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Clinical letter

Encephalopathy with electrical status epilepticus during sleep: Cognitive and executive improvement after epilepsy surgery

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1. Introduction

Electrical status epilepticus in sleep (ESES) is associated with neuropsychological disturbances, affecting cognitive and behavioural or socio-emotional domains. Attention deficit, impulsivity, lack of inhibition, difficulties in behavioural monitoring and regulation, diminished or scarcely modulated affective responses, apathy, poor social judgement, inadequate impulse control, impaired self-consciousness and poor interpersonal abilities, are indicators of executive dysfunction which variably express in children with ESES.

Perinatal vascular and other congenital brain lesions co-involving the thalamus are common aetiologies.[1] Good response may be obtained with drugs, but ESES and progressive cognitive/behavioural decline sometimes persist until the midteens and residual neuropsychological sequelae are common. Surgery is considered beneficial in patients with unilateral lesions, focal seizures and ESES. ESES resolution, seizure freedom, and improvements in development and behaviour have been described after resective surgery or callosotomy, but reported cases remain scarce,[2,3] and little consideration is given to cognitive and behavioural aspects. This study provides detailed information on neuropsychological function of two patients with ESES before and after treatment.

2. Case reports

2.1. Case 1

Boy, born at 42 weeks gestational age by vaginal delivery, with meconial amniotic liquid. Apgar score: 6/7. He had neonatal convulsions, which were controlled. Neonatal MRI showed infarctions of both middle cerebral arteries (MCA) with a left predominance. At six months of age he developed a West syndrome, controlled with ACTH (Fig. 1).

He remained seizure-free until the age of three years, when asymmetric tonic startle-related seizures began, with frequent falls. Various antiepileptic drugs (AED) were tried, obtaining only partial seizure control. Serial video-EEG studies showed occasional spike-waves over left frontal and occipital regions. No independent contralateral EEG discharges were recorded. Psychopedagogic control revealed good evolution except for expressive language, which was limited to a few poorly articulated words and the use of adapted signs and pictograms. Good adaptation to the environment was reported, as well as the absence of problematic behaviours.

A control video-EEG performed when he was five years six months old showed ESES (spike frequency >85%). Six months later, after temporary electro-clinical improvement, seizures became daily and more intense. ESES was again documented and epilepsy surgery considered.

As part of the pre-surgical work-up, a 3T-MRI performed at six years seven months of age showed porencephaly in the left MCA territory, involving the thalamus, and signs of a much milder previous infarction in the right MCA territory.

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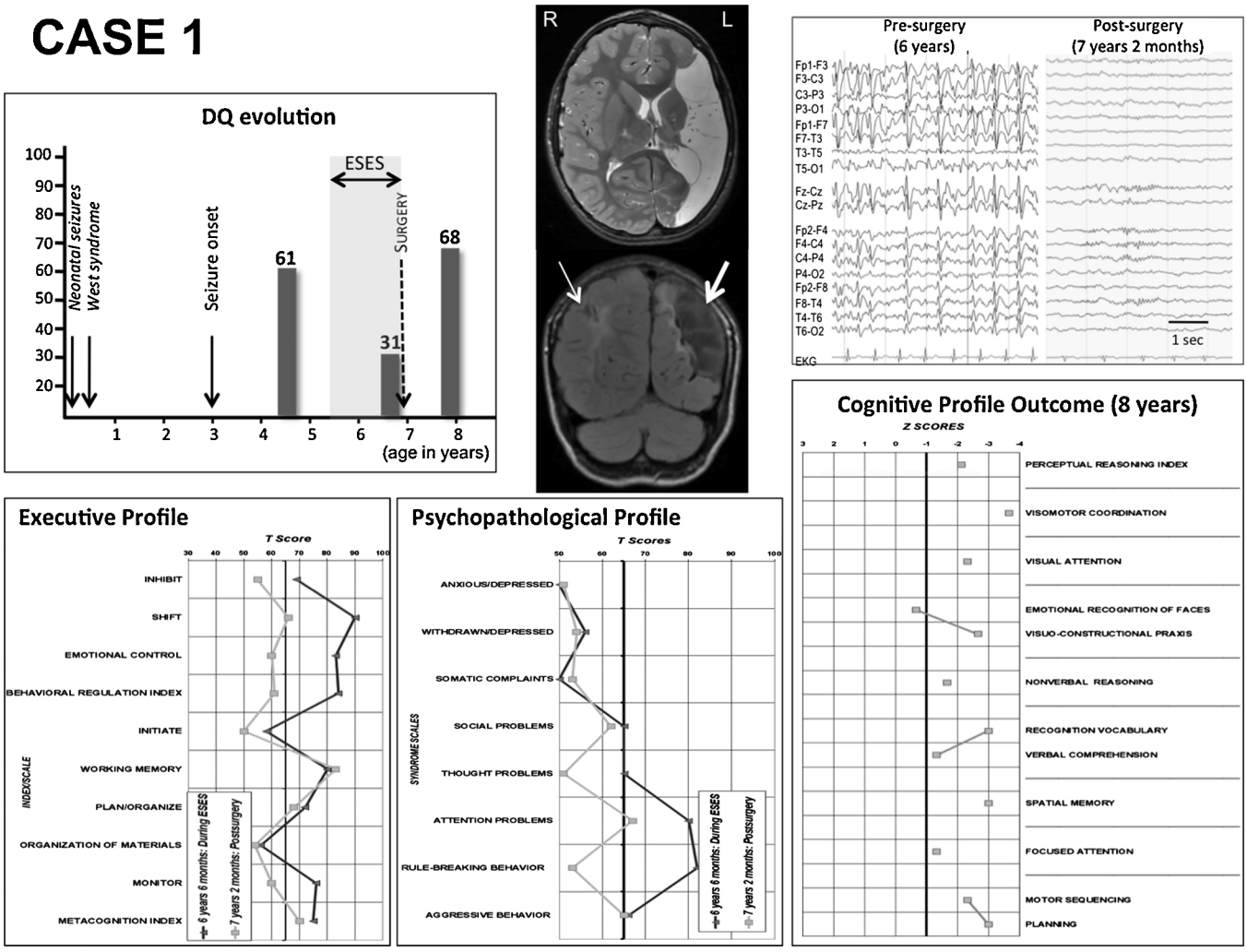

Fig. 1. Case 1 graph indicating developmental quotient (DQ) evolution over time, pre-surgical MRI scan, pre- and post-surgical sleep EEG data (gain: 20 μV/mm), pre- and post-surgical executive (Behaviour Rating Inventory of Executive Function) and psychopathological (Achenbach Child Behaviour Checklist) profile graphs and post-surgical cognitive profile graph. In the executive, psychopathological and cognitive profile graphs, the thicker vertical lines mark the limit for significant disturbances, which deviate to the right.

Neuropsychological evaluation performed one month earlier showed severe attention disturbances, restlessness and inability to persist in tasks.

Left functional hemispherectomy was performed at the age of six years eleven months.

2.1.1. Post-surgical outcome

Seizure-freedom and resolution of ESES persist at latest follow-up (three years after surgery). Sporadic left occipital spikes during sleep remained, in all postsurgical controls.

Noticeable improvements in executive functioning were documented three months after surgery, mostly in behavioural regulation aspects, emotional control and impulse inhibition (Table 1). Similarly, rule-breaking behaviour disappeared and attention problems minimized, but aggressive behaviour remained. Improvements in executive functions persist over time.

A more extensive neuropsychological evaluation was performed one year after surgery. Tests requiring expressive language were not used due to persistent absence of speech. Normalization of behavioural regulation aspects of executive functioning and cognitive improvement allowed schooling continuation at a mainstream school.

2.2. Case 2

Boy, born after 37-week gestation by emergency caesarean section due to preeclampsia. Admitted for two weeks due to hypoglycaemia and low birth weight, with no apparent complications (Fig. 2).

At the age of three months, a right hemiparesis was evident. MRI showed left hemispheric infarction. First seizure at the age of four years three months, described as eyes and mouth deviation to the right, with preserved consciousness, lasting 30 min. He had a second seizure eight months later with nausea, eyes and mouth deviation to the right, and vomiting. Similar events recurred every five or six months.

Good cognitive and satisfactory socio-emotional evolutions without significant behavioural problems were reported initially. However, progressive attention decline, impulsivity and loss of acquired academic learning were noted at later follow-up. At the age of five years eleven months, EEG showed ESES. In the following months, progressive cognitive deterioration and a sleep disorder were reported. Various AEDs and steroids were tried without improvement.

At the age of six years eight months he was referred to our hospital for surgical evaluation. Seizures remained sporadic.