Isolated, occult encephalocele as unique cause of refractory temporal lobe epilepsy. A case report

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Abstract
Encephalo-meningocele consists of a bony defect of the skull base in middle cranial fossa. This abnormality has been rarely described in patients with temporal lobe epilepsy (TLE), although its epileptogenic role is still debated. Patients with refractory TLE and encephalocele may undergo anterior temporal lobectomy or lesionectomy. We describe the case of a 20-year-old man with refractory TLE and occult temporal pole encephalocele, who underwent lesionectomy leading to seizure-freedom after a one-year follow-up. Lesionectomy may be an effective and safe treatment when isolated encephalocele is the unique cause of refractory TLE.

KEY WORDS: focal epilepsy, encephalocele, temporal lobe epilepsy, epilepsy surgery.

Introduction
Temporal pole encephalocele (TPE) is a rare pathological protrusion of brain parenchyma through congenital or acquired (traumatic, neoplastic, metabolic, or infectious damage of the skull base) defects of the skull base in the middle cranial fossa (1-3). Along with advances in neuroimaging techniques, congenital TPE is increasingly identified as a potential cause of temporal lobe epilepsy (TLE) in patients initially classified as lesion negative (occult encephalocele) (4, 5). The aim of this paper is to describe the electro-clinical features of a patient with drug-resistant TLE and occult TPE.

Case report
A 20-year-old right-handed man came to our observation at age of 18, complaining of monthly seizures that had started at the age of 17. Seizures were characterized by undefined “malaise feeling” followed by speech difficulties (expressive and receptive aphasia), lasting a few seconds. Post-ictal symptoms included headache, confusion and drowsiness. Rarely, the patient experienced tonic-clonic seizures during sleep. Physical and neurological examinations were normal, whereas neuropsychological assessment showed only a mild deficit in long-term verbal memory. He underwent 1.5 T brain MRI, reported as normal and was initially treated with valproic acid up to 1000 mg/day for 6 months, without benefit. Adjunctive treatment with levetiracetam (2 gr/day) and, subsequently, eslicarbazepine (800 mg/day) did not modify seizure frequency. At age of 19 he was evaluated for epilepsy surgery. Prolonged video-EEG with 10-20 system and supplementary zygomatic electrodes showed interictal spike-wave complexes over left temporal leads. We also recorded one of his habitual seizures during sleep, clinically manifesting with gaze and head deviation towards right, followed 10 seconds later by tonic-clonic manifestations. On EEG, 27 seconds before clinical onset, we recorded a diffuse flattening of electrical activity lasting 5 seconds, followed by a theta-delta rhythmic activity gradually increasing in frequency and amplitude, over Fp1-F3, F3-C3, Fp1-F7, F7-T3, T3-T5, T5-O1, T2-T1, T1-T3, T3-C3 lasting about 30 seconds. High-resolution CT scan (Fig. 2 a) showed a small bony defect in the mesial portion of the left great wing of sphenoid bone. A MRI was targeted to temporal pole and included T2-weighted, 3D-fluid attenuated inversion recovery, fast imaging employing steady-state acquisition and volumetric T1-weighted thin-layer sequences. A tiny encephalocele was then evidenced (Fig. 2 b) in the temporal pole, without other abnormalities. Since the personal history was negative for trauma, neoplasms or infectious diseases of the skull, a
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Figure 1 - Ictal EEG recording. A diffuse flattening of electrical activity is followed by a theta-delta rhythmic activity gradually increasing in frequency and amplitude, over Fp1-F3, F3-C3, Fp1-F7, F7-T3, T3-T5, T5-O1, T2-T1, T1-T3, T3-C3.

Figure 2 - Preoperative brain imaging. A. High-resolution CT scan showing a small bony defect in the mesial portion of the left great wing of sphenoid bone. B. MRI, coronal FLAIR sequence, showing tiny encephalocele in the temporal pole.

The congenital origin was probable. A fluoro-deoxyglucose-PET showed mild hypometabolism in left temporal lobe. Lesionectomy and bone repair were performed during an uncomplicated surgical procedure. Neuropathology showed mild chronic inflammation and gliosis, whereas cortical organization was normal. Control MRI performed a few days after surgery showed the repair of the bony defect. At twelve-month follow-up, the patient was seizure-free in monotherapy with eslicarbazepine (400 mg/day). Neuropsychological functioning was unchanged.

Discussion

We described a young man with drug-resistant TLE due to congenital, occult TPE, who has become seizure-free after lesionectomy. TPE is increasingly identified in refractory TLE. To our knowledge, 74 patients with TLE and TPE have been described so far (4-20). Of these, 45 underwent epilepsy surgery. Anterior temporal lobectomy (ATL) was performed in 35 lesionectomy or local disconnection in the remaining 10. Neuropathology almost invariably showed normal tissue or aspecific gliosis and did not document the presence of other epileptogenic lesions. Of note, in many of these patients, conventional pre-surgical brain imaging did not reveal the lesion and TPE was a chance finding during intervention. Both ATL and lesionectomy were highly effective as most patients achieved Engel Ia class, and no major outcome differences were found between the two procedures. These clinical and pathologic data support the epileptogenic role of TPE. On the other hand, Saavalainen et al. (18) described 5 patients with bilateral TPE who underwent unilateral ATL, resulting in seizure freedom in 3, thus
suggesting that some TPE may remain asymptomatic. As regards surgical approach, lesionectomy, a more sparing procedure, has proven to be as effective as ATL (6, 10, 11, 14, 18-20) and it may lead to a better neuropsychological outcome, particularly on verbal memory (4). In conclusion, the case described here further suggest that isolated, occult TPE may have an epileptogenic role and its presence should be actively searched in all patients with refractory TLE. Epilepsy surgery should be considered in patients with TPE when electroclinical and imaging data indicate a well-localized epileptogenic focus corresponding with TPE. In those cases, lesionectomy appear to be effective and safe, and may represent a valid option.

References