Spontaneous temporal encephalocele

Case report

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The authors report a 36-year-old woman with a 23-year history of simple and complex partial seizures who was treated surgically for an anteroinferior temporal encephalocele, with resolution of the seizure disorder. This patient's presentation, findings, and response to treatment are typical of those associated with anteroinferior temporal encephalocele, and different from the clinical patterns of four other types of spontaneous temporal encephalocele.

KEY WORDS • encephalocele • seizure • temporal lobe

A patient was recently evaluated and treated at our institution for an encephalocele extending through the anterior floor of the middle cranial fossa. A review of the published reports of similar and related cases indicates that there are five types of spontaneous temporal encephalocele, each with characteristic clinical features. Our case is reported and the clinical patterns of the five distinct types are reviewed.

Case Report

This 36-year-old woman was referred for evaluation of an abnormality at the right side of the skull base. She gave a history of partial seizures dating back to the age of 13 years, when she began noticing episodes that were recurrent and stereotypic. A spell would begin with a warm feeling in the lower extremities that would then extend over the trunk into the shoulders and head before subsiding; it would last 10 to 15 seconds and was not accompanied by confusion or impairment of consciousness. Initially, the episodes occurred in unpredictable flurries of several per day, but later they developed a pattern of occurrence 9 to 14 days after the initiation of menses, during which she would have between five and 20 episodes over a 2- to 3-day period.

At the age of 18 years she had two witnessed seizures that involved loss of consciousness without shaking movements or automatisms, but with postictal confusion. A routine electroencephalographic (EEG) recording was normal, but a second study with nasopharyngeal electrodes demonstrated epileptiform discharges from the right temporal region. Computerized tomography (CT) of the head was interpreted as normal and she was begun on a course of phenytoin and phenobarbital. Because these anticonvulsant drugs produced lethargy and did not affect the frequency of the episodes significantly, the patient stopped taking them.

When she was 23 years old and pregnant, the patient had two more episodes of loss of consciousness similar to the initial two. Anticonvulsant medication was not given, and she continued to have the stereotypic minor events without definite impairment of consciousness. Two months prior to her referral at the age of 36 years, she had an episode during sleep that was witnessed by her husband. He was awakened by her grunting and found her in the fetal position with her extremities rigid and her head extended. He had difficulty awakening her; she then appeared to be confused before falling asleep again. She was evaluated by her physician and magnetic resonance (MR) imaging of the head was performed. A lesion, thought initially to be an epidermoid tumor, was noted at the base of the skull in the right temporal region, and the patient was referred for surgical treatment.

Examination. When the patient was first seen at our institution on March 11, 1991, her neurological examination was normal. A CT head scan showed an abnormality in the floor of the right middle fossa, and MR imaging demonstrated a large mass centered in the right infratemporal region, projecting into the sphenoid sinus medially and abutting the maxillary sinus anteriorly (Figs. 1 and 2). Based on the CT and MR studies, the abnormal process was not thought to involve the hippocampus or amygdala. Positron emission tomography revealed a focal area of hypometabolism in the anterior aspect of the right temporal lobe.

The patient was hospitalized for 2 days in April for prolonged video and EEG monitoring. During this period she reported a series of three auras that were
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![Image](image_url)

Fig. 1. Preoperative magnetic resonance images, coronal view, showing a largely cystic subtemporal encephalocele projecting into the sphenoid sinus medially and containing malformed brain extending downward from the inferomedial aspect of the right temporal lobe (arrows).

Fig. 2. Preoperative magnetic resonance images, axial view, showing a largely cystic right subtemporal encephalocele projecting into the sphenoid sinus medially and abutting the maxillary sinus anteriorly. Malformed brain extends inferomedially from the right temporal lobe (arrows).

Fig. 3. Postoperative magnetic resonance image, axial view, showing the extent of the right anterior temporal lobectomy.

typical but shorter than usual. Electroencephalography showed a single seizure manifested by well-developed rhythmic right temporal theta activity lasting approximately 30 seconds. Behavioral testing was not carried out during the initial discharge, but subsequent mild confusion suggested possible impairment of consciousness.

Operation. The patient was readmitted to the hospital in May, 1991, and underwent a right temporal craniotomy. An anterior right temporal lobectomy was performed with the plane of separation 4 cm posterior to the tip. The surgeon noted during removal of the temporal lobe specimen that it was quite adherent to the dura lining the floor of the middle fossa. At this site, two small openings through the dura and skull were discovered. The dura and bone between the two openings were removed, revealing a cystic structure in the infratemporal area. An irregular soft nodule of abnormal brain tissue projected inferiorly from the bone defect in the floor of the middle fossa into a larger cavity lined by gliotic brain tissue and filled with clear, colorless fluid having the appearance of cerebrospinal fluid (CSF). There was no evidence of an epidermoid tumor on inspection at operation or by microscopic examination of the portions of the wall of the structure that were removed. The abnormality was thought to represent a basal encephalocele of the anterior part of the middle fossa. Gelfoam was placed within the cavity, and a graft of temporalis fascia was used to close the dural defect.

On the cortical surface of the excised portion of the temporal lobe there was a roughened defect measuring approximately 2 × 2 cm, corresponding to the area that had been adherent to the dura. The temporal lobe specimen was cut into multiple sections from one end to the other, and representative portions were studied microscopically: no abnormalities were noted. The excised extradural tissue consisted of fibrous connective tissue and malformed brain with gliosis and calcification, consistent with the diagnosis of encephalocele. There was no evidence on microscopic examination that the intradural or extradural tissue removed contained any portion of the hippocampus.

Postoperative Course. The patient had an unremarkable postoperative course. In June and September, 1991, MR imaging showed only the expected postoperative changes (Fig. 3). Review of these films showed the right amygdala and hippocampus to be intact except for the possibility of slight loss anterolaterally. The patient has been free of seizures without anticonvulsant medication for the 18 months since her operation. Based on the indirect evidence presented above, we postulate that her seizure focus was within the anterior inferior temporal cortex at or near its area of attachment to the encephalocele.

Discussion

Classification of Encephaloceles

An extension of cerebral tissue through the dura mater and skull can result from various processes such as traumatic, inflammatory, neoplastic, or surgical disruption of the normal coverings of the brain. A spontaneous encephalocele is one that develops in the ab-
enlargement of the superior orbital fissure or the optic canal, or an area of sphenoid aplasia. Defective development of the sphenoid wings, with resulting extension of temporal lobe tissue into the posterior orbit, can also occur on the basis of neurofibromatosis.4,6-12,37,65

In 1949, Strandberg44 reviewed 31 cases of encephalocele of the posterior part of the orbit that had been reported between 1841 and 1948. There was a 5:3 predominance of females. Of the 25 patients whose ages were given, 22 were under the age of 30 years at first examination. In 20 cases the condition had been accompanied with symptoms since shortly after birth, though to a comparatively slight degree. In 20 cases the bulbus was displaced in relation to its original position in the orbit, being displaced downward and laterally in 18 cases and downward and medially in only 2 cases. In 28 cases there was unilateral exophthalmos, the condition being fairly equally distributed between the right and the left eye. In 26 cases exophthalmos was of pulsating type. Cranial changes were many and characteristic: Enlargement of the orbit on the affected side, in 21 cases... and prominence of the temporal region, in 9 cases. In 4 of these 9 cases, in addition to posterior cephalocele of the orbit, there was cephalocele of the temporal region.45

Anteromedial Encephalocele

The anteromedial encephalocele extends through the anteromedial wall of the middle fossa into the sphenoid sinus and typically presents with CSF rhinorrhea.2,11,22,31,32,44,64 This lateral sphenoidal encephalocele is much less common than the midline varieties; we could find only eight cases in the medical literature. The eight patients varied in age from 41 to 69 years (average 50 years), and seven were women. In six of the eight patients, the encephalocele(s) (multiple in three patients) was on the left side.

Posterior Inferior Temporal Encephalocele

The posteroinferior temporal encephalocele (aural encephalocele) ordinarily projects through the tegmen tympani into the tympanic antrum or epitympanic recess. These typically present with CSF in the middle ear that drains laterally through an opening in the tympanic membrane (sometimes created surgically) as CSF otorrhea, or drains forward via the eustachian tube as CSF rhinorrhea. Diminished auditory acuity in the same ear is a frequent accompaniment, and meningitis may occur.

Of 30 patients with posteroinferior temporal encephalocele reported in the literature, 12 had two or more

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TABLE 1

Spontaneous temporal encephaloceles: typical features

<table>
<thead>
<tr>
<th>Description</th>
<th>Feature</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Lateral</td>
<td>defect at pterion or, occasionally, asterion apparent in infancy, especially in females soft mass on side of head</td>
</tr>
<tr>
<td>2. Anterior (sphen-orbital, posterior orbital)</td>
<td>defect in sphenoid wing area: encephalocele in posterior orbit association with neurofibromatosis, in some patients apparent in infancy or youth, more often in females slowly progressive unilateral pulsating exophthalmos, globe usually displaced laterally</td>
</tr>
<tr>
<td>3. Anteromedial</td>
<td>defect in anteromedial middle fossa, especially on left: encephalocele in sphenoid sinus presents in adult life, especially in females cerebrospinal fluid rhinorrhea, risk of meningitis</td>
</tr>
<tr>
<td>4. Posterior (aural)</td>
<td>defect in tegmen tympani: more often on left: encephalocele (frequently more than one) in tympanic antrum or epitympanic recess presents in adult life cerebrospinal fluid otorhinorrhea or otorhinorrhea, reduced auditory acuity, risk of meningitis</td>
</tr>
<tr>
<td>5. Postero-inferior</td>
<td>defect in postero-inferior middle fossa: encephalocele in infra-temporal region presents in young adulthood, especially in females complex or simple partial seizures</td>
</tr>
</tbody>
</table>

The occurrence of such factors, either because of embryological maldevelopment or from a poorly understood postnatal process that permits brain herniation to occur. Spontaneous basal encephaloceles have been classified in various ways;18,45-55 those of importance to the present discussion fall into four main groups. An additional group, the lateral temporal encephaloceles, is included to complete the list of encephaloceles that involve the temporal lobe. Each type of temporal encephalocele has characteristics that distinguish it from the other four (Table 1).

In the present review, we included only those cases in which there was surgical or pathological documentation of the existence of an encephalocele. Reported instances of brain heterotopia involving the pharynx, orbit, nasal cavity, or maxillofacial region, without a remaining connection to the brain, were not considered.66,67 Likewise, the sphenomaxillary encephalocele reported by Müller, et al.,42 and the spheno-maxillary meningo-encephalocele reported by Morris, et al.,41 were not included because there was no documentation of the origin of the displaced brain tissue. Finally, the case of "temporal encephalocele" reported by Tranmer, et al.,29 could not be classified into one of the present categories because of its large size and its diffuse extension into the subtemporal, facial, and cervical regions.

Lateral Temporal Encephalocele

The lateral temporal encephalocele is so named because it extends laterally through a defect at the pterion (or occasionally at the asterion);18,46,49,56. It is apparent in infancy as a soft mass on the side of the head. At least 15 such cases have been reported: eight of the encephaloceles were on the left side and seven on the right. Of the 13 infants whose sex was specified, 10 were female.

Anterior Temporal Encephalocele

In the second type, the anterior temporal encephalocele (sphen-orbital or posterior orbital encephalocele), there is a developmental defect in the middle fossa anteriorly, through which temporal lobe tissue extends forward into the ipsilateral orbit.6,8,15,27,28,40,49,61,65

This defect represents an enlargement of the superior orbital fissure or the optic canal, or an area of sphenoid aplasia. Defective development of the sphenoid wings, with resulting extension of temporal lobe tissue into the posterior orbit, can also occur on the basis of neurofibromatosis.7,9,12,37,65

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TABLE 2

Spontaneous anteroinferior temporal encephalocele*

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Age (yrs), Sex</th>
<th>Symptoms &amp; Signs</th>
<th>Electroencephalography</th>
<th>Treatment</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ruiz Garcia, 1971</td>
<td>30, F</td>
<td>simple (?) complex partial seizures for 6 mos</td>
<td>lt temporal seizure focus</td>
<td>lt anterior temporal lobectomy with excision of encephalocele</td>
<td>no further seizures, off medication (follow-up period unspecified)</td>
</tr>
<tr>
<td>Hyson, et al., 1984</td>
<td>12, M</td>
<td>simple partial seizures starting at 8 yrs; atomic seizures with loss of consciousness starting 6 mos later; generalized tonic-clonic seizures for 18 mos before admission</td>
<td>bitemporal independent epileptiform discharges; intraop ECoG showed epileptiform activity from middle &amp; posterior rt temporal lobe</td>
<td>rt anterior temporal lobectomy with excision of encephalocele</td>
<td>case complicated by lt temporal head injury at 7 yrs &amp; by a mixed astrocytoma/ependymoma in surgical specimen</td>
</tr>
<tr>
<td>Hyson, et al., 1984 &amp; Leblanc, et al., 1991</td>
<td>37, F</td>
<td>complex partial seizures for 6 yrs</td>
<td>bitemporal interictal epileptogenic foci; onset of seizures from lt temporal lobe by depth electrodes</td>
<td>lt anterior temporal lobectomy and amygdalohippocampectomy with excision of encephalocele</td>
<td>no further seizures (5 yrs)</td>
</tr>
<tr>
<td>Rosenbaum, et al., 1985</td>
<td>38, F</td>
<td>complex partial seizures for 7 yrs</td>
<td>bitemporal independent interictal discharges; rt temporal ictal focus by subdural strip electrodes</td>
<td>rt anterior (6 cm) temporal lobectomy; 1 large encephalocele &amp; about 30 tiny encephaloceles</td>
<td>some auras but no further complex partial seizures &quot;in postoperative period&quot;</td>
</tr>
<tr>
<td>Elster &amp; Branch, 1989</td>
<td>27, F</td>
<td>complex partial seizures for several years</td>
<td>not reported</td>
<td>no surgery; encephalocele on lt shown by MRI</td>
<td>lt-sided headaches recurred at 37 yrs; reap at 52 yrs</td>
</tr>
<tr>
<td>Whiting, et al., 1990</td>
<td>18, F</td>
<td>complex partial seizures for 3 yrs</td>
<td>not reported</td>
<td>lt anterior temporal lobectomy (3 cm along superior temporal gyrus, 5 cm along inferior temporal gyrus, to mid-mesencephalic level mesially) with excision of encephalocele</td>
<td>a few auras but no further seizures (14 mos); encephalocele showed meningioma, angio-astrocytoma - no further seizures (1 yr)</td>
</tr>
<tr>
<td>Leblanc, et al., 1991</td>
<td>36, M</td>
<td>generalized tonic-clonic seizures for 20 yrs</td>
<td>epileptic activity recorded from lt infero-mesial temporal region</td>
<td>lt anterior temporal lobectomy, amygdalohippocampectomy with biopsy &amp; amputation of encephalocele</td>
<td>no further seizures (1 yr)</td>
</tr>
<tr>
<td>Wilkins, et al., 1993</td>
<td>26, M</td>
<td>2 generalized tonic-clonic seizures at 21 yrs; subsequent complex partial seizures</td>
<td>epileptic activity recorded from mesial &amp; anterior surfaces of lt temporal lobe &amp; from lt amygdala</td>
<td>lt anterior temporal lobectomy &amp; amygdalohippocampectomy with excision of encephalocele</td>
<td>no further seizures (1 yr)</td>
</tr>
<tr>
<td>Wilkins, et al., 1993</td>
<td>36, F</td>
<td>partial seizures (some complex) for 23 yrs; generalized seizure at 36 yrs</td>
<td>epileptiform discharges from rt temporal region</td>
<td>rt anterior temporal lobectomy (4 cm) with excision of encephalocele</td>
<td>no further seizures (18 mos)</td>
</tr>
</tbody>
</table>

* ECoG = electrocorticography; MRI = magnetic resonance imaging.

Adjacent encephaloceles. For the 28 patients whose age was given, the range was from 16 to 80 years with an average of 54 years. For the 26 patients whose sex was specified, 12 were male and 14 female. The encephalocele(s) was on the left side in 16 patients and the right in 10; in four the side was not specified.

Anteroinferior Temporal Encephalocele

The fifth type of temporal encephalocele, typified by the present case, projects through the anterior floor into the infratemporal region. Of the 11 reported patients with this type of anteroinferior temporal encephalocele, eight presented with medically intractable complex or simple partial seizures (Table 2). Eight of
the 11 patients were female and eight were between 26 and 38 years old (with the remainder being younger). Six of the encephaloceles were on the left side and five were on the right. All eight patients who had seizures preoperatively and for whom postoperative information was available noted improvement in their seizures, with six being free of seizures during a follow-up period of up to 5 years. The present case is of interest in that complete seizure control was achieved with resection of the lesion alone and with no attempt at amygdalo-hippocampectomy.

Etiology of Basal Middle Fossa Encephaloceles

Two theories have been advanced to explain the presence of encephaloceles in the base of the middle fossa. The first suggests that the encephalocele results from defective embryological development of that portion of the skull and its adjacent tissues.

According to Elster and Branch,17 “the ossification of the sphenoid bone is complex, with up to 19 separate ossification centers. . . . The greater wing ossifies in both cartilage and membrane. At about the 8th week of fetal life a cartilaginous ossification center appears near the foramen rotundum. Only this root of the greater wing of the sphenoid bone is ossified in cartilage; the rest of the greater wing and pterygoid plates ossify in membrane.” And, as summarized by Hyson, et al.,18 “just before the end of the fetal period, the temporal bone consists of three principal parts: the squama, ossified in membrane from a single nucleus, which appears about the second month; the petromastoid part, developed from four centers, which make their appearance about the fifth and sixth month; and the tympanic ring, an incomplete circle, ossified in membrane from a single center, which appears about the third month. The tympanic ring unites with the squama shortly before birth; the petromastoid part and squama join during the first year.”

If this complex process of formation of the base of the skull does not follow its usual pattern, one or more gaps could occur, permitting outward herniation of the adjacent portion of the temporal lobe and its investing membranes during fetal development. Other factors, such as intermittent increases in intracranial pressure, might also take part in such a process.

Concerning the anteroinferior temporal encephalocele, Leblanc, et al.39 postulated that the bony defect “. . . results from a failure of chondrification and ossification of the base of the greater sphenoid wing which normally would produce the pterygoid process, foramen rotundum, and lingula.” They also concluded that “such a developmental defect must occur before the 2nd intrauterine month when chondrification begins.”39

An alternative explanation for the occurrence of basal temporal encephaloceles is that one or more small defects in the bone allow the gradual postnatal development of cerebral herniation. This concept is based on the fact that small pits or bony dehiscences are frequently seen along the floor of the middle fossa at routine autopsy examinations.14,19,25,31,32 For example, Ahren and Thuilin1 investigated the temporal bones of 94 subjects at autopsy and found that 21% had bony defects in the tegmen tympani, with 15% having less than five defects and 6% having between five and 10 defects. In addition, another 16% had only thin, transparent cortical bone covering the air cells of the tegmen tympani. Of more potential significance to the present case, Kaufman, et al.31,33 demonstrated pits in the anteromedial aspects of the middle fossa floor, occasionally involving a lateral extension of the sphenoid sinus. They postulated that “pneumatization of the middle fossa floor is essential to the development of bony dehiscence. Normal pressure and pulsatile forces of the CSF and brain may then lead to additional thinning of the bone from the intracranial surface, the development of small holes or enlargement of normal pit holes . . . and prolongations of the dura and the arachnoid or of the dura, arachnoid, and brain through these holes, resulting in acquired meningoceles or meningoencephaloceles. With fixation of these prolongations, the dura becomes thinner and thinner until fenestration occurs, destroying the barrier.”31

Conclusions

The present review does not answer questions about why and how temporal encephaloceles occur. However, it is apparent that there are at least five types of temporal encephalocele. Each has typical clinical characteristics and each can be diagnosed easily with the aid of modern radiological techniques. Furthermore, their detrimental accompaniments such as meningitis, proptosis, or partial seizures ordinarily can be prevented or treated by an appropriate surgical procedure.

Acknowledgment

The authors thank Robert D. Tien, M.D., of the Department of Radiology for reviewing the CT and MR studies.

References


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demonstration. AJNR 5:820–821, 1984

Manuscript received May 14, 1992.
Accepted in final form July 28, 1992.
This work was presented in part at the Annual Meeting of the Southern Neurosurgical Society, Washington, D.C., on April 3, 1992.
This work was presented in full at the First International Skull Base Congress, Hannover, Germany, June 14–20, 1992.
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