Spontaneous motor cortex encephalocele presenting with simple partial seizures and progressive hemiparesis

Case report and review of the literature

KOSTAS N. FOUNTAS, M.D., PH.D., JOSEPH R. SMITH, M.D., PATRICK D. JENKINS, PH.D., AND ANTHONY M. MURRO, M.D.

Departments of Neurosurgery and Neurology, Medical College of Georgia, Macon, Georgia

Several cases of congenital or acquired temporal encephaloceles have been reported in the literature as the causative mechanism of simple and/or complex partial seizures. In this report the authors describe a rare case of spontaneous parietal encephalocele presenting with simple partial seizures and progressively increasing contralateral upper-extremity motor deficit. The unusual anatomical location of an encephalocele associated with seizures and the delayed seizure onset represent distinctive characteristics in this case. Preoperative imaging included surface electroencephalography, computerized tomography, and brain magnetic resonance imaging. Frameless neuronavigation and intraoperative cortical mapping were used to aid resection of the encephalocele, and the dural and bone defects were reconstructed. The surgical outcome in this case was excellent, and the patient has remained seizure free. The pertinent literature is reviewed in this report.

KEY WORDS • encephalocele • motor cortex • simple partial seizure • neuronavigation • cortical mapping

An association between seizures and encephaloceles, although rare, is well established. There are several reports in the literature describing cases of medically intractable epilepsy, in which the operative findings revealed that small encephaloceles were the epileptogenic focus. In the vast majority of these cases the underlying encephaloceles had remained undiagnosed for prolonged periods and were intraoperatively identified as the cause of usually complex partial seizures. However, a few cases of acquired encephaloceles associated with medically refractory seizures have also been described.

In this communication we report a case of spontaneous motor cortex encephalocele, which presented with progressive hemiparesis and simple partial seizures and was successfully treated at our institution. In addition, we review the pertinent literature.

CASE REPORT

History and Examination. This 61-year-old Caucasian, right-handed woman was evaluated in our epilepsy clinic for seizures marked by an inability to use her left hand, followed by left thumb jerking that progressed to generalized tonic-clonic seizures. Her seizures had begun 5 years previously and progressively increased in frequency. Initial evaluation revealed dysarthria, left-sided facial weakness, and dysphagia that had started approximately 3 months earlier and had been worsening. Her neurological examination revealed mild facial weakness (motor strength 4+/5) with no other focal abnormalities. She was evaluated periodically in the adult epilepsy clinic for several years. Initially, there was no report of a change in neurological status. On her last visit before surgery, however, the patient reported a slight but definite increase in gripping strength in her left hand. Serial MR images demonstrated no progression of the gliosis or traction on the right lateral ventricle (Fig. 1).

The patient’s medical history revealed no concussion, birth complications, febrile seizures, central nervous system infection, stroke, or brain tumor. Her family history was unremarkable. She was currently receiving 1500 mg levetiracetam per day, and several other anticonvulsive medications (valproate, phenytoin, ethosuximide, and carbamazepine) had been tried previously. Her seizures had been well controlled by her latest medication until recently.
Neuroimaging. Her laboratory workup included surface interictal electroencephalography and brain MR imaging. Her electroencephalogram demonstrated no abnormalities. Serial MR imaging studies revealed a right parietal defect of the inner table of the skull, with an associated protruding encephalocele. There was also an associated gliotic track creating traction on the right lateral ventricle.

The imaging and electrophysiological findings were extensively discussed with the patient and the option of resection of her encephalocele was offered to her. After considering the advantages and disadvantages of such a surgical intervention, the patient decided to proceed with resection.

Operation. After endotracheal induction of general anesthesia, the patient was placed on the operating table in a left semidecubitus position with her torso rolled up approximately 40°. The patient’s head was secured in a three-point Mayfield fixation device. Frameless stereotaxy (BrainLAB AG, Heimstetten, Germany) was applied for localization of the encephalocele and of what we thought was motor cortex (based solely on anatomical criteria; Fig. 1). A 10-cm linear skin incision extending from the midline longitudinally to a point just in front of and above her right ear was centered over the encephalocele. A high-speed air drill (Midas Rex; Medtronic, Inc., Minneapolis, MN) was used to make a doughnut-shaped craniectomy circumferentially to the lesion, after meticulous dissection of the exposed encephalocele from the adjacent bone edges (Fig. 2A). The exposed underlying normal-appearing dura mater was cruciated around the encephalocele, and after meticulous dissection the stalk of the lesion was identified (Fig. 2B).

Subsequently, intraoperative corticography was performed and revealed no epileptogenic activity in the area of encephalocele. At that point, cortical motor mapping using direct stimulation was performed. Stimulation (pulse width 1 msec, 4 V, 60 Hz) of a site immediately superior to the encephalocele caused flexion of all the fingers of the left hand (Fig. 3A).

After completion of the motor mapping, the stalk of the encephalocele was transected en bloc by using bipolar cautery, and was then sent for pathological examination (Fig. 3B). Hemostasis was obtained and the overlying dura mater was closed using 4-0 silk sutures (Fig. 4A). When that was done, a 5 × 6-cm piece of polysorbate mesh was secured with 4-mm absorbable screws. After that, bone putty was used to fill in the cranial defect and then a second outer plate of polysorbate mesh (of the same dimensions) was secured over the bone putty with 4-mm absorbable screws (Fig. 4B). The surgical wound was closed in anatomical layers.
Pathological examination of the resected specimen revealed gliotic and edematous cerebral cortex consistent with encephalocele.

Postoperative Course. The patient was observed in the intensive care unit for 24 hours and then was transported to the neurosurgical ward for 48 hours, after which she was discharged home. She has remained seizure free (26-month follow-up duration) with no evidence of focal neurological deficits and no other symptoms.

DISCUSSION

Congenital encephaloceles represent a quite rare neural tube defect; the incidence has been reported to be 1:5000 live births, but it might be even less frequent in North America.\(^7,12\) The incidence of encephaloceles in specific anatomical locations varies significantly according to sex,
with occipital encephaloceles more common among female patients, whereas anterior lesions are more common among males.\textsuperscript{7,12} Several classification systems have been proposed based on the anatomical location of the encephalocele;\textsuperscript{6} the most widely accepted and simple one appears to be that proposed by Matson.\textsuperscript{10} According to his classification system, encephaloceles can be separated into basal, sincipital, convexity, and atretic ones.\textsuperscript{10} Each of these groups can be further subdivided into subgroups (Table 1). The most commonly found among these groups is the convexity encephalocele, as in our parietal case.\textsuperscript{7}

Encephaloceles have been clearly associated with seizures, particularly in lesions located in the temporal lobe. In their case report, Wilkins, et al.\textsuperscript{17} presented a 36-year-old patient with a bilateral temporal encephalocele in patients with complex partial seizures after undergoing a surgical procedure for resection of a right maxillary sinus polyp and an ethmoidectomy 2 weeks earlier. Acquired temporal or frontal encephaloceles may occur as a consequence of surgical procedures, traumatic injuries, or infections.\textsuperscript{2,5,13}

Regarding the diagnosis of encephaloceles, it needs to be emphasized that in a significant number of the reported cases the diagnosis was established intraoperatively; their presence had not been suspected preoperatively.\textsuperscript{14,15,17} The recent advances in neuroimaging, along with increasing suspicion of this rare but definitely existent pathological entity can enable surgeons to identify these lesions preoperatively. The synergistic contribution of CT and MR imaging to the preoperative workup in patients with partial complex seizures or in high-risk patients who have undergone previous surgical procedures or who have experienced multiple episodes of recurrent meningitis cannot be overemphasized.\textsuperscript{5} In our patient the anatomical location of the encephalocele, along with the alarming clinical symptomatology, significantly facilitated its diagnosis. Nevertheless, in cases of small temporal encephaloceles, their diagnosis might be challenging despite high-resolution MR imaging and CT studies.

In regard to the surgical management of encephaloceles in adults presenting with seizures, this appears to be less challenging than the treatment of these lesions in neonates. Blood loss is usually better tolerated in adults and the size of these lesions is significantly smaller than the ones present in neonates.\textsuperscript{6} Nevertheless, particular attention needs to be paid to the postoperative functional status of the patient while planning the resection of such lesions, as was done in our case. Our patient’s encephalocele was critically located in the proximity of the hand motor area; this was implied by her presenting clinical symptomatology and confirmed by intraoperative cortical mapping. Additional preoperative functional imaging such as functional MR imaging could be beneficial in the surgical planning for these procedures. In our patient, magnetic source imaging of evoked sensory potentials verified that the hand sensory cortex was immediately posterior to the encephalocele in the motor cortex.

The use of a frameless neuronavigational system further enhances the accuracy of the procedure and helps not only in identifying the lesion through a smaller exposure but also in localizing the preregistered adjacent functional areas. Intraoperative cortical mapping can delineate the relationship of the encephalocele with the motor cortex and further minimize the chance of postoperative focal deficits. The importance of meticulous dissection of the encephalocele

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**TABLE 1**

Classification scheme for encephaloceles proposed by Matson

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<thead>
<tr>
<th>Type</th>
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<tr>
<td>basal</td>
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<tr>
<td>sphenopharyngeal</td>
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<td>intranasal</td>
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<td>sphenoorbital</td>
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<tr>
<td>sphenomaxillary</td>
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<tr>
<td>sincipital</td>
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<tr>
<td>nasofrontal</td>
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<tr>
<td>frontoethmoidal</td>
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<tr>
<td>nasoorbital</td>
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<tr>
<td>convexity</td>
</tr>
<tr>
<td>frontal</td>
</tr>
<tr>
<td>parietal (or sagittal)</td>
</tr>
<tr>
<td>occipital</td>
</tr>
<tr>
<td>cervicooccipital</td>
</tr>
<tr>
<td>atretic</td>
</tr>
<tr>
<td>alopecic (parietal)</td>
</tr>
<tr>
<td>nodular (occipital)</td>
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from the adjacent bone and dural defect (as was the case in our patient) cannot be overemphasized. Finally, reconstruction of the dural (if present) and bone defects can be easily accomplished in these cases, as was demonstrated in our patient.

CONCLUSIONS

Spontaneous or acquired encephaloceles constitute a rare but known pathological entity in patients with simple or complex partial seizures. Temporal encephaloceles represent the vast majority of the reported cases, but a parietal anatomical location, as in our patient, may also be observed. A high index of suspicion, especially in patients with previous ear, nose, and throat procedures; trauma; middle ear infections; and episodes of recurrent meningitis, along with high-resolution CT and MR studies, can facilitate detection of the presence of an encephalocele. Thorough preoperative planning and meticulous dissection during resection and reconstruction of the bone defect ensure good functional outcome in the vast majority of these patients.

Disclaimer

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References


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