Case Report

Occipital intermittent rhythmic delta activity (OIRDA) in pediatric focal epilepsies: A case series

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Abstract

In this case series, we have identified an atypical pattern of OIRDA (Occipital intermittent rhythmic delta activity) on the electroencephalograms (EEGs) of three pediatric patients with self-limited focal epilepsies, including Childhood Epilepsy with Centrottemporal Spikes (CECTS), and Panayiotopoulos syndrome (PS). Previously, OIRDA was described as a symmetric sinusoidal occipital-maximal activity, often associated with childhood idiopathic generalized epilepsies, although it was also reported among other physiologic or pathological entities including focal epilepsy. We have observed in our case series that OIRDA, without prominent field effect, is lateralized or maximal on the hemispheric side ipsilateral to the more defining epileptiform discharges in these focal epilepsies. They also exhibit a notched morphology due to the intermixed sharp wave activities, although the sharp waves are not occurring repetitively. This report provides additional evidence that OIRDA can be associated with a spectrum of idiopathic focal epilepsies and may suggest a cortical origin of OIRDA in these patients as opposed to a hypothesized subcortical generator in patients with primary generalized absence epilepsy, even though further investigation is warranted for either hypothesis.

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Introduction

Occipital intermittent rhythmic delta activity (OIRDA) is an interictal electroencephalographic (EEG) finding, described as a sinusoidal, 3 Hz, high-amplitude, symmetrical, occipital-maximum activity which may have a spatial field to the posterior temporal or parietal channels, most commonly associated in children with idiopathic generalized epilepsies [1], such as Childhood Absence Epilepsy (CAE) [2,3]. In some instances, OIRDA is associated with better prognosis among patients with absence seizures [4]. This finding was initially thought to be associated with posterior fossa structural lesions or midline structural lesions [5], but has since been found in cases of juvenile Huntington’s disease [6] and encephalopathy, including one case of CNS salmonellosis [7] and two children with anti-NMDA receptor encephalitis [8]. OIRDA-like rhythms may even be a physiologic subcortical process associated with eye closure, i.e. the “phi rhythm” [9,10].

Nevertheless, the electrographic features of OIRDA associated with focal epilepsies have not yet been well-characterized. We identified OIRDA in three pediatric patients with focal epilepsies, including Self-limited Childhood Epilepsy with Centrottemporal Spikes (CECTS) [11,12], and Benign Childhood Epilepsy with Occipital Spikes, formerly known as Panayiotopoulos syndrome (PS). We have identified some ‘atypical’ or focal features of OIRDA in patients with focal epilepsies, including lateralized OIRDA, a notched morphology or embedded sharp wave forms ipsilateral to focal centrottemporal or occipital spikes, and lack of prominent field of distribution, implicating a focal origin, and perhaps a link between this ‘atypical’ OIRDA and a spectrum of idiopathic focal epilepsy syndromes [13,14].

Case presentations

Case 1. A 5-year-old boy with normal developmental history presented for evaluation of sporadic seizures, which first occurred at age 4 years and 7 months. Semiologically, he would deviate his head and eyes to the right, associated with clenching of the jaws, staring, and unresponsiveness, lasting 10–15 minutes. On one occasion, he had non-bloody, non-bilious emesis. No clonic convulsions were observed. He had two EEG studies performed. The first,
a routine inpatient study which captured only wakefulness, had independent left central (C3 maximum) spike and slow wave discharges seen more prevalently than right central discharges (C4). The second EEG, a six-hour, prolonged outpatient study which captured sleep, was performed five months after the first, and now revealed left centroparietal (C3 maximum), left occipital (O1), and right centroparietal (C4 maximum) spike and slow wave discharges, with sleep potentiation, and left-sided only OIRDA (Fig. 1-A-B, Table 1). The OIRDA was observed during the awake state, separate from hyperventilation; it was 3 Hz in frequency, primarily over O1 without a prominent field of distribution, and of medium (120–190 uV) amplitude, and with a notched morphology (Fig. 1A). Brain MRI was normal, and the child was presumptively diagnosed with CECTS following the first EEG, despite a semiology more suggestive of PS. He has been treated with oxcarbazepine with good effect on seizure control.

Case 2. A 7-year-old girl with normal developmental history was evaluated for staring spells following a generalized convulsive seizure. The child’s first seizure was at age 2 years and 11 months, described as left arm twitching followed by 10–15 min of full body twitching, which occurred out of sleep, with subsequent post-ictal sleepiness. The child had a routine outpatient EEG, which captured sleep, with frequent bicantral (C3, C4) sharp or spike wave discharges with sleep potentiation, of shifting hemispheric predominance. The child was lost to follow up, for nearly 5 years, during which time she developed frequent spells wherein the child would “stare and smile, as if looking through out,” without automatism or behavioral interruption, of an unclear etiology, as well as nodding and throat clearing tics, not yet meeting criteria for Tourette’s syndrome. Repeat routine outpatient EEG, which captured sleep, revealed independent bilateral centroparietal (C3/T3, C4/T4) spike or sharp wave discharges with presence of tangential dipole, as well as left more prevalent than right occipital (O1 > O2) epileptiform discharges (Fig. 1C-D, Table 1). In addition, bilateral (O1, O2), 3 Hz, medium to high (150–250 uV) amplitude OIRDA without a field was present (Fig. 1C). The OIRDA was seen during and outside of hyperventilation, at times showing a notched morphology due to admixed occipital sharp waves. Brain MRI subsequent to this EEG study revealed a 2 mm right frontal gray matter heterotopia, thought not to be correlative with the EEG focus. Although some clinical features and the EEG findings suggest CECTS, this child has never been on anti-seizure medicines, due to lack of electrographic correlation with the staring spells, only one lifetime convulsive seizure. A decision was made with the parents to observe the spells with plan for a future evaluation in the epilepsy monitoring unit (EMU).

Case 3. A 3-year 6-month-old boy with normal developmental history was evaluated in the neurology clinic after an unprovoked seizure, which occurred 2 months prior. It was described as vomiting at night, loss of awareness, associated with eyelid fluttering and clonic activity of the right hand. The episode lasted approximately 15 minutes with postictal confusion. Routine outpatient EEG, which captured sleep, revealed O1 and O2 spike and slow wave complexes, which were greatly potentiated during drowsiness and sleep. A maximum amplitude was over O1 with a field involving P3 and P4. In addition, there was 2.5 Hz, medium amplitude (120–150 uV), also left more prominent than right, OIRDA during wakefulness, often not associated with hyperventilation (Fig. 1E-F, Table 1). At times, there is a notched morphology within the OIRDA due to admixed occipital sharp waves. Brain MRI was normal. The child was diagnosed with PS and started on oxcarbazepine. He never developed recurrent seizures. Oxcarbazepine was weaned due to significant weight gain and eventually converted to topiramate.

Discussion/Conclusions

In accordance with the American Clinical Neurophysiology Society’s standardized critical care EEG terminology (2021 version), Rhythmic Delta Activity (RDA) is defined as a waveform with relatively uniform morphology and duration, without an interval between consecutive waveforms, and a frequency of 0.5 to ≤4.0 Hz [15]. Although OIRDA has been documented previously in cases of focal epilepsies [14] in addition to CAE, repeat studies have not corroborated this as a statistically significant correlation [13]. Our case series, albeit limited in number, illustrates the presence of OIRDA in children with focal epilepsies and some atypical features as compared to that published in cases of CAE. Therefore, this series highlights a less-well recognized association between OIRDA and idiopathic childhood focal epilepsy syndromes, including CECTS and occipital epilepsies.

All three cases of OIRDA exhibit occipital spikes or sharp waves, in addition to a CECTS-like spike-wave discharges in the first two cases, suggesting a link between occipital epileptiform discharges and OIRDA. Furthermore, case 1 presents with unilateral OIRDA in the left side with concurrent O1 spike–wave discharges. The other two cases show occipital spikes or sharp waves of a left-sided predominance as well as bilateral OIRDA; yet, asynchrony (case 2) or amplitude asymmetry (case 3) are appreciable between the left and right-sided OIRDA. Watemberg et al also reported different electrographic features of OIRDA in patients with focal epilepsies when comparing to those with primary generalized epilepsies [14]. We therefore speculate that OIRDA in focal epilepsies represents a focal cortical irritability, as opposed to the previous hypothesis of a subcortical or thalamic origin of OIRDA in patients with CAE. However, it remains to be investigated in a large cohort whether there is a positive correlation between the laterality of occipital discharges and OIRDA distribution when both present.

It has been documented that, in serial interictal EEGs of patients with CECTS, generalized discharges or focal discharges in the midline, parietal, frontal and occipital regions can be observed, where occipital discharges are typically first to present [16]. Furthermore, several reports in the literature [11,12,17] have described a link between occipital and centroparietal spikes. Case 1 highlights the observation of co-existent CECTS-like spikes, O1 spike-wave discharges, and unilateral OIRDA (O1). Interestingly, the previous EEG obtained 5 months prior showed only bilateral central spikes; no OIRDA or occipital spikes were detected despite a semiology more suggestive of PS. This observation led us to question whether this represents simply an overlap between CECTS and PS, or a temporal evolution of centroparietal spikes to subsequently include occipital discharges and OIRDA. The caveat to this interpretation is that the second EEG performed was running longer than the first (6 hours, as compared to 40 minutes), which might have increased the detection sensitivity.

Interestingly, OIRDA was also seen in children with independent frontal or temporal epileptiform discharges [14], and reported in two children presenting with focal epilepsies associated with NMDA receptor encephalitis [8], suggesting a wider spectrum of this link between OIRDA and focal epilepsies. Similarly, TIRDA (temporal intermittent rhythmic delta activity) is considered as a potential EEG marker of focal epileptogenesis in temporal lobe epilepsy and likely has a neocortical origin based on the concurrent scalp EEG and stereoecephalography recordings [18,19]. This neocortical origin of TIRDA was further corroborated by the observation that de novo TIRDA in patients who underwent laser interstitial thermal therapy (LiTT) for mesial temporal sclerosis was associated with a poor post-LiTT seizure outcome [20]. The clinical relevance remains uncertain based on this observational...
series; however, lateralized OIRDA, with admixed epileptiform discharges and without a prominent field of distribution may be demonstrative of focal cortical abnormality which predisposes to epileptogenic discharges, especially when concurrent with other defining focal discharges, as opposed to the previous hypothesis of a nonspecific abnormality of subcortical or thalamic origin.

Fig. 1. Electroencephalographic (EEG) samples of OIRDA and associated epileptiform discharges (in dashed circles and square). (A) A 5-year old boy (case #1) with unilateral (O1) 3 Hz OIRDA concurrent with ipsilateral C3 spikes during wakefulness. At times, a notched morphology is appreciated within the OIRDA (arrow). There are also sleep potentiated O1 and C4 spikes in this patient (B). (C) A 7-year old girl (case #2) demonstrates bilateral 3 Hz OIRDA with concurrent bi-hemispheric centro-temporal spikes/sharps during wakefulness. At times, the OIRDA displays a notched morphology (arrow). This patient has also bilateral occipital yet O1 predominant spike-slow wave discharges (shown in dashed square in D). (E) A 4.5-year old boy (case #3) with a diagnosis of Panayiotopoulos syndrome has bilateral occipital yet O1 predominant spike-slow wave complex, as well as 2.5 Hz OIRDA more prominent in O1 than in O2 (F), which also exhibit a notched morphology at times (arrow). All EEG tracings are displayed in AP bipolar montages, except D in average referential montage. Setting: LFF 1 Hz; HFF 70 Hz; Notch 60 Hz; sensitivity 10 uV/mm in A-B, 15 uV/mm in C and F, 20 uV/mm in D and E; time base 30 mm/sec.

Table 1
Characteristics of the EEG findings in three patients with OIRDA and focal epileptiform discharges.

| Case # | Age at onset | Gender | PDR (Hz) | OIRDA description | Associated EDs | ASMs |
|--------|--------------|--------|----------|-------------------|----------------|------|
| #1     | 4 y 7 mo     | M      | 8        | O1, 3 Hz, 120–190 uV, 3–10 seconds in duration, during wakefulness, outside HV, at times with a notched morphology | Independent frequent C3, C4, and O1 spike-slow wave; sleep potentiation of O1 and C4 spikes | OXC |
| #2     | 2 y 11 mo    | F      | 8        | O1 and O2, 3 Hz, 200 uV, during wakefulness, during and outside HV, at times with a notched morphology | Independent C3/T3 (with sleep potentiation) and C4/T4 spikes; O1 and O2 sharp waves (O1 predominant) | None |
| #3     | 3 y 4 mo     | M      | 8        | O1 and O2 (O1 predominant), 2.5 Hz, 120–150 uV, during wakefulness, during and outside HV, at times with a notched morphology | O1 and O2 (O1 predominant) spike-slow wave complex with sleep potentiation | OXC, then TPM |

PDR: posterior dominant rhythm; OIRDA: occipital intermittent rhythmic delta activity; yo: years old; mo: months old; uV: microvolts; HV: hyperventilation; EDs: epileptiform discharges; ASMs: anti-seizure medications; OXC: oxcarbazepine; TPM: topiramate.
OIRDA appears to be a relatively rare finding, with rates being reported between 0.75% [13] to 3.4% [14] of studies showing this finding. Given the rare relative rarity of OIRDA, and potential pathophysiologic implication, further studies are necessary to determine if a correlation exists between OIRDA spatial distribution and localization, epileptiform features, cortical versus subcortical origin, or if this proposed link between CECTS/PS, and OIRDA represents a temporal progression or resides on a spectrum of focal and generalized abnormalities when OIRDA is seen in these patients.

Disclosure

The authors don’t have any disclosures.

Study funding

We didn’t receive any funding.

Ethical statement

We have consulted Lifespan IRB (Institutional Review Boards) and were told ‘an anecdotal report on a series of patients seen in one’s own practice and a comparison of these patients to existing reports in the literature is not research and would not require IRB approval’.

Verbal consent on publishing anonymized data was obtained from parent or legal guardian of each patient.

Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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