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EPILEPSY SURGERY IN FOCAL CORTICAL DYSPLASIA IN CHILDREN. A REPORT OF TWO CASES AND LITERATURE REVIEW.

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ABSTRACT

Introduction: Focal cortical dysplasia is a malformation that leads to seizures which are candidates for surgical resection of the epileptogenic focus. The objective is to report two cases and review literature because there are few reports in children.

Clinical case 1: A 3-year old male with global neurodevelopmental delay and 7 seizures per day with fixed gaze, sudden and brief contractions in flexion. Right frontal partial lobectomy was performed.

Clinical case 2: A 11-year-old male with language disorder and 1 seizure per night with fixed gaze, tonic posture, and 30-second automatisms. Right frontal hemispherectomy was decided.

Conclusions: In case 1 a type IIB cortical dysplasia was identified and surgery was performed at the age of 3 years 9 months, while in case 2 cortical dysplasia type IA was observed and surgery was performed at the age of 11 years, presenting better surgical results with classification of Engel type I in the first case, unlike the second case which resulted in a classification of Engel type IV. The early age of surgery and the histological type of dysplasia with globoid cells are predisposing factors for better surgical results.

KEYWORDS : malformations of cortical development, epilepsy, hemispherectomy.

INTRODUCTION

Epilepsy is a frequent cause of attention in neurology and 15 cases per 1000 residents in Mexico are reported.[1] Within epilepsy etiology, it has been found that focal cortical dysplasia is a malformation that determines epileptogenic foci associated to epileptic seizures which are resistant to drug treatment. An estimation states that 30% of the patients with focal epilepsy are resistant to antiepileptic drugs[2] which is why surgical resection of the epileptogenic foci may be the curative treatment.

Clinical case 1

Male patient with 3 years and 9 months of age, with global neurodevelopmental delay records and epileptic seizures, with approximately 7 seizures a day since the age of 11 months.

The epileptic seizures are described as 3 seizures with fixed gaze, followed by abrupt and brief contractions with flexion of the head and four limbs over the trunk, with 2-3 bursts with falls to the floor, irritability and postictal drowsiness, with durations of 10 seconds and 4 palpebral focal clonic seizures with alterations in the alert status, with 15-second duration. It was handled with levetiracetam, vigabatrin, and topiramate, with a poor response, being drug resistant.

The epilepsy surgical protocol was followed, finding in the nuclear magnetic resonance of the skull previous to surgery a hyperintensity in T2 and FLAIR in the right middle frontal gyrus (Figure 1a); in the brain SPECT previous to surgery an hypoperfusion of the orbital gyrus of the right frontal lobe was found (Figure 1b); and in the electroencephalogram previous to surgery outbreaks of right frontal slow spike-wave of 2 Hz and of 1-2 seconds duration, in vigil and increasing during photostimulation and sleep, were documented (Figure 1c).

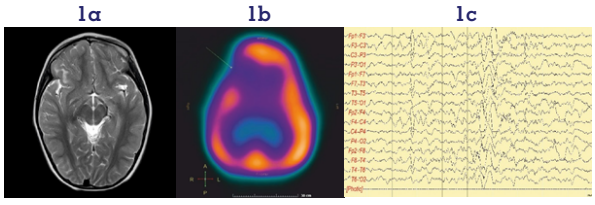

Figure 1. a. Nuclear Magnetic Resonance of Skull before surgery.
b. Brain SPECT before surgery.
c. Electroencephalogram before surgery.

During surgery, the area of dysplasia was located, and a resection of the abnormal cortical area was carried out (partial right frontal lobectomy). The result of the surgery was satisfactory, and the findings of the histopathological study were: right frontal injury F1, hard operculum, off-white, blackish spots of 2x3x3 cm not much vascularized. Delamination with gigantic, dysmorphic neurons, and globoid cells in a white substance. The neuropathological diagnosis was: Focal cortical dysplasia Taylor IIB.

In the cranial scan following surgery, post-surgical changes were found in the right frontal region. In the EEG after surgery a right hemispheric dysfunction was observed.

Post-surgical results: The patient is currently free from seizures of the Engel type I classification, with left body hemiparesis sequela with muscle strength 3/3/3, the patient can emit monosyllables and attends comprehensive rehabilitation therapy.

Clinical case 2

Male patient of 11 years of age, with language disorder records that are present since the age of 6 years, epileptic seizures during the second third of sleep with fixed gaze, tonic posture and automatisms with a duration ranging from 30 seconds to 1 minute, with a postictal period of 2 hours, with frequency of one even per night. It is being handled with polypharmacy with clonazepam, topiramate, levetiracetam, oxcarbazepine and lamotrigine.

An epilepsy surgical protocol was followed, finding in the nuclear magnetic resonance of the skull previous to surgery a right side hemiatrophy (Figure 2); in the brain SPECT previous

to surgery an increase in the cerebral perfusion in both frontal lobes was observed with a right side predominance of the superior gyrus; in the electroencephalogram previous to surgery frequent paroxysms of spikes, sharp waves and slow theta delta waves in the right frontal region with spread to the contralateral hemisphere and secondary generalization were found.

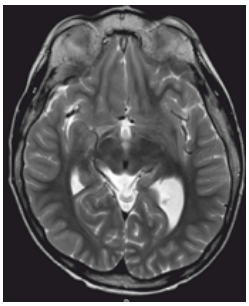

Figure 2. Nuclear Magnetic Resonance of Skull before surgery.

Due to the poor response, it is decided on a right frontal hemispherectomy with lateral resection of the frontal lobe up to F4, resection of middle frontal lobe up to the medial longitudinal fissure and the right gyrus, with resection of precentral convolution.

Histopathological study: dysplastic and swollen frontal, with a firmer consistency than usual. Delamination: the neuropathological diagnosis was: Focal cortical dysplasia Taylor IA.

In the nuclear magnetic resonance of the skull, after the surgery, post-surgical changes were found in the right frontal region. In the electroencephalogram after surgery, two-way fronto-temporal slow waves with right predominance were observed.

Post-surgical results: Post-surgery evolution has not been satisfactory due to the fact that focal crises have persisted with left eye version, switch-off episodes with tonic phase in the right hand and a tendency to generalize with 2-minutes duration, with a frequency of a crisis per day, and of Engel class IV classification. Left body hemiparesis sequela with muscle strength 5/4/4 with low intellectual disability.

DISCUSSION

In literature, the location of dysplastic injuries in focal epilepsy are predominantly extratemporal: frontal (26%), temporal (12%), multilobar/frontal including the central region (19%), posterior multilobar (without the central area) (34%), parietal (4%), and occipital (4%).[3] In the cases presented, the predominant location was of the frontal type.

According to Wyllie E. and col. (1998), after a two-year follow-up after the epilepsy surgery, 50% of the children remained in the Engel[4] I classification (free of seizures), 10% in the Engel II classification, 33% in the Engel III classification and 7% in the Engel IV classification (without improvement).[5]

In case 1, it was observed that surgery was conducted at the age of 3 years 9 months with an evolution of 1 year and 10 months, while in case 2 surgery was carried out at 11 years of age, with an evolution of 5 years, showing better surgical results with an Engel type I classification in the first case, in contrast to the second case with an Engel type IV classification. According to worldwide reports[6], carrying out the surgery at an early age favors a better post-surgical prognostic.

According to Hader WJ and col. (2004) patients with complete excision of the injury have better post-surgical results.[7,8] In case 1 the complete excision was carried out giving a positive evolution.

In the study presented by Kral and col. (2003) patients with focal cortical dysplasia and globoid cells had a better post-surgical prognosis with Engel type I/II classification in 78% of the patients, while patients with focal cortical dysplasia and hippocampal sclerosis or a low-grade glioma presented a worst prognosis with Engel type I/II classification in 50% of the patients.[9,10,11,12] In our patients, in case 1 a cortical dysplasia of type IIB (dysmorphic neurons and globoid cells) was identified with a better post-surgical prognosis with Engel type I classification, while in case 2 a cortical dysplasia of type IA (irregular lamination) was observed with a worse post-surgical prognosis with Engel type IV classification.

In scientific literature on the Focal Cortical Dysplasia of Taylor, favorable results of surgical ablation of the epileptogenic focus exist in a significant number of cases.[13] The surgical ablation of the type IIB cortical malformation reduced the epileptic seizures by 80%. In a number of cases, 63% of the patients remained free of epileptic seizures for a year after the surgery[14], which is similar to the evolution of patient 1, who currently remains free of seizures, with an neurodevelopment improvement, especially in the language area.

The hemispherectomy is one of the most used surgical procedures in cases of secondary refractory epilepsy to cases of cortical malformations.

The epilepsy surgery has as its objective to stop epileptic seizures with the purpose of improving the patient's cognitive status and avoiding deleterious effects of refractory epilepsy. The surgical result will depend on the histo-pathological level, intensity, and frequency of epileptic seizures and on the preoperative time of evolution.

In our Pediatric Neurology services two patients with different post-surgical evolution were reported, finding favorable results in the first one. While doing a worldwide literature review, it was found that surgery performed at an early age, as well as the histologic type of dysplasia with globoid cells, are predisposing factors for obtaining better surgical results.[15-18] For this reason, it is important to report these cases and to analyze the importance of carrying out a timely surgery and thus improve the post-surgical and neurodevelopment prognosis of patients with refractory epilepsy.

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