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Unusual Presentations of Middle Fossa Encephaloceles: Report of Two Cases

ABSTRACT—Basal encephaloceles are rare occurrences with occult and often varied presentations. Frequently, the need for surgical treatment is not clear to the patient or the physician, leading to potentially fatal complications. We report the case of a 44-year-old woman with an 8-year history of nonspecific complaints who presented to us with trigeminal neuralgia. Also, an unusual case is presented of a 60-year-old man with bilateral middle fossa encephaloceles who suffered acutely from widespread pneumocephalus acting as a mass lesion. Both are cases of nontraumatic middle fossa encephaloceles, which are of particular interest due to the age of the patients, their atypical presentations, and the associated complications.

Cranial base encephaloceles are rare occurrences with an estimated incidence of 1:35,000 live births according to Van Nouhuys and Bruyn.¹ Of these, approximately 10–25% affect the anterior skull, while the remaining 75–90% occur in the occipital skull base.^{2,3} As basal encephaloceles protrude into normally developed cavities such as the paranasal sinuses, their presentation is often occult unlike the more common calvarial encephaloceles, which present as obvious deformities at birth. Furthermore, because basal encephaloceles are usually not outwardly visible, the need for surgical correction may not be clear to the patient or the physician.⁴

Clinically, patients with occult trans-sphenoidal encephaloceles can present with a variety of signs and symptoms. Typical presentations include headache and cerebrospinal fluid (CSF) rhinorrhea. In some cases, patients may first present with recurrent meningitis, which in about 80% of cases is caused by *Streptococcus pneumoniae*.^{4,5} Less common presentations include de-

creased visual acuity,⁶ ocular abnormalities such as morning glory syndrome arising from the abnormal development of the optic disc,⁷ endocrine defects,^{8,9} and cranial nerve deficits.¹⁰ Midline cerebral anomalies such as congenital malformations of the fornix, septum pellucidum, and corpus callosum have also been associated with basal encephaloceles.¹¹

While spontaneous CSF rhinorrhea, followed by retro-orbital and/or frontal headaches is the most common presenting symptom, the rare occurrence of these occult malformations, the nonspecificity of clinical signs and symptoms, and the possibility of misdiagnosis based on routine computed tomography (CT) scanning, all indicate the need for heightened clinical awareness of these lesions and their varied presentations. Recently, we encountered two cases of nontraumatic middle fossa encephaloceles, which were of particular interest due to the age of the patients, their atypical presentations, and the associated complications.

Figure 1. Case 1, preoperative CT scans. (A) Coronal image and (B) 3-D reconstruct showing the bony defect of the medial middle fossa floor on the right.

CASE REPORT 1

A 44-year-old female had an 8-year history of multiple nonspecific complaints, including right-sided retro-ocular headaches, dizziness, and tremors. Additionally, she complained of paresthesias and diminished sensation in the second distribution of the trigeminal nerve (V_2), affected by position, especially quick turning of the head or flexion of the neck. She had no evidence or history of CSF rhinorrhea, meningitis, or seizures. Her right sided retro-ocular headaches as well as progressive loss of near vision in the right eye were attributed by several ophthalmologists to a diagnosis of unilateral preglaucoma; topical B-blocker therapy had failed to improve her symptoms, however. She also complained of tinnitus on the right, which had recently increased in intensity. Previously, her physical complaints had been attributed, at least in part, to a diagnosis of Post-Traumatic Stress Disorder, for which she had undergone several years of cognitive and psychotherapy without much success. Her physical exam was otherwise unremarkable and the patient was referred for further diagnostic work-up.

On CT scanning, the patient was found to have a bony defect of the medial middle fossa floor on the right side (Fig. 1A and B). There had been erosion of the foramen rotundum and foramen ovale, consistent with a diagnosis of sphenoid encephalocele. Magnetic Resonance Imaging (MRI) confirmed the diagnosis, indicating a significant amount of temporal lobe herniation into the posterior aspect of the maxillary sinus by the encephalocele (Fig. 2A and B).

The patient subsequently consented to surgical repair of the encephalocele. This was done through a temporal craniotomy and a zygomatic osteotomy. An intra/extradural approach to the middle fossa was used to resect the infratemporal fossa encephalocele, and reconstruction was performed utilizing a dural graft and a temporalis muscle pedicle flap. The procedure was tolerated well and the patient was discharged 7 days postop.

Follow-up examination approximately 18 months later revealed none of the previously encountered signs and symptoms except for a frontalis palsy.

CASE REPORT 2

A 60-year-old man with a known history of CSF rhinorrhea for approximately 3 years, presented to the emergency room (ER) with new onset of fever, complaining of severe headaches and numerous episodes of emesis over the prior several days. The patient had previously been diagnosed as having bilateral sphenoid encephaloceles and had been cautioned about meningitis and seizures; he had refused surgical treatment at the time.

While in the ER, the patient's condition rapidly deteriorated with an abrupt decline in mental status; he became aphasic and quickly lost the ability to follow commands, lapsing into a coma. A CT scan was urgently obtained, which showed massive pneumocephalus (Fig. 3A and B). The patient was subsequently intubated and admitted to the intensive care unit where he underwent

Figure 2. Case 1, preoperative MRI. (A) Sagittal T_1 image and (B) Coronal T_2 image indicating temporal lobe herniation into the sphenoid sinus and the posterior aspect of the maxillary sinus with clear intracranial connections.

lumbar puncture, placement of an Intracranial Pressure (ICP) monitor, and was started on antibiotics and steroids.

Over the next 24 hours, the patient maintained positive corneal, gag, and cough reflexes. His pupils remained fixed at 3 mm bilaterally. He demonstrated extensor posturing and occasionally localized to pain bilaterally, the left side being worse than the right. Plantar responses were down-going. Spontaneous movements, though never purposeful, were occasionally observed.

Analysis of the CSF was consistent with a bacterial meningitis, although all cultures remained negative for any growth. On Day 2, the patient remained febrile but a follow-up head CT showed significantly decreased pneumocephalus with a small subdural hematoma layering out over the orbital roof.

Over the course of 10 days, he remained intubated, being severely impaired but slowly regaining consciousness. He exhibited spontaneous eye-opening and verbalization of one to two word phrases and was following commands briskly on the right side. His left side demonstrated a dense hemiparesis, though he did withdraw to pain. Fine-cut CTs and an MRI of the head showed bilateral hypothalamic infarcts, a small calcified lesion in the right pontine peduncle, as well as a left

frontal subdural collection that was stable with little mass effect. At this point, a clearer evaluation of the bilateral encephaloceles—left larger than right—was obtained (Figs. 4A and B). After consultation with Otorhinolaryngology, it was decided that the left encephalocele was not amenable to endoscopic treatment. He, therefore, underwent a middle fossa craniotomy to resect the encephalocele and reconstruct the defect in the middle fossa floor.

Using a transzygomatic middle fossa approach with a frontotemporal craniotomy, a zygomatic osteotomy was performed and the middle fossa encephalocele was resected. A temporalis fascia dural graft was used and a temporalis muscle reconstruction of the middle fossa skull base was performed. Approximately 10 days after surgery, the patient was alert and awake, but still failed to follow commands and had no verbal communication. Spontaneous movements were observed on the right side only. He was subsequently discharged to a rehabilitation center in stable condition.

On follow-up approximately 1 month later, the patient was conversant, but with clear cognitive impairment. In addition, his vision was markedly impaired in his right eye and his mild left-sided hemiparesis persisted.

Less than 5 months after this surgery, the patient presented with right-sided rhinorrhea and was referred

Figure 3. Case 2, preoperative CT scan. (A) Widespread pneumocephalus acting as a mass lesion. (B) Coronal image indicating bilateral defects of the sphenoid bone.

Figure 4. Case 2, preoperative MR scan. (A) Sagittal T₁ image showing the left sphenoid encephalocele. (B) Coronal T₁ image postcontrast, showing bilateral encephaloceles.

to the Otorhinolaryngology service for endoscopic resection of his persistent right sphenoid encephalocele. An uncinectomy was performed followed by the complete removal of the anterior wall of the sphenoid sinus, which gave wide exposure to the encephalocele. As had been appreciated on the CT scan, the encephalocele proceeded laterally into a recess of the sphenoid sinus. It was at the roof of this lateral recess that the bony defect of the middle cranial fossa existed. Accordingly, the encephalocele was reduced and the defect was reconstructed using septal bone and cartilage. Postoperatively, no evidence of CSF rhinorrhea was found after repeated examinations including rigid nasal endoscopy and the patient was discharged to a rehabilitation facility as before.

DISCUSSION

The development of encephaloceles may be linked to the complex embryology of the skull base. Up to 19 separate centers have been associated with ossification of the sphenoid sinus,¹² with the body of the sinus itself developing from paired chondrocranial ossification centers located in the corpus.¹³ The definitive sphenoid bone is not formed until the first year of life when these centers, growing independently, finally fuse. By the third year of life, gradual erosion of the sphenoid corpus leads to the development of epithelial outpouchings of the superior and dorsal portions of the nasal fossa.¹³ One possible explanation for the development of sphenoid encephaloceles relates to early gestational ossification problems and abnormal coalescence of the ossification centers.^{12–15} Another possibility, as hypothesized by Kaufman et al.,¹⁶ is herniation of tissue into the sphenoid sinus as a result of enlargement of congenital sphenoid “pitholes”; it has been estimated that up to 10% of the normal population may possess an aerated middle fossa floor.^{15,17} The hypothesis is that these small dehiscences may enlarge as a result of CSF pressure changes that occur during straining, coughing or normal physiological variations in intracranial pressure, thus allowing herniation of brain matter into sinuses.^{15,16}

Five types of basal encephaloceles have been identified based on anatomical location.^{1,18,19} Exophthalmos and neurofibromatosis are associated with the sphenoorbital type in which the herniated sac is found in the orbit, protruding through the superior orbital fissure.²⁰ Herniations into the nasopharynx are of the sphenomethmoidal type in which the encephalocele extrudes through the posterior ethmoid cells and the sphenoid bone.²¹ Protrusion into the nasal cavity through the cribriform plate comprises the trans-ethmoidal type,²¹ while encephaloceles placed at the pterygopalatine fossa are of the sphenomaxillary type, which herniate through the inferior orbital fissure.²¹ In the least common type, the

trans-sphenoidal, brain matter is found in the sphenoid sinus or the oropharynx, via defects in the body of the sphenoid bone.¹⁴ It must be noted that not all cases fall into the above categories; Leblanc et al.²² have reported three cases in which meningoencephaloceles were found in the pterygopalatine fossa protruding through a defect at the base of the greater sphenoid wing near the foramen rotundum and the pterygoid process without the involvement of the orbit. These encephaloceles were not readily noticeable on CT scans of the middle fossa and were difficult to diagnose; all three patients presented with medically intractable temporal lobe epilepsy and were successfully treated by surgery.

The first case presented in this report is of particular interest for its clinical presentation. After a thorough MEDLINE literature search, including years 1959 to present, we found only one case, reported by Soyer et al.²¹ in which the patient presented with pain and sensory symptoms in the distribution of the trigeminal nerve. In their case the encephalocele, having protruded through a defect in the greater wing of the sphenoid bone, was described as transalar—an unclassified type—and was differentiated from the sphenoid type by its laterality and normal morphology. Two other transalar cases described by Elster et al.¹² included patients who did not present with parasthesias of the fifth nerve or any other cranial neuralgia. In our case, absence of CSF rhinorrhea or history of meningitis combined with the presence of an array of numerous non-specific complaints, led to an atypical presentation which effectively delayed proper diagnosis and treatment. This is significant as prolonged delay in surgical intervention may lead to numerous complications including acute meningitis with its associated morbidity,^{4,23} pneumocephalus, hydrocephalus, and epilepsy.

The second case presented in this report demonstrates the potentially fatal nature of the possible complications that may arise acutely from an encephalocele. It is also unique due to the patient's age, his presentation and the bilaterality of his skull defect. A MEDLINE search of years 1959 to present, indicated only one case with a similar presentation. Cartwright et al.²⁴ reported an elderly man with no previous history of CSF rhinorrhea or meningitis, who presented acutely with a change in mental status and a tension pneumocephalus that had developed from rupture of a nontraumatic, middle fossa encephalocele. The authors describe a “ball-valve” mechanism in which the intracranial contents prevent the escape of air that has entered the cranial cavity through open fistulous tracts. Without timely surgical intervention, the subsequent exertion of mass effect by the developing tension pneumocephalus can cause focal neurological deficits, parenchymal dissection, herniation, and death. Although our case is only the second in which a spontaneous, nontraumatic middle fossa encephalocele is associated with acute pneumocephalus, it becomes imperative, in the setting of intracranial air and

acute mental status change, to consider the possibility of an existing skull base defect, especially in older, atypically symptomatic patients.

The early diagnosis of sphenoid encephaloceles remains a challenge clinically and radiographically. Indeed, midline masses may be asymptomatic unless large, and even then they are often mistaken for neoplastic or inflammatory processes.¹⁵ Although most encephaloceles are diagnosed radiographically, smaller skull base bone defects can be easily missed on routine axial CT scans. Willner et al.¹⁵ reported a case in which the patient, presenting with recurrent headaches as the only initial symptom, was intermittently treated for rhinitis and sinusitis for a period of 8 years before the appropriate diagnosis was made. Sphenoid sinus encephaloceles can also be missed if the scans indicate mass or fluid in the sinus without delineating a clear intracranial connection.¹⁵ Therefore, a complete evaluation of occult defects in the skull base typically necessitates the use of multiple radiological and clinical diagnostic studies. Without MRI, the condition could be mistakenly diagnosed as sinusitis, especially if the patient does not present with the more common signs and symptoms associated with dural defects such as CSF rhinorrhea and meningitis.

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