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Occult Spontaneous Lateral Temporal Meningoencephalocele: MR Findings of a Rare Developmental Anomaly

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Summary: We describe a case of an occult lateral temporal meningoencephalocele discovered in a 14-year-old boy during a work-up for minor head trauma. This spontaneous encephalocele resulted from a closure defect at the former sphenoidal fontanelle. Preoperative MR images are presented.

Index terms: Brain, hernia; Head, injuries; Temporal bone, abnormalities and anomalies

Temporal lobe meningoencephaloceles may develop spontaneously as congenital maldevelopments or they may occur subsequent to acquired processes, such as otologic infection, trauma, surgical defects, or neoplasms (1). Although autopsy studies have shown that congenital defects of the temporal bone occur frequently (2), spontaneous meningoencephaloceles of the temporal bone are rare. Among these, meningoencephaloceles projecting within the lateral temporal bone are the least frequent. Each patient we found reported in the literature (3–7) presented with a lesion that was overt at birth or early in childhood. We report a particularly unusual lateral temporal meningoencephalocele that was occult until adolescence, when it was detected during imaging for minor head trauma.

Case Report

A 14-year-old boy struck his head during a fall from a bicycle, transiently experiencing both minor depression in level of consciousness and difficulty with his speech. Computed tomography and magnetic resonance (MR) imaging at another institution showed an acute, focal brain contusion and adjacent subdural hematoma. In addition, directly adjacent to the contusion was a soft-tissue lesion within the left temporal calvaria, which was reported to be an incidental congenital intradiploic epidermoid tumor. The patient was referred to our institution 3 months later for further evaluation.

On admission, the patient described intermittent headaches, suggestive of postconcussion syndrome. He was alert and oriented, spoke without difficulty, and had no neurologic deficits. An area of irregularity was palpable in the left temporal region.

Subsequently, thin-section, high-resolution MR imaging was performed, which showed a 10-mm defect within the anterior squamosal portion of the left temporal bone, through which both dura mater and cerebrospinal fluid (CSF) were seen to project (Fig 1). A small mass with intermediate signal intensity on T1-weighted (500/25/1 [repetition time/echo time/excitations]) spin-echo images (Fig 1A) and high signal intensity on proton density–weighted (2000/30/1) and T2-weighted (2000/80/1) spin-echo images (Fig 1B and C) projected through the defect and was contiguous with the underlying temporal lobe. A small area of the underlying temporal lobe had similar signal intensity properties, suggestive of adjacent encephalomalacia or gliosis. These findings were consistent with a lateral temporal meningoencephalocele with probable encephalomalacia from prior trauma.

After group consultation and patient consent, a left temporal craniotomy was performed. Surgery confirmed the diagnosis of a congenital meningoencephalocele, in which a knob of encephalomalacic, herniated cerebrum was found protruding through a smoothly marginated, oval defect of the cranium bifidum. The encephalocele was resected and the calvarial defect repaired with titanium mesh and temporalis muscle. Pathologic evaluation of the resected specimen showed gliosis and hemosiderin deposition. The patient had no complications from the procedure. After a brief postoperative course, he was discharged home in excellent condition, with no deficits evident on his neurologic examination.

Discussion

A cranium bifidum, or meningoencephalocele, is a gap in the skull with herniation of adjacent meningeal and brain substances. Meningoencephaloceles can result from various

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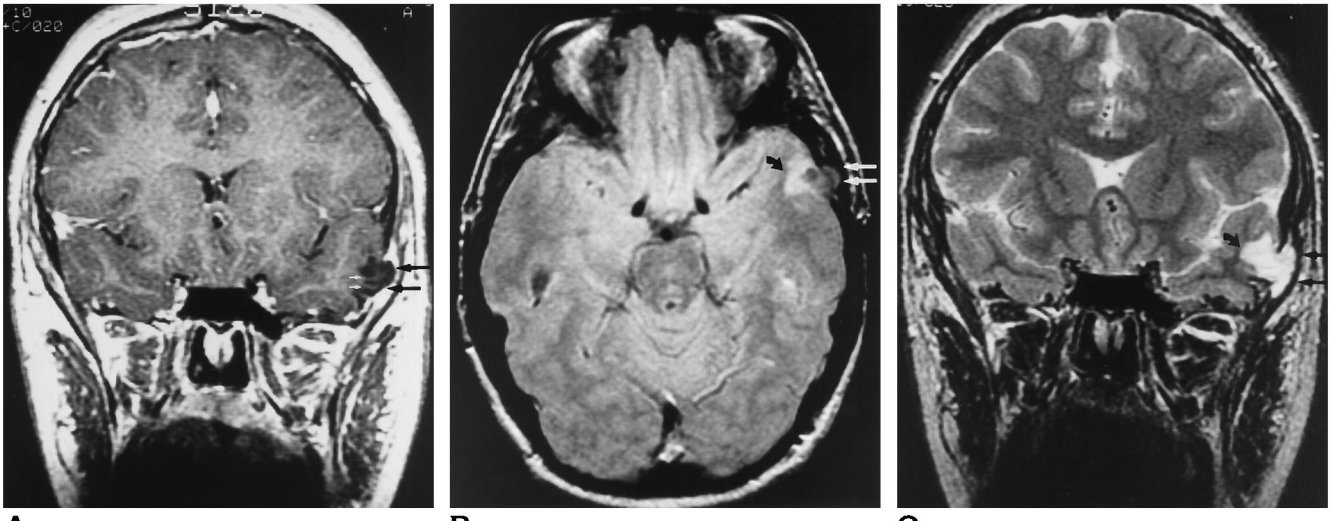

Fig 1. A 14-year-old boy with occult spontaneous lateral temporal meningoencephalocele.

A, Coronal T1-weighted (450/27/1) contrast-enhanced MR image shows a defect in the lateral temporal bone (*large arrows*) that contains CSF and herniated cerebral tissue (*small arrows*). The CSF is continuous with the subarachnoid space.

B, Axial proton (spin) density–weighted sequence (2000/30/1) shows a temporal lobe defect (*straight arrows*) with regions of both low signal intensity (isointense with CSF) and intermediate signal characteristics (isointense with white matter). An adjacent region of high signal in the temporal lobe may be gliotic change from prior trauma (*curved arrow*).

C, Coronal T2-weighted sequence (2000/80/1) shows a region of high signal (*straight arrows*) within the bony defect that is isointense with CSF. An adjacent region of high signal in the temporal lobe may be gliotic change from the prior trauma (*curved arrow*).

acquired processes, including infection, trauma, surgical damage, and neoplasms (1). Meningoencephaloceles, which develop in the absence of such acquired processes, are congenital or early postnatal maldevelopments termed spontaneous meningoencephaloceles (8). These spontaneous lesions usually occur at the site of a cranial suture, and most are the result of a primary or secondary midline defect in closure of the neural tube (4). Nagulich et al (3) listed the most frequent sites of occurrence of spontaneous meningoencephaloceles as nasal, nasopharyngeal, buccal, nasoorbital metopic, interparietal, occipital, and suboccipital.

The majority of spontaneous meningoencephaloceles associated with the temporal bone fall within the category of spontaneous basal meningoencephaloceles, and they are classified according to the anatomic location of their basal temporal defect: anterior (a defect of the sphenoid wing), anteromedial (a defect of the anteromedial middle fossa), posteroinferior (a defect of the tegmen tympani), and anteroinferior (a defect of the anteroinferior middle fossa) (8). The present case illustrates a rare fifth, nonbasal subtype, termed a *spontaneous lateral temporal meningoencephalocele*.

Spontaneous lateral temporal meningoencephaloceles represent lateral closure defects of the neural tube at the pterion, a craniometric point corresponding to the former sphenoidal (or anterolateral) fontanelle, or the asterion, which corresponds to the former mastoid (or posterolateral) fontanelle. Embryogenesis of the temporal bone involves the development and union of its squamous, tympanic, petrosal, and styloid components (9). During the eighth week of development, the squamous component undergoes membranous ossification from a single center. Similarly, at 12 weeks, the tympanic ring arises from the fusion of four intramembranous ossification centers. In contrast, the petrosal and styloid components undergo endochondral ossification from 16 ossification centers. Both the pterion and asterion are points of late closure of the membranous cranium where squamous temporal bone forms beveled sutures with the greater sphenoid wing and membranous portions of the frontal, parietal, and supranuchal occipital bones (3–5).

The pathogenesis of spontaneous temporal meningoencephaloceles is unclear. Autopsy work by Åhrén and Thulin (2) showed that defects of the petrous temporal bone are common. In their randomized series, they found perforations of the tegmen tympani in 21% of patients, and a further 16% were noted to have only a thin

layer of cortical bone covering the pneumatic cellulae of the tegmen tympani. In the absence of infection or increased intracranial pressure, however, the mechanism of meningoencephalic herniation into the bony defect is controversial. Using a rabbit model, Falconer and Russell (10) showed that meningoencephaloceles may develop initially as hematomas and cysts at sites of contact of gray matter and bony cortex. Some investigators have also suggested that dural defects at the site of pacchionian bodies and arachnoid granulations may play a role in pathogenesis (1, 11). Referring specifically to lateral temporal defects, Nagulich et al (3) contended that the herniation results from an ectopic deposit of meninges into the fontanellar island during the fetal stage.

We found approximately 15 lateral temporal meningoencephaloceles reported in the literature (3–7). Among these cases, 14 herniations occurred at the pterion and one at the asterion. None of the lesions described was occult. In the present case, the anterior location of the lesion defines it as a herniation at the pterion. In all these previously reported cases, the patients had a mass or deformity at physical examination. As with other spontaneous meningoencephaloceles, they appeared during infancy or early childhood, after a period of gradual enlargement. At presentation, the lesion is compressible, fluctuant, nonpulsatile, and becomes increasingly tense with crying. Other symptoms common to congenital defect temporal meningoencephaloceles, such as CSF rhinorrhea or otorrhea, progressive hearing loss, seizure activity, and recurrent meningitis (1), have not been described in cases of lateral defects.

In the present case, we found that multiplanar, thin-section MR spin-echo imaging provided an excellent three-dimensional definition of the lesion that was useful for both diagnosis and surgical planning. As with any meningoencephalocele, diagnosis relies on the demonstration of a sac that is directly contiguous with the subarachnoid space and that contains a component of herniated cerebrum. The herniated brain tissue can have signal characteristics of normal nervous tissue or, as in our case, increased signal on sequences with long repetition times. MR imaging, with its capacity to define and characterize the contents of the sac, provided the information necessary to confidently differentiate this lesion from epidermoid tumors, leptomeningeal cysts, arachnoid cysts, sinus pericranii, and lytic primary or metastatic bone tumors that may occur in the same region. MR imaging also provides the best method for identifying any associated intracranial anomalies. Meningoencephaloceles can be associated with Chiari malformations, holoprosencephaly, Dandy-Walker complex, aqueductal stenosis, agenesis of the corpus callosum, and other midline abnormalities (12).

In conclusion, occult congenital lateral temporal meningoencephalocele needs to be considered in the differential diagnosis of a patient with an incomplete bony defect at the pterion or asterion with adjacent meningoencephalitic herniation.

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