg* 104: 279–281, 1991
- 3) Alfieri A, Schettino R, Taborelli A, Pontiggia M, Reganati P, Ballarini V, Monolo L: Endoscopic endonasal treatment of a spontaneous temporosphenoidal encephalocele with a detachable silicone balloon. Case report. *J Neurosurg* 97: 1212–1216, 2002
- 4) Bikmaz K, Cosar M, Iplikcioglu AC, Dinc C, Hatiboglu MA: Spontaneous cerebrospinal fluid rhinorrhoea due to temporosphenoidal encephalocele. *J Clin Neurosci* 12: 827–829, 2005
- 5) Blaivie C, Lequeux T, Kampouridis S, Louryan S, Saussez S: Congenital transsphenoidal meningocele: case report and review of the literature. *Am J Otolaryngol* 27: 422–424, 2006
- 6) Bolger WE: Endoscopic transpterygoid approach to the lateral sphenoid recess: surgical approach and clinical experience. *Otolaryngol Head Neck Surg* 133: 20–26, 2005
- 7) Buchfelder M, Fahlbusch R, Huk WJ, Thierauf P: Intrasphenoidal encephaloceles—a clinical entity. *Acta Neurochir (Wien)* 89: 10–15, 1987
- 8) Clyde BL, Stechison MT: Repair of temporosphenoidal encephalocele with a vascularized split calvarial cranioplasty: technical case report. *Neurosurgery* 36: 202–206, 1995
- 9) Deasy NP, Jarosz JM, Al Sarraj S, Cox TCS: Intrasphenoid encephalocele: MRI in two cases. *Neuroradiology* 41: 497–500, 1999
- 10) Firat AK, Firat Y: Spontaneous bilateral intrasphenoidal lateral encephaloceles: CT and MRI findings. *Ear Nose Throat J* 83: 831–833, 2004
- 11) Fraioli B, Conti C, Lunardi P, Liccardo G, Fraioli MF, Pastore FS: Intrasphenoidal encephalocele associated with cerebrospinal fluid fistula and subdural hematomas: technical case report. *Neurosurgery* 52: 1487–1490, 2003
- 12) Gürkanlar D, Akyuz M, Acikbas C, Ermol C, Tuncer R: Difficulties in treatment of CSF leakage associated with a temporal meningocele. *Acta Neurochir (Wien)* 149: 1239–1242, 2007
- 13) Herman P, Sauvaget E, Guichard JP, Tran Ba Huy P: Intrasphenoidal transsellar encephalocele repaired by endoscopic approach. *Ann Otol Rhinol Laryngol* 112: 890–893, 2003
- 14) Jabre A, Tabaddor R, Samaraweera R: Transsphenoidal meningoencephalocele in adults. *Surg Neurol* 54: 183–188, 2000
- 15) Kennedy EM, Gruber DP, Billmire DA, Crone KR: Transpalatal approach for the extracranial surgical repair of transsphenoidal cephaloceles in children. *J Neurosurg* 87: 677–681, 1997
- 16) Kumar R, Jenkins A: Spontaneous CSF rhinorrhoea due to temporosphenoidal encephalocele. *Br J Neurosurg* 12: 66–68, 1998
- 17) Lai SY, Kennedy DW, Bolger WE: Sphenoid encephaloceles: disease management and identification of lesions within the lateral recess of the sphenoid sinus. *Laryngoscope* 112: 1800–1805, 2002
- 18) Landreneau FE, Mickey B, Coimbra C: Surgical treatment of cerebrospinal fluid fistulae involving lateral extension of the sphenoid sinus. *Neurosurgery* 42: 1101–1104, 1998
- 19) Myssiorek D, Cohen NL: Intrasphenoidal meningoencephalocele: a case report. *Am J Otolaryngol* 8: 391–394, 1987
- 20) Sautter NB, Batra PS, Citardi MJ: Endoscopic management of sphenoid sinus cerebrospinal fluid leaks. *Ann Otol Rhinol Laryngol* 117: 2–9, 2008
- 21) Sharma RR, Mahapatra AK, Pawar SJ, Thomas C, Al-Ismaily M: Trans-sellar trans-sphenoidal encephaloceles: report of two cases. *J Clin Neurosci* 9: 89–92, 2002
- 22) Smith DE, Murphy MJ, Hitchen PW, Babin RW, Abu-Yousef MM: Transsphenoidal encephaloceles. *Surg Neurol* 20: 471–480, 1983
- 23) Tsutsumi K, Asano T, Shigeno T, Matsui T, Ito S, Kaizu H: Transcranial approach for transsphenoidal encephalocele: report of two cases. *Surg Neurol* 51: 252–257, 1999
- 24) Vergoni G, Antonelli V, Veronesi V, Servadei F: Spontaneous cerebrospinal fluid rhinorrhoea in anteromedial temporal occult encephalocele. *Br J Neurosurg* 15: 156–158, 2001
- 25) Willner A, Kantrowitz AB, Cohen AF: Intrasphenoidal cephalocele: diagnosis and management. *Otolaryngol Head Neck Surg* 111: 834–837, 1994

Address reprint requests to: Ealmaan Kim, M.D., Ph.D., Division of Skull Base Surgery, Department of Neurosurgery, Dongsan Medical Center, Keimyung University School of Medicine, 194 Dongsan-dong, Jung-gu, Daegu 700-712, R.O.K.

Commentary

This is a nice report of a rare case of a sphenoidal encephalocele arising from the lateral recess of the sphenoid sinus.

The authors successfully repaired this through a craniotomy and an extradural approach. The reader should be aware of this entity and this extradural approach to the area of the leak in relation to the divisions of the trigeminal nerve. An endonasal endoscopic approach is also an option that some surgeons may prefer to use.

Chandranath SEN, M.D.
Department of Neurosurgery
St. Luke's-Roosevelt Hospital Center
New York, New York, U.S.A.