Meningoencephalocele in the Lateral Sphenoid Sinus Showing Malformation of Cortical Development: A Case Report

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Abstract

Meningoencephalocele in the lateral sphenoid sinus (SS) has been determined to be a rare entity often detected by cerebrospinal fluid (CSF) rhinorrhea. To date, the pathology of meningoencephalocele in the lateral SS has remained to be unclear in many cases. In this study, we report on a case of a 72-year-old woman with an arteriovenous malformation who presented with CSF rhinorrhea. Radiologic investigations revealed a left temporal meningoencephalocele in the lateral SS. We removed the meningoencephalocele and performed skull base repair, after which the CSF rhinorrhea resolved. Pathological examination showed congenital cortical abnormalities with dysmorphic neurons in various shapes and acquired chronic tissue alterations including fibrillary gliosis and scattered Rosenthal fibers. These findings may further aid in understanding the etiopathogenesis of meningoencephalocele in the lateral SS.

Introduction

Meningoencephaloceles consist of cerebral parenchyma and meninges that herniate into the extracranial compartment through a cranial bony defect.[1] Meningoencephalocele in the lateral sphenoid sinus (SS) most commonly occurs after trauma or skull base erosion due to inflammatory or neoplastic disorders. Spontaneous meningoencephalocele in the lateral SS is considered rare, and it is often accompanied by various clinical symptoms, such as cerebrospinal fluid (CSF) rhinorrhea, chronic headache, epileptic seizures, and meningitis.[2-5] Surgical repair of the skull base is often attempted to prevent central nervous system infection in patients with CSF rhinorrhea. Previously, the etiopathogenesis of meningoencephalocele in the lateral SS was believed to be correlated with a persistent skull base bone defect in the lateral craniopharyngeal (Sternberg's) canal. More recently, dynamic disturbances in CSF and increased intracranial pressure (IICP) were determined to be associated with this rare entity, which is sup-

ported by radiologic and clinical observations.[4]

In this study, we have recently encountered gradually protruding meningoencephalocele in the lateral SS presenting as CSF rhinorrhea in a patient with arteriovenous malformation (AVM). We resected the meningoencephalocele and repaired the bony defect in the middle fossa with temporal fascia and an autologous bone fragment to prevent a central nervous system infection. There are only few studies on the pathological findings of meningoencephalocele in the lateral SS.[1,2,6] Furthermore, the pathological findings of meningoencephalocele in the lateral SS are not well described; thus, the pathological features of meningoencephalocele in the lateral SS are yet to be elucidated. Here, we report our experience of a rare, spontaneous meningoencephalocele in the lateral SS case and provide pathological details and a literature review.

Case Report

A 72-year-old female with no previous history of trauma

was diagnosed with a right parietal lobe cerebral AVM 23 years ago (Fig. 1A). The patient has reportedly experienced intermittent, massive watery rhinorrhea for the past 5 years, accompanied by chronic headache. She was overweight with a body mass index of 26.0 kg/m^2. Plain computed tomography (CT) of the head revealed a bony defect in the left middle skull base and a continuous structure extending into the left lateral SS (Fig. 1B). Three-dimensional CT revealed a bony defect that was integrated into the foramen rotundum (Fig. 1C), whereas coronal CT images showed multiple small bony defects (Fig. 1D). Magnetic resonance imaging (MRI) revealed a trabecular structure with low signal on T1-weighted images and high signal on T2-weighted images, and the trabecular structure was noted to be continuous from the tip of the temporal lobe to the lateral SS (Fig. 1E). MRI constructive interference in the steady-state sequence showed an empty sella (Fig. 1F). The mass gradually increased for over 12 years (Fig. 1G-I). Based on these findings, the mass that spanned the left middle cranial fossa to the lateral SS was diagnosed as meningoencephalocele. We concluded that surgical repair of the meningoencephalocele was necessary to avoid the risk of central nervous system infections due to repeated intermittent CSF rhinorrhea, meningoencephalocele, and potential IICP due to the ipsilateral AVM. We opted for a combined transcranial and endoscopic endonasal approach to thoroughly repair the defects.

A lumbar drain was inserted before surgery. After performing a left frontotemporal craniotomy, we separated the middle fossa dura from the skull base, and the V2 and V3 nerve roots and several arachnoid pits that invaginated into bone were thereafter identified (Fig. 2A-C). On closer inspection, the suspected V2 root was enlarged, and the covered dura was stretched to the enlarged foramen rotundum. The brain tissue was slightly exposed through the broken dura. The brain tissue covered with dura mater was coagulated and incised. Next, the V2 nerve was identified in the medial foramen rotundum. Using endonasal endoscopy, the natural ostium of the SS was magnified and observed using a 70-degree endoscope. We then observed the pulsatile mass through the enlarged natural ostium in the SS (Fig. 2D), and the incised brain tissue was removed from the nasal cavity. A bony defect was seen outside the foramen rotundum. The dural defect was closed using a galeal patch with sutures (Fig. 2E). The defects of the middle fossa, enlarged foramen rotundum, and several caves by arachnoid pits were covered with the bone fragment consisting of the inner plate of the frontotemporal bone and pedunculated temporalis fascia (Fig. 2F). After surgery, no obvious CSF rhinorrhea was observed, and she was discharged from the hospital. She has not experienced recurrence 1 year since the surgery.

Pathological findings

Histological examination of the resected mass revealed that it consisted of brain tissue and a cyst partially covered with ciliated columnar epithelium and a thick fibrocollagenous tissue membrane (Fig. 3A). Microscopically, the brain surface was not clearly demarcated from the submucosal fibrocollagenous tissue. Glial tissue immunolabeled with glial fibrillary acidic protein was intermixed with fibrocollagenous tissue, which is then stained green when the surface membrane was stained with Elastica-Goldner staining (El-Gold) (Fig. 3B-D); the gliotic cortical tissue was noted to be irregularly extended under the fibrocollagenous tissue (Fig. 3E-G). Epithelial membrane antigen-labeled cells that were meningothelial in nature were evident in the sparse fibrous tissue covering the cortex (Fig. 3 E inset). The resected cortex exhibited severe cortical laminar disorganization accompanied by large dysmorphic neurons (arrows in Fig. 3H and I) including binuclear neurons (Fig. 3I inset). The cortical tissue around the cyst showed dense fibrillary gliosis with scattered Rosenthal fibers (arrowheads in Fig. 3H). Furthermore, abnormal clustered arterioles and venules with thickened fibrous adventitia were also observed to be evident on the cortical surface (Fig. 3 J).

Discussion

Spontaneous meningoencephalocele in the lateral SS is known to be extremely rare. Recently, the etiopathology of meningoencephalocele in the lateral SS favors a hypothesis that involves an interaction of physiologic and anatomic factors.[4] As a physiologic factor of meningoencephalocele in the lateral SS, progressive erosion of the skull base in patients with IICP and well-pneumatized sphenoid sinuses may result in focal areas of dehiscence and herniation of the brain parenchyma.[5] Generally, whether CSF pressure is increased in patients with unruptured brain AVMs, as in our case, remains to be unclear. Rossitti reported a theory for the pathophysiologic development of increased CSF pressure in patients with brain AVMs and applied a basic hydraulic hypothesis related to cerebral intravascular and CSF pressures.[7] Additionally, vascular malformations have been considered as the cause of bone defects.[8] Bony defects are considered to be acquired and occur during development, rather than being caused by local pressure on the bone due to vascular malformations.[2] Salehian et al. reported that bony defects are often caused by changes in local temperature, oxygen content, and venous flow caused by the vascular malformation, regardless of whether the malformation is in contact with the bone.[9]

One anatomical factor of meningoencephalocele in the lateral SS assumes that errors in the embryological development of the sphenoid bone may also result in congenital defects of the skull base.[5] Indeed, the development of the sphenoid bone is complex, and it involves the fusion of multiple cartilaginous precursors into a single osseous structure. Incomplete fusion of the precursor of the greater
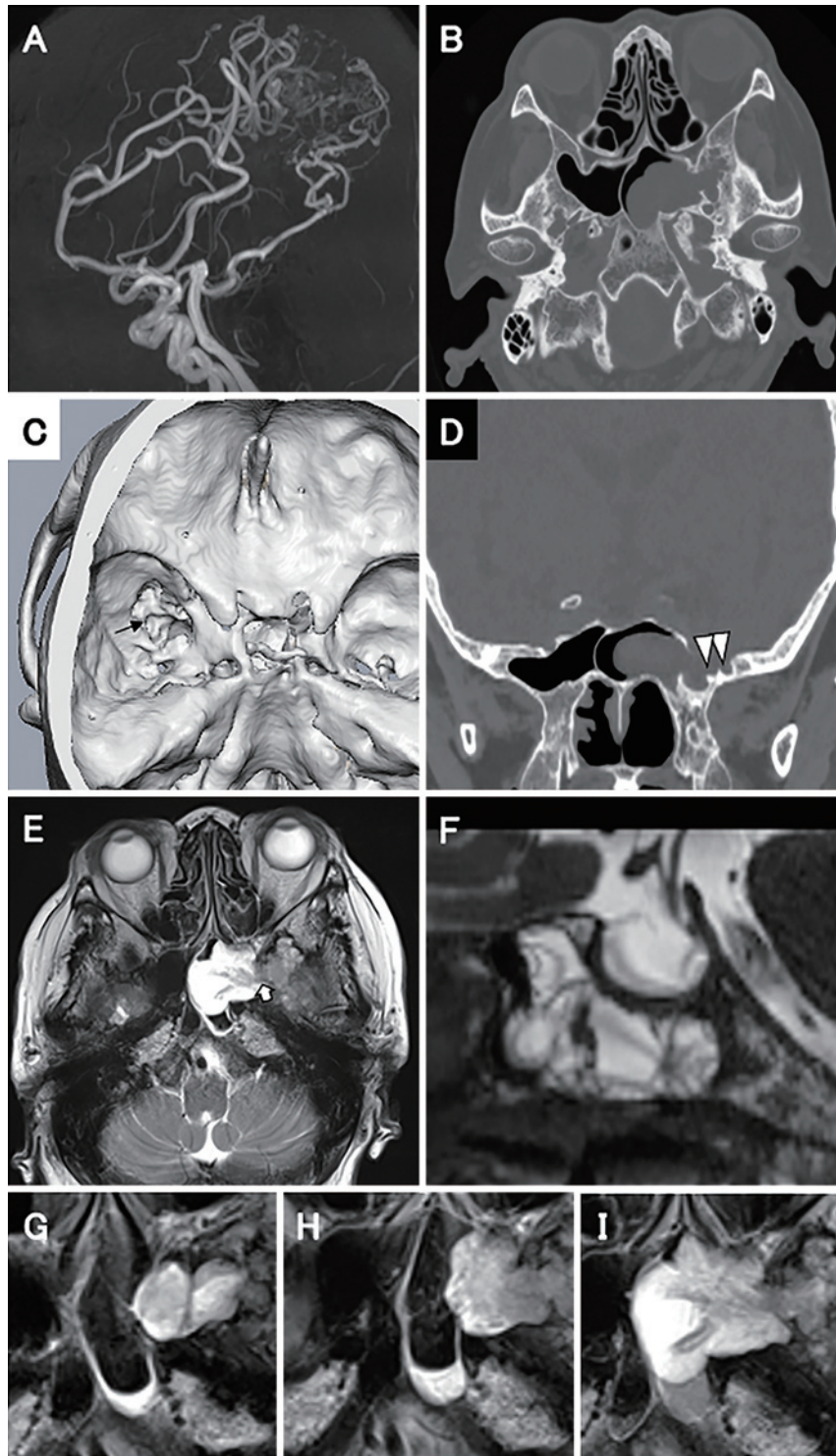
Fig. 1 Preoperative radiological images show a meningoencephalocele in the lateral sphenoid sinus and a bony defect in the left middle skull base.
A: MR angiography images show an arteriovenous malformation in the right parietal lobe. B: CT image reveals a bony defect in the left middle skull base and a continuous structure that protrudes into the left lateral sphenoid sinus. C: Three-dimensional bone computed tomography image reveals a large bony defect outside the foramen rotundum (black *arrow*). D: Coronal CT image with multiple arachnoid pits in the greater wing of the sphenoid bone (white *arrowheads*). E: Axial magnetic resonance imaging (MRI) reveals a trabecular structure with a low signal on a T1-weighted image (not shown) and a high signal on a T2-weighted image. F: Sagittal constructive interference in the steady-state sequence shows the empty sella. G–I: Retrospective MRI on T2-weighted images shows how the mass gradually increased (G: 12 years before surgery, H: 9 years before surgery, I: 2 years before surgery).
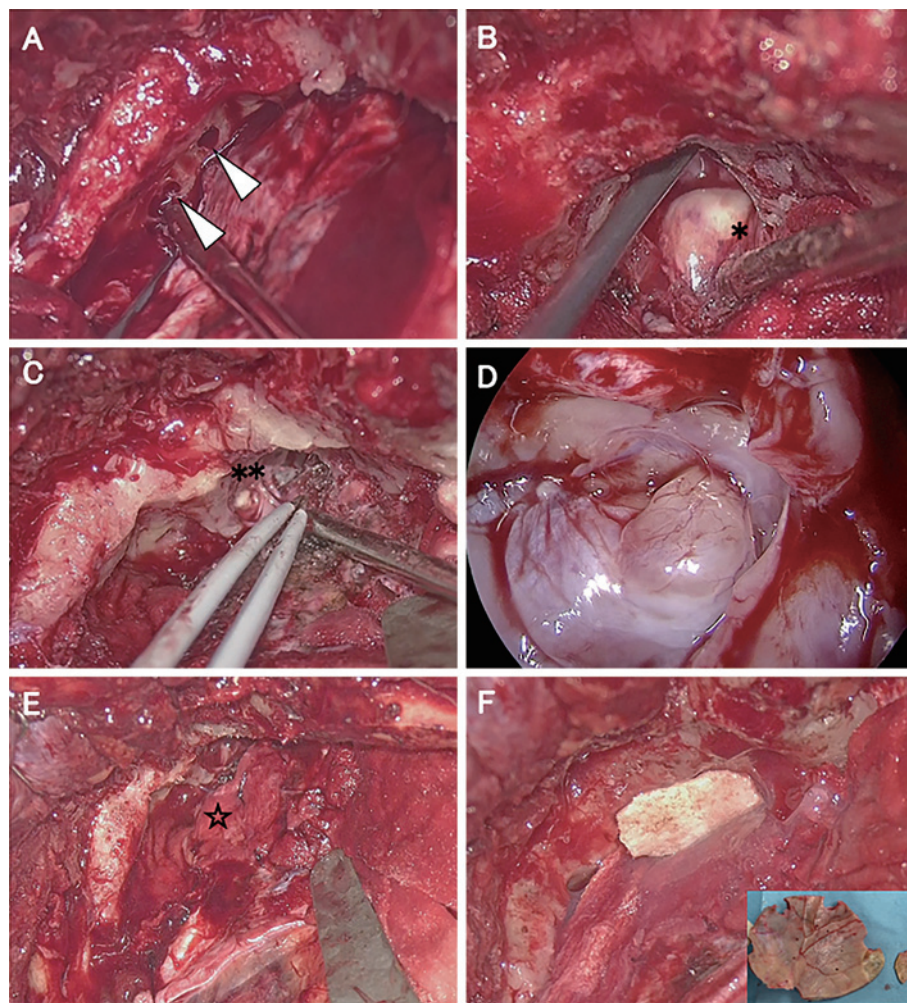
Fig. 2   Intraoperative images.
A: Multiple arachnoid pits (white *arrowheads*) in the middle fossa exposed after a left frontotemporal craniotomy. B: Partially removed brain tissue covered with dura. Asterisk (*) indicates the V2 root through the medial bone defect. C: Double asterisks (**) indicate the large bony defect and arachnoid pit. D: 70-degree endonasal endoscopic view. Brain tissue covered with mucosa was revealed in the sphenoid sinus. E: The dural defect was patched with temporal fascia after resection of the meningoencephalocele. F: The large bony defect and multiple bone pits in the middle fossa were repaired with the pedunculated temporalis fascia and a bone fragment made of the inner plate of the frontotemporal bone.

wing of the sphenoid with the presphenoid and basisphenoid areas can result in a persistent channel termed Sternberg's canal.[4,5,10] Arachnoid pits, pneumatization of the lateral recess of the sphenoid sinus, and an empty or partially empty sella are associated radiological signs based on CT and MRI observations.[4,10] Another anatomical factor is that the extent of pneumatization of the lateral SS develops completely only by adolescence. Lateral pneumatization in the sphenoid sinus has been observed in 23%-43% of patients and has been postulated as a predisposing factor for lateral recess leaks.[7] Development of the SS is completed by adolescence.[11] Settecase et al. reviewed the radiologic images of 26 patients with meningoencephalocele in the lateral SS to identify the anatomical factors that contribute to its pathogenesis and proposed that patients with

meningoencephalocele in the lateral SS may be divided into two types.[10] The first type of meningoencephalocele in the lateral SS herniates into a pneumatized lateral recess of the sphenoid sinus. In this type, patients typically present with CSF leakage and/or headache. The second type herniates into the great sphenoid wing with no involvement of the sphenoid sinus. Patients with this type most commonly present with seizures and other neurologic signs. All patients have arachnoid pits on their ipsilateral or contralateral inner plate of the greater sphenoid bone as a radiological feature. Additionally, they postulated that the development of meningoencephalocele in the lateral SS is related to altered CSF dynamics and increased hydrostatic pressure and pulsatile forces, which could all lead to the development and formation of arachnoid pits.[9] This
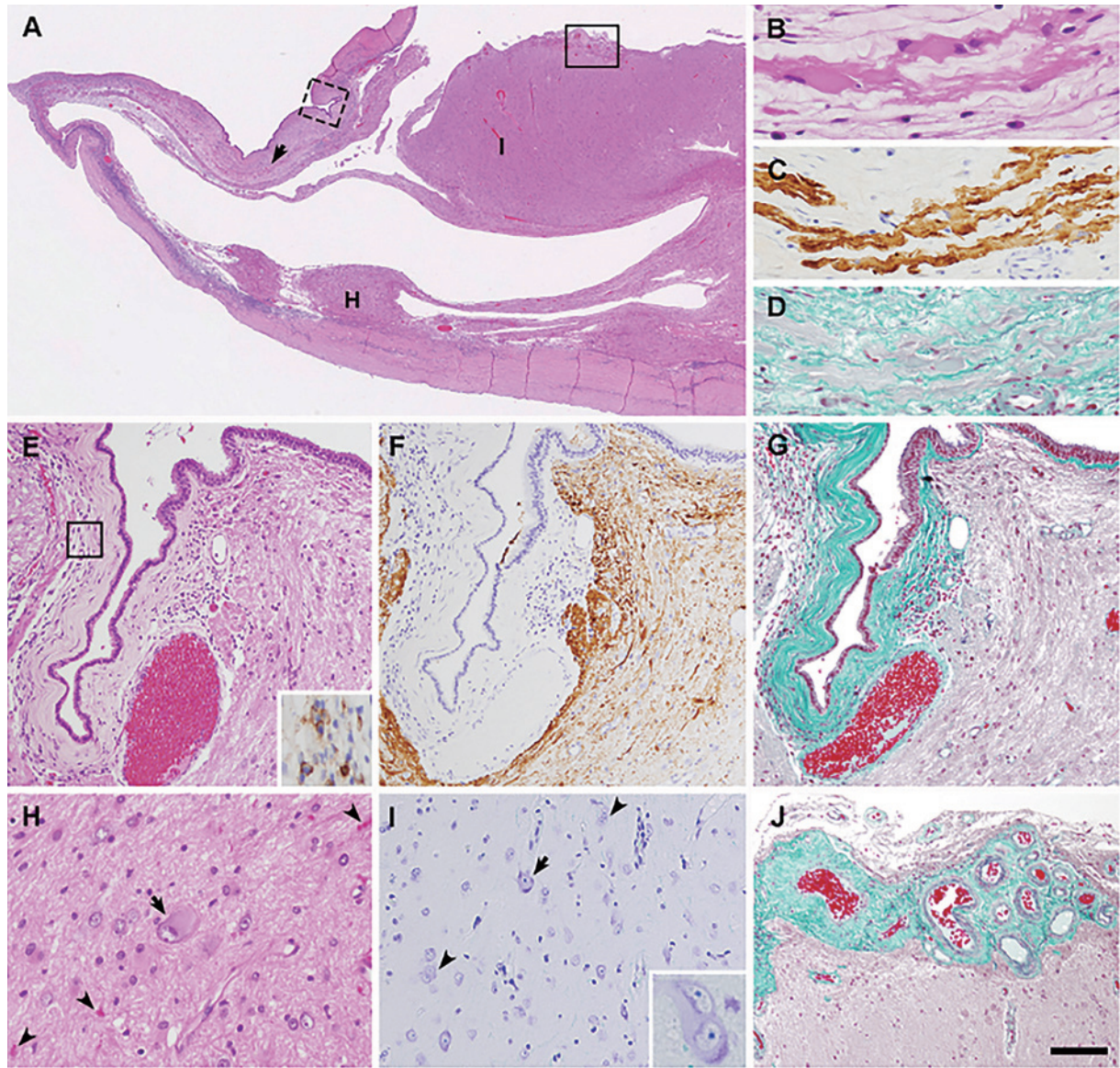
Fig. 3   Histological findings of the resected specimen.
A: The mass consists of brain tissue with a cyst partially covered with epithelium and a thick connective tissue membrane. H&E.
B–D: Higher magnification of the area indicated by an *arrow* in A. Glial tissue immunolabeled with glial fibrillary acidic protein (GFAP) is intermixed with fibrocollagenous tissue, whose surface was stained green with Elastica-Goldner staining (El-Gold). B, H&E; C, GFAP; D, El-Gold. E–G: Higher magnification images taken from the consecutive section of A. The area indicated by the *dotted square* shows an irregular surface of gliotic cortical tissue covered with ciliated columnar epithelium lined with a dense fibrocollagenous band and sparse fibrous tissue. Inset: Sparse fibrous tissue indicated by a *square* in E includes epithelial membrane antigen (EMA)-labeled cells that are meningothelial in nature. E, H&E; F, GFAP; G, El-Gold; inset in E, EMA. H: A higher magnification image taken from the area indicated by H in A shows cortical tissue with fibrillary gliosis and a dysmorphic large neuron (*arrow*) and scattered Rosenthal fibers (*arrowheads*). H&E. I: A higher magnification image taken from the area indicated by I in A shows uneven neuronal distribution with a dysmorphic neuron (*arrow*) and closely adjacent small neurons (*arrowheads*). Inset: A binuclear dysmorphic neuron. Klüver-Barrera staining. J: A higher magnification image taken from the area indicated by the *square* in A exhibits abnormal clustered arterioles and venules with thickened fibrous adventitia. El-Gold. Bar = 800 μm for A, 30 μm for B, and inset in E, 50 μm for C, D, and H, 100 μm in E–G, 75 μm for I, 115 μm for J, and 10 μm for inset in I.

mechanism may play a significant role in the pathogenesis of meningoencephalocele in the lateral SS.[4,10,12] Moreover, according to previous studies, an empty or partially empty sella is evidence suggestive of altered CSF hydrodynamics in patients with meningoencephalocele in the lateral SS.[4] Patients with meningoencephalocele in the lateral SS share common clinical features, such as female sex, middle age, obesity, and IICP.[3-5] The female preponderance may be ge-

netic in origin.

Recently, the endonasal endoscopic repair of spontaneous meningoencephalocele in the lateral SS, specifically the transpterygoid approach, has been identified as a major reconstructive method.[3,5,13] However, the management of patients with spontaneous meningoencephalocele in the lateral SS remains to be controversial, as various surgical approaches and reconstruction methods are reported in the literature. Determination of the ideal approach is based on various factors, including the degree of lateral SS pneumatization, location and size of the meningoencephalocele, history of epilepsy, and ability to perform an adequate skull base repair through a given exposure.[5,13] In the presented case, similar to a previous report, we performed a combined transcranial and endonasal endoscopic approach to remove the large meningoencephalocele that protruded into the SS and robustly repaired the comparatively large dural and bony defects.[1,14,15] We selected this approach for three reasons. One reason is the annual and long-term risk for meningitis of 10% and 40%, respectively, in patients with meningoencephalocele in the lateral SS.[16] The second reason is that patients with AVM potentially have steady IICP, as has been mentioned above. Furthermore, the patient was found to have multiple other risk factors, such as female sex, obesity, middle age, and a highly pneumatized sphenoid sinus.[4,5,17] The third reason is that the therapeutic long-term results of the different reconstruction methods are not known. If the AVM remains untreated, elevated intracranial pressure also remains. Therefore, we elected to repair the defect as thoroughly as possible. After the rigorous repair surgery, the patient has experienced neither CSF rhinorrhea nor chronic headache for more than 1 year.

Pathologic confirmation of resected meningoencephalocele in the lateral SS was not presented in many reported cases, and few detailed reports on the pathological findings of meningoencephalocele in the lateral SS have been published. The reported pathologic features of meningoencephalocele in the lateral SS include gliosis, unspecified or benign brain parenchyma, astrocytosis, and cortical dysplasia.[1-3,17,18] In this present case, the resected meningoencephalocele consisted of cerebral cortical tissue that exhibited severe cortical laminar disorganization accompanied by dysmorphic neurons suggestive of cortical dysplasia. Furthermore, vascular malformations were observed on the cortical surface. These findings were considered congenital abnormalities rather than acquired changes. On the contrary, fibrillary gliosis and Rosenthal fibers observed in the cortical tissue around the cysts were believed to be secondary chronic degenerative changes. Interestingly, although various-shaped dysmorphic neurons were observed in the resected cortical tissue, the patient had no obvious history of epilepsy. Focal cortical dysplasia (FCD) characterized by cortical architectural dysplasia, dysmorphic neurons, and balloon cells is known to be a major cause of epilepsy.[8,19,20]

Dysmorphic neurons, which are observed in the epileptic brain and are considered to be related to seizure generation, are first observed as neural precursors during the development of the cerebral cortex.[4,10,17] However, whether the various-shaped dysmorphic neurons in this case are physiologically identical to dysmorphic neurons in FCD is yet to be determined.[16,20] Most patients who underwent only lesionectomy of an epileptogenic encephalocele were seizure-free during follow-up.[1,2,6,18,21] Considering this finding, the headaches observed in this case before surgery may have been a symptom of epilepsy. No reports on the resected brain tissue of meningoencephalocele in the lateral SS showing the various histopathological findings seen in this case have been published thus far.

We present this case of meningoencephalocele in the lateral SS development and its gradual progression in a patient with AVM, which caused chronic elevation of intracranial pressure. This patient had a pneumatized lateral recess of the sphenoid sinus, arachnoid pits, and empty sella and also possessed risk factors for meningoencephalocele in the lateral SS, such as female sex, obesity, and middle age. The etiology of meningoencephalocele may be related to multiple factors including preexisting Sternberg's canal. The pathological findings in our case may support the above-mentioned multifactorial theory.

Funding

Ethical Approval

This case report is in accordance with the ethical standards of the institutional ethics committee and COPE guidelines and complies with the CARE statement.

Informed Consent

The participant has consented to the submission of the case report to the journal.

Conflicts of Interest Disclosure

None.

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