Ictal nystagmus: acquired nystagmus of infrequent cause in pediatrics

Nistagmo ictal: causa infrecuente de nistagmo adquirido en pediatría

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Abstract

Ictal nystagmus is a rare phenomenon secondary to an epileptic seizure, usually in the temporo-occipital region. For its characterization, we must rely on clinical history, examination, and ideally observation of the episodes. Objective: To describe a case of this unusual entity and highlight the characteristics that should increase diagnostic suspicion in order to avoid treatment delay. Clinical Case: An 8-year-old schoolboy, with no relevant history, consulted due to 5-6 episodes a day in the last year of conjugate horizontal eye movements with rapid jerks and associated slight miosis, lasting 5-10 seconds, with doubtful disconnection from the environment or consciousness impairment in some of the episodes, with no other accompanying signs or symptoms. Neurological examination between episodes was normal. He was evaluated by ophthalmology and otolaryngology, which ruled out pathology in these areas. Video-electroencephalogram showed electro-clinical correlations, with epileptiform activity in the left temporal and occipital region, which subsequently generalized during episodes. Brain MRI showed no pathological findings. After initiation of carbamazepine treatment, the patient had a good evolution, without recurrence of the episodes at 2 years of follow-up. Conclusions: When faced with a case of acquired nystagmus, epileptic etiology should be included in the differential diagnosis, especially if the frequency of episodes is high, of short duration, and associated with consciousness impairment. The diagnosis is based on a video-electroencephalogram with electro-clinical correlations and a good response to treatment with antiepileptic drugs is expected.

Keywords: Epilepsy; Nystagmus; Electroencephalogram; Ictal Nystagmus
Introduction

Nystagmus is an involuntary, rhythmic, and repetitive eye movement that can be congenital or acquired in origin. Acquired nystagmus can be a manifestation of stimulation of the vestibular or ocular system, involvement of the cerebellum, subcortical structures, or the cerebral cortex. The most frequent causes are ocular and inner ear diseases, head trauma, cerebrovascular accident, multiple sclerosis, anoxia, drug or toxic administration, brain tumors, or intracranial infection but it can also be secondary to epileptic activity (epileptic or ictal nystagmus)1,2.

It is important to describe the characteristics of the nystagmus (pendular or jerking, direction, oscillation in the plane, amplitude, and frequency) for which the clinical history and physical examination of the patient are essential3, and video recording of some of the episodes by the family can be very useful. It also helps to determine whether it is related to gaze fixation, changes in position, or head movements, as well as to exclude other causes of abnormal movements such as opsoconus, ocular flutter, or ocular bobbing, and thus try to elucidate between central (lesions of the brainstem, cerebellum, subcortical structures, or cerebral cortex) or peripheral (ocular, extraocular musculature, or vestibular diseases) causes of nystagmus4.

Epileptic or ictal nystagmus was first described by Féré in 18904 and, since then, less than 100 cases have been described in the literature with ages ranging from 10 days to 75 years, with a more frequent presentation in childhood5,6. It is most often accompanied by other signs such as oculocephalic deviation, visual hallucinations, amaurosis, vertigo, blinking, focal seizures, autonomic changes, or even altered level of consciousness but, exceptionally, it can appear in isolation4, which makes diagnosis even more difficult. Given the scarce evidence on the subject, the description of new cases of ictal nystagmus could contribute to its early identification.

We present a clinical case of ictal nystagmus in a pediatric patient to recall the existence of this rare entity as a presenting sign of an epileptic seizure since it is not included in most of the algorithms for the management of nystagmus in the pediatric age and it is a pathology little known by the general pediatrician. It is important to highlight the characteristics that should increase its diagnostic suspicion in order not to delay the initiation of treatment.

Clinical Case

An 8-year-old boy, with no personal or family history of interest, attended the Pediatric Emergency Department due to a 1-year history of abnormal eye movements. The family reported that the child presented 5-6 episodes per day, lasting 5 to 10 seconds, consisting of horizontal conjugated eye movements with rapid jerks and associated slight miosis (Video 1), without associated head or limb movements. They were not related to gaze fixation, position or head movements. The patient could not self-induce the episodes but could identify their onset which was of abrupt onset and end, some of them seemed to be disconnection from the surroundings or altered states of consciousness and did not refer to postictal period or headache. They always occurred during wakefulness and never during sleep. The parents had observed a higher frequency of episodes on days of greater stress and physical fatigue, without being able to identify other triggers and there were no associated ataxia, tinnitus, autonomic symptoms or hearing loss. They denied a history of head trauma and the intake of medications or other toxic substances. The symptoms had not changed or progressed from the onset to the day of consultation and the neurological examination between episodes was normal.

The ophthalmologic evaluation ruled out visual pathology and the otolaryngologic evaluation ruled out pathology in that area and described an episode of high-intensity and low-amplitude spring nystagmus. He was hospitalized for further study, performing general laboratory tests (including thyroid hormones, liver function, autoimmune profile, and vitamin D), which showed no pathological findings. Video recording of the episodes by the family was helpful for diagnostic guidance and a video-EEG showed electro-clinical correlates of the episodes. The video-EEG was performed in wakefulness, recording two separate events lasting about 8-10 seconds during a total recording of 25 minutes. The background tracking was normal for his age and, coinciding with the two clinical episodes of nystagmus, epileptiform activity was recorded in the EEG in the left temporal and occipital region, which subsequently generalized (Figure 1). After demonstrating focal onset epileptic activity in relation to the patient’s

Video 1. Video of one episode. We can see a jerk nystagmus with quick phase to the left followed by slow phase, not crossing middle line. Note: to scan the video codes you must focus the picture with your smartphone camera and open the link. Sometimes you should install the correct mobile application.
CLINICAL CASE

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Figure 1. The ictal EEG registry shows epileptic activity that begins in the left temporo-occipital region (arrows) with sharp waves which subsequently generalized.

symptoms, treatment was started with carbamazepine orally at ascending doses.

A brain MRI with contrast was performed, showing no evidence of structural alterations or abnormalities of cortical development, and the final diagnosis was ictal nystagmus. He was discharged home with carbamazepine at 13mg/kg/day dose which was later increased to 17mg/kg/day. In the outpatient follow-up in Neuropediatrics 2 years after starting treatment, he showed good adherence and absence of side effects to treatment, with no new episodes.

Discussion

Early etiological orientation of patients with nystagmus is important, as it may be a manifestation of a serious underlying pathology. Its etiology can be very varied and diagnosis is usually complicated. The clinical history and video recording of the episodes were fundamental in our case to guide the diagnosis of ictal etiology. The main clinical feature of our patient was nystagmus, in addition to the suspicion of disconnection from the surroundings or altered states of consciousness referred by the family during some of the episodes, which was difficult to verify due to the short duration of these and having ruled out visual pathology.

Coinciding with what has been described in the literature, ictal nystagmus episodes are very frequent with short duration (less than one minute). In some cases, the triggers of the episodes that have been described are stress, as in our patient, and light stimulation. In most cases, the nystagmus is binocular, horizontal, and conjugated, and, very characteristically, its direction (fast phase) is usually contralateral to the epileptogenic zone.

Several mechanisms involved in the generation of ictal nystagmus have been described depending on the region of the cerebral cortex in which the epileptic discharges are initiated. If they originate in the saccadic regions (frontal and temporo-occipital cortex), they cause saccadic eye movements with a fast phase directed to the side opposite to the epileptogenic zone, followed by a slow phase in which the eyes are directed to the midline, but without crossing it. In contrast, if they originate in the cortical regions of eye tracking or optokinetic reflex (temporo-parieto-occipital region), they result in slow eye movements ipsilateral to the epileptogenic zone, followed by a fast phase in which the eye crosses the midline. In our case, we suspect that the origin would be in the saccadic regions of the temporo-occipital cortex. However, the direction of the nystagmus in the child was ipsilateral to the area of electrical activity, which has been described in some studies since there may be electrical activity in...
other areas simultaneously or propagation from one area to another\textsuperscript{a}, so that the nystagmus will not always have a localizing value.

The onset of the discharges in our patient was focal at the temporo-occipital level with subsequent generalization. In most of the cases described, the epileptogenic focus is in the occipital lobe, although more than one lobe is frequently involved such as temporo-occipital, fronto-temporal, parieto-occipital, or temporo-parieto-occipital, and in some cases, generalized epileptic activity is observed\textsuperscript{12,13,15} and because it is one of the drugs approved as the first choice in monotherapy for focal epilepsy in this age group; treatment with phenytoin, lamotrigine\textsuperscript{a}, and topiramate\textsuperscript{a} has also been described.

Conclusions

The ictal etiology of nystagmus should be considered in the differential diagnosis of a patient with acquired nystagmus, especially if the frequency of episodes is high, the duration is short, or they are associated with altered states of consciousness. The diagnosis is based on a video-EEG with electro-clinical correlate and a good response to treatment with antiepileptic drugs is expected.

Ethical Responsibilities

Human Beings and animals protection: Disclosure the authors state that the procedures were followed according to the Declaration of Helsinki and the World Medical Association regarding human experimentation developed for the medical community.

Data confidentiality: The authors state that they have followed the protocols of their Center and Local regulations on the publication of patient data.

Rights to privacy and informed consent: The authors have obtained the informed consent of the parents (tutors) of the patients and/or subjects referred to in the article. This document is in the possession of the corresponding author.

Conflicts of Interest

Authors declare no conflict of interest regarding the present study.

Financial Disclosure

Authors state that no economic support has been associated with the present study.

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