Case Report

Occipital intermittent rhythmical delta activity in a schoolgirl with microcephaly

José Guevara Camposa,*, Lucía González-Guevarab and José Guevara Gonzálezc

aDepartment of Pediatrics, Felipe Guevara Rojas Hospital, El Tigre- Anzoátegui, Venezuela
bDepartment of Epilepsy and Encephalography Unit, El Tigre, Anzoátegui, Venezuela
cDepartment of Pediatrics, Health Sciences School “Dr. Francisco Battistini Casalta”, Universidad de Oriente, Ciudad Bolívar, Bolívar, Venezuela

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Abstract. The clinical significance of occipital intermittent rhythmic delta activity (OIRDA) on the electroencephalogram (EEG) has not been fully established. Over the years, it has been suggested that the interpretation of the OIRDA electroencephalographic pattern may be related to metabolic problems, structural lesions, infections and epilepsy. Recent studies suggested that this pattern occurs almost exclusively in children and is probably of epileptic origin in most cases. It has been associated with primary generalized epilepsy syndromes, such as childhood absence. An 8-year-old schoolgirl, attending the 3rd grade, with easy distractibility, inattention, low school performance, and microcephaly is described. EEG was performed awake during hyperventilation revealed occipital intermittent rhythmic delta with a frequency of 3 Hz, bilateral and synchronous, with 2nd and 4th sec of duration. Antiepileptic therapy with valproic acid was begun. After being treated for 8 months, no OIRDA was detected on follow up EEG. The presence of OIRDA activity on EEG in a child should alert us to the presence of absence epilepsy.

Keywords: Occipital rhythmic, absences, electroencephalogram, epilepsy, delta activity

1. Introduction

Occipital intermittent rhythmic delta activity (OIRDA) was described by Cobb [1] in 1945 in patients with convulsive absence seizures. Due to the difficulty in differentiating the physiology from the pathology in the posterior waves, in 1959 Aird and Gastaut [2] examined several patients and described four groups of posterior rhythms: a) a slow alpha variant; b) slow posterior waves; c) pathological posterior rhythm, and lastly, d) slow posterior rhythm associated with petit mal. This latter rhythm, is characterized by a 3 Hz frequency, was of high amplitude, occurred symmetrically and synchronously in bursts. It was blocked by eye opening and was accentuated during hyperventilation. Currently there are three patterns of intermittent delta activity with different clinical symptoms [3]. The first is frontal intermittent rhythmic delta activity (FIRDA). This activity was associated with injuries of the central nervous system [1]. Recent studies have shown that FIRDA also occurs in metabolic processes in adults prior to cerebral ischemia [4]. The clinical correlate of FIRDA in children is unclear. The second is temporal intermittent rhythmic delta activity (TIRDA) which is associated with temporary epilepsy [1,5]. Lastly, OIRDA, which
occurs almost always in children, has been associated with epilepsy, particularly in primary generalized epilepsy syndromes. These findings were reported in a work by Gullapalli and Fountain [6]. Recent studies have shown that OIRDA is probably an epileptiform pattern [7,8]. While studying the occipital intermittent delta rhythm in pediatric patients, Rivello and Foley [7] found that the localization of the rhythm was 87% occipital in most of the cases, and 81% of the patients had epileptic activity. Convulsions were significantly more frequent in patients who had OIRDA. The frequency of the occipital rhythm was 2.89 Hz. This rhythm pattern was 21% unilateral and hyperventilation was present in more than half of the cases [8]. Moreover, it intermixed with other epileptiform discharges in 30% of the epilepsy patients, compared with 12.5% of patients without seizures [6]. However, this pattern was not considered to be pathognomonic for epilepsy because it can occasionally be seen in patients with encephalopathies [8,9]. Monotherapy with valproic acid controlled convulsions in 92.85%, and in 71.42% with simultaneous disappearance of the OIRDA rhythm [9]. This work describes the case of a schoolgirl showing OIRDA who was treated with anti-epileptic medication, resulting in the disappearance of the epileptiform pattern as well as clinical improvement.

2. Case report

An 8-year-old schoolgirl with learning difficulties, inattention, easy distractibility, hyperactivity, is described. These symptoms were interpreted as possible absence seizures. She was in the 3rd grade with poor school performance but had not repeated any school year. She was referred to a neuropediatric for neurological examination. She was the term product of a uncomplicated 3rd pregnancy. Parents were non-consanguineous and delivery was normal. Apgar scores were 7 at 1st min, 9 at 5th and 10th min. Birth weight was 2.700 kg (50th percentile) and height birth 47 cm (50th percentile). Head circumference was 35 cm (50th percentile). Her siblings were healthy. Psychomotor development was normal. She started to sit up when she was 7 months old and cruised at 13 months. Acquisition of the language was appropriate. No history of convulsive episodes or neurological disease was noted.

On physical examination, weight was 22 kg (50th percentile), height 124 cm (50th percentile) and head circumference 47.5 cm (< 10th percentile). No dysmorphic features or cutaneous blemishes were observed. There were no cranial nerve palsies. Deep tendon reflexes were normal. Respiratory, cardiovascular, and gastrointestinal systems were normal. On laboratory tests, hemoglobin was 12.7 g/dL, hematocrit 42%, platelets count 204,000/mm³, glucose 71 mg/dL, urea 22 mg/dL, and creatinine 0.8 mg/dL. Sodium was 140 mEq/L, potassium: 4.9 mEq/L, and chlorine 104 mEq/L. Thyroid stimulating hormone was 1.41 ng/mL (normal 0.3–3.8 ng/mL) and thyroxine 0.88 ng/mL (normal 0.7–1.3 ng/mL). Cystine-homocystine test was negative. Urinary organic acids were normal. Lactate and pyruvate levels were normal. Serologic examination for toxoplasmosis, rubella, cyto-
megalovirus, herpes simplex, and human immunodeficiency virus were negative. Cranial magnetic resonance imaging (MRI) was normal. On electroencephalography (EEG) during awake and natural sleep, intermittent, symmetrical and synchronous occipital delta wave activity with a frequency of 3 Hz was observed in both hemispheres lasting 2–4 seconds. This was present during hyperventilation and visual stimuli and was blocked by eye opening (Figs 1–3). Valproic acid at 15 mg/kg/day was prescribed. Eight months after treatment was started, EEG showed disappearance of OIRDA. The patient had been promoted to the next higher grade in school and improved clinically (Fig. 4).

3. Discussion

The meaning of OIRDA on EEG has intrigued neurologists, epileptologists and electroencephalographers for decades. Over the years, the range of clinical interpretation of this EEG pattern by researchers varied because it is unspecific, and had been associated with metabolic, infectious and epileptic etiologies. Furthermore, there are two more patterns of intermittent delta activity in addition to OIRDA with important clinical significance namely, frontal intermittent rhythmic delta activity, and temporal intermittent rhythmic delta activity [8].

OIRDA occurs exclusively in children. This activity has been associated with epilepsy, particularly in generalized epileptic syndromes such as in children with absence episodes [9]. OIRDA cases have also been described in juvenile Huntington’s disease, salmonellosis of the central nervous system, and in subacute sclerosing panencephalitis [10,11]. Gullapalli and Fountain [6] found that the most frequent convulsive episodes in patients with OIRDA are generalized tonic-clonic and absence episodes. Thirty percent of epileptic discharges occur in epileptic patients as compared to 12.5% in patients without convulsive episodes [6,12]. Occipital intermittent delta activity in most patients occurs during wakefulness and hyperventilation [8], as in our case. The presence of lack of attention and hyperactivity suggested the possibility of attention deficit hyperactivity disorder (ADHD); which was discarded after the Diagnostic and Statistical Manual of Mental Disorders (4th ed) criteria was not met. However, OIRDA has not been described as being associated with ADHD [13].

Forty-five percent of the most frequent electroencephalographic alterations in ADHD occur in the temporal region, followed by 45% in the frontal region, with no alterations in the occipital region such as those described in our case [13], although there is certainly no electrical pattern for children with ADHD, as the diagnosis is eminently clinical. Posterior fossa tumors, particularly in the 4th ventricle, may show
abnormalities in the EEG, such as arrhythmic posterior slow waves [14]; however, this was ruled out by the normal results of the MRI of the brain. The lack of head trauma history in the preceding weeks ruled out the possibility that the OIRDA was brought on by trauma [15]. In most cases, neuroimaging does not show structural pathologies associated with OIRDA [8].

Due to the characteristics of OIRDA during wakeful EEG, during hyperventilation and visual stimuli-, given the clinical symptoms, of inattention and distractedness, we suspected the possibility of absence epilepsy. Consequently, we decided to treat with valproic acid at a dose of 15 mg/kg every 12 h, with good clinical and encephalographic results.
OIRDA is considered as a sign of good prognosis in absence epilepsy. In 76.92% of cases, valproic acid controls the seizures and normalizes the EEG [9].

References