Arachnoid Pit and Extensive Sinus Pnematization as the Cause of Spontaneous Lateral Intrasphenoidal Encephalocele

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INTRODUCTION

ABSTRACT

Lateral sphenoid encephalocele, especially within the lateral aspect of the sphenoid sinus, when the sphenoid sinus has pneumatized extensively into the pterygoid recess, are considered exceedingly rare. We report a rare case of lateral intrasphenoidal encephalocele with spontaneous cerebral spinal fluid (CSF) rhinorrhea. Computed tomography demonstrated bilateral arachnoid pit, extensive sphenoid sinus pneumatization, and a defect in the superior wall of the left lateral recess of the sphenoid sinus. Magnetic resonance imaging demonstrated anteromedial temporal lobe herniating through the bony defect.

Key words: Arachnoid pit, lateral recess, sphenoid sinus, spontaneous CSF-leaks, spontaneous encephalocele

An encephalocele is the protrusion of intracranial contents, including brain matter and meninges through a defect in the cranium or skull base. Abiko et al. noted two types of transsphenoidal meningoencephaloceles: The intrasphenoidal and the true transsphenoidal. Intrasphenoidal encephaloceles describe those extending into the sphenoid sinus but confined by its floor.^[1]

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Intrasphenoidal encephaloceles are subdivided by location into medial perisellar and lateral sphenoid recess types.^[2] Perisellar encephaloceles within the sphenoid sinus are considered more common, whereas basal encephaloceles limited to the lateral sphenoid sinus are rare. Temporal lobe encephalocele protruding through a middle fossa defect is rare and has few distinctive features, so is difficult to identify clinically.^[3] In a study of 15 patients, the mean delay between onset of symptoms such as CSF rhinorrhea and the diagnosis was 13.1 months.^[4] The clinical manifestations included CSF rhinorrhea, chronic headache, and a history of meningitis.^[5]

Arachnoid pits are smooth, lobulated bony defects that are related to aberrant arachnoid granulations. CSF pulsation and its pressure also play an important role in the creation of these defects. This role of the CSF is evident from the

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Figure 1: Axial CT scan demonstrates arachnoid pit lateral to sphenoid sinus (black arrow). An approximately 9-mm size defect in the left superolateral wall of the sphenoid sinus (white asterisk) just lateral to foramen rotundum, with a soft tissue density mass filling the adjacent part of the sphenoid sinus. Notice the pneumatization of most far lateral recesses of sphenoid sinus (white arrow).



Figure 3: Arachnoid pit lateral to the right sphenoid sinus is noted (arrow).

pattern of the bony remodeling, with outward concave orientation of adjacent bones.^[6]

CASE REPORT

A 40-year–old male patient presented to the ENT clinic with watery nasal discharge that had persisted for 10 months. This patient had a history of headache associated with nasal obstruction that had been diagnosed by another doctor as hypertrophied turbinate and was advised surgery. But the patient refused to have surgery.

For the past 2 years, the patient had started to experience anosmia. Though gradual in onset and not associated with any watery discharge initially, it was found to be associated with nasal obstruction. For the past 10 months he was having watery nasal discharge which increased when he bent forward or coughed. The discharge trickled into the nasopharynx, though he could not taste any of this



Figure 2: Coronal-reformatted CT scan demonstrates osseous erosion lateral to the left foramen rotundum and lateral sphenoid sinus recess pnematization. An approximately 9-mm size defect in the left superolateral wall of the sphenoid sinus (white asterisk) just lateral to foramen rotundum (black arrow), with a soft tissue density mass filling the sphenoid sinus (curved arrow).



Figure 4: Coronal-reformatted CT scan demonstrates arachnoid pits (white arrows) lateral to sphenoid sinus. A soft tissue density mass filling the sphenoid sinus is noted (black arrow).

discharge it was associated with headache. The patient has no past history of any nasal trauma or surgery and no sign of meningitis.

Computed tomography of brain and paranasal sinuses demonstrated bilateral arachnoid pit lateral to sphenoid sinus and osseous erosion lateral to the foramen rotundum, lateral sphenoid sinus recess pneumatization, and an approximately 9-mm size defect in the left anterolateral wall of the sphenoid sinus just lateral to foramen rotundum, with a soft tissue density mass filling the adjacent part of the sphenoid sinus [Figures 1-4]. Magnetic resonance imaging demonstrated left temporal lobe herniation through a wide defect in the left inferiolateral wall of sphenoid sinus surrounded by a cerebrospinal fluid filled meningeal sac which contains the herniated brain parenchyma [Figures 5, 6].



Figure 5: Coronal IR-FSE T2-weighted image demonstrates left temporal lobe herniation through a wide defect in the left inferiolateral wall of sphenoid sinus surrounded by a cerebrospinal fluid filled meningeal sac.

DISCUSSION

The most common site of CSF leakage is through the floor of the anterior fossa, which communicates with the ethmoid or frontal sinuses, or with the nasal fossa. The sphenoid sinus is rarely implicated as a source of spontaneous CSF fistula.^[6] Communication with the middle cranial fossa and CSF fistula is more likely if the sphenoid sinus is laterally pneumatized.^[6]

CSF pressures and the hydrostatic pulsative forces may lead to the development of pit holes on the middle fossa at the sites of arachnoid villi with herniation of dura/arachnoid or brain tissue.^[6] If such defects are located over the underlying lateral extension of the sphenoid sinus, encephalocele can develop and lead to CSF leakage into the sinus. Therefore, erosion of bone by arachnoid granulations is only significant when it affects a pneumatized part of the skull.^[7]

In a study on a subgroup of 11 patients with spontaneous sphenoid sinus fistulas, the authors concluded that the constellation of empty sella, arachnoid pits, and extensive pneumatization plays a role in the etiopathogenesis of sphenoid sinus fistulas.^[6,7]

A persisting Sternberg's canal should be considered the source of spontaneous CSF-leaks with or without meningoencephaloceles in sphenoid sinuses with extensive lateral pneumatisation, especially when located laterally and below the maxillary nerve.^[8] However, in another study, the conclusion was Sternberg's canal, as historically defined, is not nearly as prevalent as previously reported. Furthermore, the presence of arachnoid pits in all sphenoid CSF leaks and the predominant leak location lateral to the sites of fusion of ossification centers suggests that



Figure 6: Axial T2-weighted spin-echo imaging demonstrate the left temporal lobe herniation through a wide defect in the left inferiolateral wall of sphenoid sinus surrounded by a cerebrospinal fluid filled meningeal sac.

the leaks are acquired. Contributing factors may include arachnoid pits/weaknesses in the skull base and intracranial hypertension.^[9]

The leaks related to arachnoid granulations along the lateral sphenoid were significantly larger than those along the ethmoid and midline sphenoid osteodural defects with an average size of 3.5 ± 0.80 mm.^[7] In this case, the defect was approximately 9 mm and considered one of the largest acquired defect.

Plain high-resolution CT can give indirect evidence of the CSF fistula by revealing bone defect and opacification of sinuses or air cells. MR cisternography can non-invasively reveal CSF leakage in multiple planes without the disadvantage of ionizing radiation. On the T2-weighted spin-echo sequence, the bright signal emanating from the CSF column is well visualized against the paranasal sinuses background of air.^[10] MRI is essential to confirm the extension of the lesion and any associated abnormalities. MRI is able to provide mulitplanar images of the encephalocele that is useful for both diagnosis and surgical planning.

Without repair of the defect, patients with CSF leakage have increased risk of developing ascending infection. Landreneau et al. in a study of four patients, conclude that fistulas involving lateral extension of the sphenoid sinus require a transcranial approach for direct visualization and obliteration of the defect, whereas fistulas involving the central portion of the sinus may be successfully obliterated transsphenoidally.^[11]Transcranial multilayered closure of the defect is safe and reliable, particularly for large CSF fistula at the far lateral sphenoid sinus.^[3]The patient refused the transcranial approach. Endoscopic management is technically challenging, nevertheless its advantages are a good view of the surgical field while being less traumatic than transcranial approaches.^[8]

Determination of the ideal approach is based on various factors, including the degree of lateral sphenoid pneumatization, location, and size of the meningoencephalocele, and ability to perform an adequate skull base repair through a given exposure.^[5]

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