Temporal Lobe Encephalocele in the Lateral Recess of the Sphenoid Sinus Presenting with Intraventricular Tension Pneumocephalus

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ABSTRACT

A basal encephalocele often shows an insidious clinical course. Only two cases of temporal lobe encephalocele accompanied with tension pneumocephalus have previously been reported. In this paper, we describe a case of lateral sphenoid sinus encephalocele presenting with intraventricular tension pneumocephalus. A 54-year-old man was referred to our institution presenting with intraventricular tension pneumocephalus. He had undergone ventriculoperitoneal shunt placement for postmeningitis hydrocephalus 3 months before this admission. Precise imaging examinations detected evidence suggestive of a lateral sphenoidal sinus recess encephalocele. Endoscopic transnasal approach was performed for surgical repair of the encephalocele. The encephalocele was removed with subsequent repair of the bony defect. Histological examination showed that the encephalocele includes a part of the ventricular system. This indicates that air might enter directly into the ventricular system after rupture of the temporal lobe encephalocele. A lateral sphenoid sinus encephalocele would potentially cause intraventricular tension pneumocephalus, although pneumocephalus is an extremely unusual complication of this type of basal encephaloceles.

KEYWORDS: Encephalocele, endoscopic surgery, pneumocephalus, sphenoid sinus

Since basal encephaloceles protrude into physiological cavities such as the paranasal sinuses and are therefore outwardly invisible, the clinical manifestations are often occult in contrast to those of calvarial encephaloceles with apparent deformities. Among several types of basal encephaloceles, a temporal lobe encephalocele protruding into the lateral sphenoid sinus, particularly in the lateral recess of the sinus, is the least common type.^{1–3} This type of basal encephaloceles is difficult to identify in the clinical setting because it rarely occurs and has few characteristic clinical features.^{4–6}

Moreover, lateral sphenoid sinus encephaloceles can easily be missed on routine radiological examinations due to its peculiar location. The insidious nature of lateral sphenoid sinus encephalocele can lead to a delay in diagnosis unless patients present with active cerebrospinal fluid (CSF) rhinorrhea that can motivate physicians to explore cranial base pathologies.^{7,8}

We here present a case of lateral sphenoid sinus encephalocele presenting with intraventricular tension pneumocephalus. Whereas only two cases of tension pneumocephalus caused by rupture of a basal

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Figure 1 A plain CT image showing a marked pneumocephalus exclusively in the lateral ventricle.

encephalocele have been so far reported in the literature,^{9,10} no case of pneumocephalus occurring exclusively in the lateral ventricle has previously been described as a manifestation of basal encephaloceles. The pathophysiological mechanism for this unusual presentation of lateral sphenoid sinus encephalocele is discussed.

CASE REPORT

This 54-year old man was referred to the authors' institution presenting with impaired consciousness. He had past history of bacterial meningitis twice over the last 4 years. He had undergone ventriculoperitoneal shunt placement for postmeningitis hydrocephalus 3 months before the current admission. He had been uneventful and experienced no clinical sign indicating

CSF rhinorrhea during those periods. He had no history of significant head trauma or cranial surgery before.

Admission computed tomography (CT) imaging of the head revealed a large amount of air present predominantly in the bilateral lateral ventricles (Fig. 1). He was somnolent with no focal neurological deficits. Physical examinations found no abnormality in the skin along a subcutaneous CSF shunt tube system. On CT and magnetic resonance imaging (MRI) studies immediately after the admission, no abnormality was found in the cranial base as well as the neighboring paranasal sinuses. Whereas a fistula point through which such a lot of air had entered into predominantly in the lateral ventricle was not determined, lowered intracranial pressure due to CSF shunting was thought to be one of potential mechanisms for tension pneumocephalus. To prevent worsening of pneumocephalus, therefore, extracranial diversion of CSF was occluded by ligating a precordial subcutaneous shunt tube. One week thereafter, CT and MR imaging showed amelioration of intraventricular pneumocephalus, and detected evidence suggesting an extension of soft tissue from the left middle cranial fossa into the left sphenoidal sinus (Fig. 2). Although the lesion was suspected to be temporal lobe encephalocele in the lateral recess of the sphenoid sinus, the mechanism by which air might directly enter the ventricular system remained unclear.

Endoscope-assisted transnasal approach was performed for surgical repair of the encephalocele. After endoscopic sphenoethmoidectomy and maxillary antrostomy, the posterior wall of the maxillary sinus was removed with subsequent partial resection of the pterygoid process. Then, a 30-degree angled endoscopic view clearly demonstrated the lesion as a pulsating membranous tissue bulge (Fig. 3A) in the lateral recess of the sphenoid sinus. The herniated tissue protruding through a small round bony defect was removed. A resection stump was found to have a lumen with a smooth surface from which CSF flowed out. After the tissue stump was sealed with a fibrin glue-soaked autologous fat

Figure 2 (A) Preoperative coronal CT and (B) T2-weighted MRI demonstrating a temporal lobe encephalocele protruding in the left lateral recess of the sphenoid sinus.

Figure 3 (A) Intraoperative view with a 30-degree angled endoscope showing a pulsating membranous tissue bulge (arrow) in the lateral recess of the sphenoid sinus. (B) Endoscopic view illustrating a bony defect repaired with a bone fragment (arrow).

graft, the bony defect was repaired with a bone fragment of the nasal septum (Fig. 3B). Histological examination of the surgical specimen revealed a degenerative brain tissue including a layer of ependymal cells (Fig. 4). Postoperatively, CSF shunting was restored to its original condition by untying the ligature of a precordial subcutaneous shunt tube. On a series of

postoperative CT scans, pneumocephalus was disappeared associated with consequent recovery of his consciousness over time. Nonetheless, he suffered from ventriculitis caused by multidrug-resistant *Pseudomonas aeruginosa*. Despite having undergone the ventricular drainage and received systemic antibiotics, he died 3 weeks after the operation.

Figure 4 Photomicrographs of histological sections of the surgical specimen. (A) Hematoxylin and eosin staining showing a layer of degenerative ependymal cells in line (original magnification, $200\times$). (B) A layer of ependymal cells exhibiting dotted immunoreactivity for epithelial membrane antigen (arrows; epithelial membrane antigen immunohistochemistry, original magnification, $200\times$). (C) A focal staining for neurofilamentous proteins (neurofilamentous proteins immunohistochemistry, original magnification, $100\times$).

DISCUSSION

Tension pneumocephalus is an extremely unusual presentation of basal encephaloceles. To our knowledge, only two cases of tension pneumocephalus associated with a lateral sphenoid sinus encephalocele have been reported in the literature.^{9,10} In both cases, air was found to accumulate within brain parenchyma of the temporal lobe ipsilateral to the side of the encephalocele. Accordingly, the authors ascribed pneumocephalus to rupture of the temporal lobe encephalocele, and they suggested that alterations in the extracranial and intracranial pressure balance play a role in the intracranial entrance of air, although the exact mechanism was unclear. In the present case, unlike the two reported patients, a lot of air was present exclusively in the lateral ventricle. Even when CT and MRI showed evidence to indicate temporal lobe encephalocele in the lateral sphenoid sinus, it still remained unclear whether or not air could directly enter the ventricular system through this cranial base lesion. However, intraoperative findings raised the possibility that temporal lobe encephalocele may contain a part of the inferior horn of the lateral ventricle, and that rupture of the encephalocele may enable air to enter directly into the ventricular system. The possibility was confirmed by histological examination on the surgical specimen revealing that the herniated brain tissue includes a layer of ependymal cells. Intracranial pressure alterations potentially induced by previous placement of ventriculoperitoneal CSF shunt might play a role in facilitating the development of intraventricular tension pneumocephalus. Furthermore, as previously suggested elsewhere,⁹ brain tissue around the fistula point might act as a ball-valve mechanism, consequently preventing the escape of air already trapped in the ventricular system.

A temporal lobe encephalocele protruding in the lateral sphenoid sinus is one of the most unusual types of basal encephalocele.¹⁻³ Previous reports have shown that temporal lobe encephaloceles in the lateral sphenoid sinus present with a variety of signs and symptoms, such as CSF rhinorrhea, meningitis, headache, vertigo, seizure, and some somatic complaints.¹¹⁻¹⁴ In addition to the low incidence, the fact that some of them are not related to a specific pathophysiology of this disease makes the early diagnosis difficult. Moreover, lateral sphenoid sinus encephaloceles are not readily noticeable on routine radiological exams unless the lesion is large enough to be detectable on ordinary axial CT scans. In our case, indeed, routine CT and MRI studies failed to detect abnormalities in the cranial base at first just after admission. Radiological evidence for a lateral sphenoid sinus encephalocele was barely obtained after intracranial pressure was intentionally elevated by obstruction of CSF shunting.

The repair of encephaloceles in the lateral recess of the sphenoid sinus is a unique surgical

challenge. Because of its peculiar location, this type of basal encephaloceles has previously been approached either via transcranial or transfacial route.^{3,15,16} Endoscopic transnasal approaches have recently been employed as an effective and less invasive means to treat basal encephaloceles.^{6,17-19} The endoscopic technique that can provide a wide angled or side angled view has a great advantage particularly in the treatment of lateral sphenoid sinus encephaloceles over conventional transcranial and transfacial approaches that require avoid scalp and facial incisions, respectively. In the present case, indeed, we performed an endoscopic transnasal approach to successfully repair a lateral sphenoid sinus encephalocele in a less invasive way.

CONCLUSION

Our case indicates that a lateral sphenoid sinus encephalocele would potentially cause intraventricular tension pneumocephalus. Whereas this type of basal encephaloceles is difficult to clinically identify, the importance of clinical awareness of this disease and of the early diagnosis should be stressed to prevent the occurrence of such a devastating complication.

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