Microencephaloceles: another dual pathology of intractable temporal lobe epilepsy in childhood

Case report

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Several reports describe the association of temporal lobe epilepsy with encephaloceles of the middle cranial fossa.3,6,9,13–19 All reported cases exhibited definite temporal lobe herniations through defects in the bone constituting the anteroinferior part of the middle cranial fossa floor, occurring in young to middle-aged adults. We present the case of an adolescent with multiple microencephaloceles, in the anterolateral middle fossa floor, identified at surgery (temporal lobectomy) for intractable partial-onset seizures of temporal origin. Magnetic resonance imaging revealed only hippocampal atrophy. Subdural electrodes demonstrated ictal activity arising primarily from the anterior and lateral temporal lobe, close to the microencephaloceles, spreading to the anterior and posterior mesial structures. Pathological examination revealed diffuse temporal gliosis involving the hippocampus, together with microdysgenesis of the amygdala. The literature on epilepsy secondary to encephaloceles is reviewed and the contribution of the microencephaloceles to the seizure disorder in this patient is discussed. (DOI: 10.3171/2009.11.PEDS09275)

KEY WORDS • temporal lobe epilepsy • microencephalocele • dual pathology

Abbreviations used in this paper: EEG = electroencephalography; MEG = magnetoencephalography.
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**Histological Evaluation.** Histological examination demonstrated diffuse chronic inflammation and gliosis throughout the cortex and white matter of the left temporal lobe. Gliosis and perivascular chronic inflammation were also evident in the hippocampus; neuronal loss in CA1 was not prominent. The amygdala exhibited gliosis and foci of microdysgenesis.

**Discussion**

The association between medically refractory epilepsy and encephaloceles is well documented for both the temporal and frontal skull base as well as for the skull vault. A Medline literature search using the key words “epilepsy,” “temporal,” and “encephalocele,” from 1966 to 2009, together with a review of the bibliography of articles obtained, yielded 10 publications reporting on 13 patients.6,9,13–19 One patient was a 12-year-old boy in whom, in the resected temporal lobe, a glioma was also found;6 he is excluded from this discussion. There were 9 females and 3 males, between 18 and 46 years of age (median 33 years). Six encephaloceles occurred on the right, 6 on the left, and 1 was bilateral. Seizures were complex partial in 8, secondarily generalized in 1, and a combination in 2. In 1 patient, only abnormal epileptiform activity on electroencephalography was identified.9 Prior to diagnosis, seizures were present for between 5 and 25 years (median 13.5 years).

In all of these patients clear, well-defined encephaloceles were demonstrated, with a documented defect in the bone of the middle fossa floor. The smallest lesion was 1 cm in diameter.8 All except 2 cases were diagnosed preoperatively using CT scanning or MR imaging.14 Most encephaloceles were located in the anteromedial region of the middle fossa floor, at the base of the pterygoid process, anterolateral to the foramen rotundum and anterior to the Meckel cave. The lesions projected toward, or extended into, the sphenoid sinus and pterygopalatine fossa. In 1 patient, the encephalocele projected through the ala of the sphenoid and was associated with posterior displacement of the carotid artery.8 In another patient, a large right temporal encephalocele was associated with several smaller ones.14 One case was exceptional in that the encephaloceles occurred through bony defects, 2 and 5 cm in diameter respectively, in the tegmen of the temporal bone on both sides; these were associated with CSF otorrhea.19 In contrast to these descriptions, in the case we describe, the encephaloceles were smaller, up to 4 mm in maximal diameter, and only 2 mm deep; they were located in the anterolateral region of the middle fossa floor (Fig. 3A). They occurred through dural defects, and we were unable to demonstrate a clear defect in the bone of the skull base. They were too small to be appreciated on thin-section MR imaging using epilepsy protocols and could only be visualized adequately under the operating microscope.

In the literature reviewed, 7 patients underwent temporal lobectomy, including resection of the amygdala and hippocampus.9,14,15,17 Amputation and resection of the encephalocele was also conducted. One patient underwent neocortical temporal lobectomy only.18 In another patient, a large temporal encephalocele was approached and re-

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**Fig. 1.** Coronal MR image, FLAIR sequence, showing the smaller hippocampus on the left (arrow), with a larger temporal horn on the left than the right.

**Operation and Postoperative Course.** A left temporal lobectomy, including amygdalohippocampectomy, guided by intraoperative electrocorticography, was performed. The posterior superior temporal gyrus was preserved. On reflection of the anterior half of the inferior temporal gyrus from the skull base, we observed that the temporal lobe was adherent to the dura covering the anterolateral region of the middle cranial fossa floor. Gentle traction revealed 3 strands of tough, scarred brain tissue, extending into small perforations in the dura. The largest dural defect measured up to 4 mm in diameter. Microencephaloceles, up to 2 mm deep, could be seen within these 3 dural pockets (Fig. 3). The patient's postoperative course was uneventful; she is on a single antiepileptic drug and has been seizure free for 9 months.
sected extradurally, with primary repair of the dura. In the patient with herniation through the tegmentum, the encephaloceles were evacuated from the mastoid cavities and the tegmental defects repaired with bone and fascia. In 1 patient, surgery was not performed prior to publication, and in another treatment and outcome details were not given.

Pial and arachnoid adhesions were identified at the site of herniation in several cases. In 1 patient, the dura around the encephalocele was thickened and reactive. Histologically, gliosis was evident and extended from the encephalocele into the amygdala, hippocampus, and uncus in some patients. The temporal neocortex in proximity to the encephalocele was not always histologically abnormal. One encephalocele exhibited meningoangiomaticosis.

Two groups performed temporal lobectomy with amygdalohippocampectomy and reported freedom from seizures for up to 5 years postoperatively (range 1–5 years). The patient in whom resection of the amygdala and hippocampus was not performed was also seizure free 18 months after surgery, despite a 23-year history of complex partial seizures; this would suggest that the

Fig. 2. A: Ictal scalp EEG showing onset of seizure activity from T1, F7, and T3 electrodes over the anterior and middle left temporal cortex (arrow). B: Source estimates of spontaneous activity obtained on MEG, projected onto axial and coronal structural MR images, demonstrating interictal discharges arising from the left temporal lobe. C: Operative photograph of left temporal lobe exposed through a temporal craniotomy; a 32-channel subdural grid has been placed over the lateral temporal surface; anterior and posterior subtemporal, inferior frontal, orbitofrontal, and parietal (reference) strips are also evident. D: Ictal subdural EEG demonstrating seizure onset from anterior and middle temporal cortex (Electrodes 4–6 and 13–15, arrows); the subtemporal electrodes do not show any seizure activity at this time. E: Ictal subdural EEG recorded 3 minutes after that in D, showing initiation of seizure activity in the anterior subtemporal region (arrow). AST = anterior subtemporal; G = grid; IF = inferior frontal; OF = orbitofrontal; PST = posterior subtemporal.
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Fig. 3. A: The skull base showing the region where the microencephaloceles report were located (large arrow) in relation to the location of the anteroinferior encephaloceles described in the literature (small arrow). B and C: Operative images of one of the attachments of the inferior surface of the temporal lobe to the dura (B, arrow), where it was tethered across the dural defect to the herniated brain situated subdurally (C, arrow). D: Operative image, with arrows pointing to the 4 dural defects, evident once the temporal lobectomy was completed.

mesial temporal structures are not always necessarily involved in epilepsy related to temporal encephaloceles.

Possible explanations for encephaloceles in the temporal and sphenoid area have been reviewed. Briefly, the temporal and sphenoid bones ossify in both membrane and cartilage from multiple ossification centers. Interruptions to this complex process could leave gaps between bone segments, allowing herniation of meninges and temporal lobe parenchyma. Another possibility is suggested by the presence of multiple small “pit holes” often identified in the anterior middle cranial fossa in adult skulls. These defects may be related to enlargement of normally occurring arachnoid villi or draining veins that penetrate the middle fossa floor in this region. The dura in this area often appears to be fenestrated. It has been argued that physiological elevations and fluctuations in intracranial pressure encourage the development of herniation through the floor. The location and size of the dural defects identified in our patient correspond to those found in the literature.

Our patient correspond to those found in the literature. We discuss the case of a patient with multiple middle cranial fossa microencephaloceles associated with intractable temporal lobe epilepsy. Temporal lobe herniations were small and occurred through small dural defects, probably congenital, near the temporal pole; they were not identifiable on preoperative imaging. They were associated with chronic inflammation and gliosis of the temporal lobe, including the mesial structures. This case suggests that even small basal temporal encephaloceles, herniating only through small dural defects, rather than significant herniations associated with bone defects in the middle fossa floor, may be associated with an intractable seizure disorder; this may be particularly relevant to the pediatric population.

Conclusions

We discuss the case of a patient with multiple middle cranial fossa microencephaloceles associated with intractable temporal lobe epilepsy. Temporal lobe herniations were small and occurred through small dural defects, probably congenital, near the temporal pole; they were not identifiable on preoperative imaging. They were associated with chronic inflammation and gliosis of the temporal lobe, including the mesial structures. This case suggests that even small basal temporal encephaloceles, herniating only through small dural defects, rather than significant herniations associated with bone defects in the middle fossa floor, may be associated with an intractable seizure disorder; this may be particularly relevant to the pediatric population.

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