Successful surgical outcome in a patient with coexistence of focal epilepsy due to DNET and Jeavons Syndrome

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Abstract

The coexistence of symptomatic and idiopathic epilepsy has rarely been described and may pose diagnostic and therapeutic challenges. We herein describe a patient with focal epilepsy due to dysembryoblastic neuro-epithelial tumors (DNET) associated with Jeavons Syndrome, who underwent successful lesionectomy. At the age of 13 years, this boy presented rare generalized tonic-clonic seizures controlled by valproic acid. Two years later, dyscognitive focal seizures occurred. Eyelid myoclonia with or without absences (EMA) occurring after eye-closure, photoparoxysmal response and focal seizures arising from left temporal lobe were recorded by video-EEG. Brain MRI revealed a left temporal DNET. The combination of different antiepileptic drugs gave no benefit. Due to drug-resistant disabling temporal lobe seizures, patient underwent lesionectomy. At 33-month follow-up, patient was free from temporal lobe seizures. Notably, despite the persistence of EMA, patient’s quality of life greatly improved. This report highlights the importance to consider surgery even in patients with coexistence of both symptomatic and idiopathic epilepsies.

KEY WORDS: epilepsy surgery, double pathology, eyelid myoclonia, photosensitivity, quality of life.

Introduction

The coexistence of symptomatic and idiopathic epilepsies has rarely been described (1-7) and may pose diagnostic and therapeutic challenges. Dysembryoblastic neuro-epithelial tumors (DNET) represent an important cause of refractory focal epilepsies. Surgery for DNET usually results in high seizure-freedom rate, with 70-90% of favorable outcomes (8). Jeavons Syndrome, a condition not yet recognized by ILAE Commission, is characterized by: 1) eyelid myoclonia with and without absences (EMA); 2) eye closure-induced seizures, EEG paroxysms, or both; 3) photosensitivity (9, 10). We herein describe a patient with the coexistence of both drug-resistant focal epilepsy due to DNET and Jeavons Syndrome. In this subject, the surgical removal of DNET allowed freedom from temporal lobe seizures and improvement of quality of life (QoL) despite persistence of EMA.

Case report

This previously healthy 19-year-old boy presented sporadic apparently generalized tonic-clonic seizures (GTCS), at time occurring while he was watching television, since the age of 13 years. Treatment with valproic acid (VPA) up to 1250 mg/day was started at 15 years, with full control of his GTCS. Two years later he came to our observation due to the occurrence of monthly seizures characterized by déjà-vu, unmotivated fear, palpitations, followed by confusion and drowsiness. Neurological as well as neuropsychological evaluations were within normal limits. Video-EEG recording disclosed EMA, occurring after eye-closure, associated with diffuse polyspike-wave discharges, predominant over posterior head regions (Fig. 1a). Intermittent photic stimulation provoked generalized photoparoxysmal responses (PPR) at frequencies ranging from 9 to 35 Hz (Fig. 1b). The use of optical filter lens abolished photoparoxysmal response. Interictal EEG also showed epileptic abnormalities over the right temporal regions. Brain MRI revealed a multicys-
tic lesion, characterised by hypointense signal in T1 and hyperintense signal in T2-FLAIR sequences, without contrast enhancement, in the left temporal mesial region, in keeping with DNET (Fig. 2a). Combination of different antiepileptic drugs (AEDs) (carbamazepine, lamotrigine, levetiracetam, lacosamide, VPA) gave no benefit. Patient was then admitted to our unit for a long-term video-EEG monitoring. Three seizures characterized by déjà-vu, tachycardia and profuse sweating, associated with left temporal ictal discharges, were recorded (Fig. 1c). Eye closure-induced epileptic discharges and EMA were also recorded. QoL was assessed by means of QoL in Epilepsy Inventory (QOLIE)-31 (Global score: 57). Because of refractory temporal lobe seizures, patient underwent surgery with full resection of DNET (Fig. 2b). Histopathological examination confirmed grade 1 DNET. At 33-month follow-up, patient was free from temporal lobe seizures, although EMA persisted despite VPA 1250 mg/day, and his QoL much improved (QOLIE-31 global score: 82).

Discussion
To our knowledge, this is the first patient with surgical proven coexistence of symptomatic focal epilepsy due to DNET and Jeavons Syndrome. Moreover, this report emphasizes that the presence of these two different seizure disorders do not represent a contraindication for surgery, since patient had a striking improvement of his QoL after DNET removal, despite EMA persistence. Jeavons Syndrome is not yet characterized by new ILAE diagnostic scheme (11) since EMA may occur in several epileptic conditions of idiopathic, cryptogenic, and symptomatic origin (12). Jeavons Syndrome has been proposed as a form of idiopathic generalized epilepsy (IGE) (10, 13, 14) although an occipital generator for generalized seizures and photosensitivity has been proposed (15-17). The presence of EMA, eye closure-induced seizures or EEG paroxysms, and photosensitivity, characterizes this peculiar condition with onset in childhood and seizure persistence over the years. Our subject fulfilled the proposed criteria for Jeavons syndrome (9) presenting EMA and epileptic abnormalities occurring after eye-closure, as well as photosensitivity, that persisted at follow-up. Moreover, our patient also had drug-resistant temporal lobe seizures due to DNET. Ictal EEG showed focal seizures arising from left temporal lobe. After DNET removal, the patient obtained a full control of his temporal lobe seizures. It is of paramount importance to recognize the coexistence of both idiopathic and focal symptomatic epilepsies since it can cause unique diagnostic and therapeutic chal-

Figure 1 - a) EEG showing diffuse polyspike-wave discharges, associated with eyelid myoclonia, immediately after eye-closure; b) generalized PPR provoked by intermittent photic stimulation at 21 Hz; c) EEG performed with additional zygomatic electrodes (T1: left zygomatic; T2: right zygomatic). Note the left temporal ictal discharge.
lenges (4). Indeed, it could be difficult to hypothesize an idiopathic seizure disorder in a patient with clear-cut focal symptomatic epilepsy. Moreover, some drugs used for focal seizures (carbamazepine, vigabatrin, phenytoin, tiagabine, lacosamide) may aggravate generalized seizures (18-20) but this was not the case in our subject. Of course, AEDs with broad-spectrum activity against both focal and generalized seizures such as VPA, lamotrigine, topiramate, levetiracetam and phenobarbital are preferable in patients with coexistence of both generalized and focal seizures. Good seizure control can be achieved with surgical treatment for cases of medically intractable focal seizures coexisting with idiopathic epilepsies (2-5). In our patient, despite the coexistence of Jeavons Syndrome with focal epilepsy cause by DNET, surgery was decided since the temporal lobe seizures were the most disabling. The surgical resection of DNET allowed to completely control the temporal lobe seizures but, as expected, EMA and photoparoxysmal response persisted at follow-up.

Conclusions

This report highlights the importance of considering surgery even in patients with coexistence of both focal symptomatic and idiopathic epilepsies, when drug-resistant focal seizures are disabling and epileptogenic zone is clearly demonstrated.

References