Fixation-off sensitivity in a girl with symptomatic occipital epilepsy admitted in non-convulsive status epilepticus

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Abstract

Fixation-off sensitivity occurs mainly in children with idiopathic occipital epilepsies, with only few cases reported in symptomatic epilepsies. We describe an 8-year-old girl with a history of symptomatic epilepsy admitted in non-convulsive status epilepticus. The video-EEG revealed the presence of high-amplitude occipital paroxysmal activity, which occurred continuously as long as the eyes were closed and with other conditions of fixation-off, and were inhibited when eyes were opened. Initially this phenomenon was unrecognized and misdiagnosed as simple partial status epilepticus, being treated energetically, and achieving control of discharges. We discussed the unique phenomenon of fixation-off sensitivity, which can electrically mimic an epileptic status.

KEY WORDS: fixation-off sensitivity, epileptic syndromes, eyes closed sensitivity, occipital epilepsy.

Introduction

The term *Fixation-Off Sensitivity* (FOS) was coined by Panayiotopoulos to denote the forms of epilepsy or electroencephalographic (EEG) abnormalities, or both, which are elicited by the elimination of central vision and fixation (e.g. closed eyes, complete darkness, Ganzfeld stimulation) (1-6).

The FOS EEG pattern consists of repetitive high-amplitude spikes/spike-wave/sharp and slow wave complexes either generalized or localized in the occipital regions (unilateral or bilateral), that consistently occur within 1-3 seconds from eye closure, persist throughout the eye-closed state and disappear immediately with eye opening (1-4, 6).

FOS occurs mainly in children who have idiopathic focal epilepsies with occipital paroxysms, such as Gastaut syndrome and, less frequently, in Panayiotopoulos syndrome. It may also be observed in cryptogenic focal and generalized epilepsies; there are even reports of non-epileptic children and adults. Patients with symptomatic occipital epilepsy and FOS have only rarely been reported (1-6).

We describe a girl with symptomatic occipital epilepsy, who was admitted with the diagnosis of status epilepticus and was subsequently found to have FOS during EEG monitoring.

Case report

A 8-year-old girl presented with a history of perinatal hypoxia associated with bilateral schizencephaly; as sequelae she had a cerebral palsy with spastic tetraparesis, moderate-severe mental retardation and symptomatic occipital epilepsy.

She started having seizures in the first year of life, apparently well-controlled, increasing in frequency to twice a week from the age of 6. At the age of 7 she was admitted for brief, but multiple daily episodes of eyes deviations and loss of consciousness. Pharmacologic treatment included carbamazepine (CBZ), oxcarbazepine (OXC), valproic acid (VPA) and topiramate (TPM) as serial monotherapies.

At the age of 8, she was brought to the emergency room because of recurrence of these previously described episodes lasting several hours, followed sometimes by a tonic posturing involving all four limbs; she was admitted in the Pediatric Intensive Care Unit with the diagnosis of non-convulsive status epilepticus (NCSE).

The first video-EEG was performed 24 hours after admission, being the child in her baseline condition (Fig. 1). The EEG showed, with the patient staring vacantly, rhythmic, continuous 2- to 3-Hz high-voltage sharp and slow wave discharges (SSWDs), maximally in the left témporo-occipital region. The SSWDs increased 1 to 2 seconds upon eye-closure, and persisted, “waxing and waning”, throughout the record.

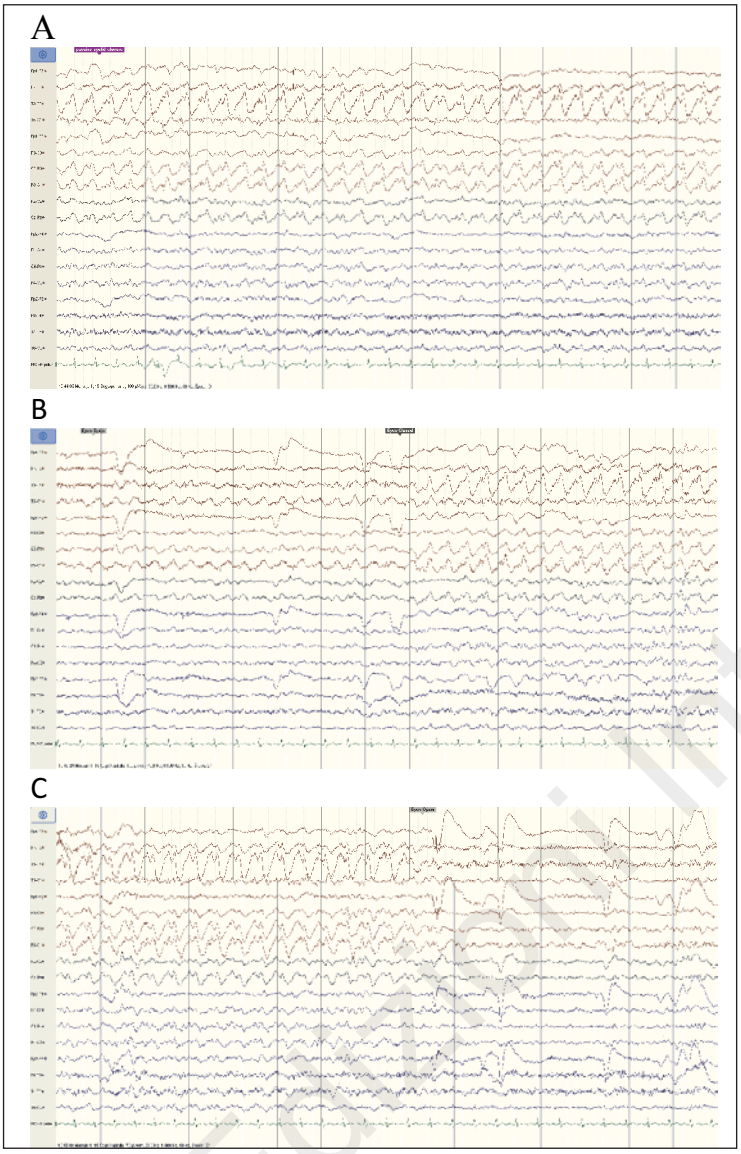

Figure 1 - A. Continuous high-amplitude left posterior sharp and slow waves discharges during the eyes-closed state in bipolar montage. B. There was an attenuation of the discharges amplitude with patient´ eyes opening without focusing on a spot, but recurred immediately on eye closure, and finally disappearing with eyes opening and fixation C.. Sensibility: 100 µV/cm, HFF: 70 Hz, LFF: 0.3, 15 seconds/epoch.

The patient reported no symptoms during occipital paroxysm, and she was able to talk. The situation was interpreted as a simple partial status and, levetiracetam (LEV) was added to the treatment.

Due to the persistence of clinical seizures, midazolam (MDZ) infusion was begun, and in the follow-up video recording in the next day, (Fig. 2 A) the discharges had completely disappeared. Sequential video-EEGs after MDZ discontinuation, showed the SSWDs reappearance with similar features, except a delay of 5-8 seconds between eye closure and the onset of paroxysmal activity. Passive eyes closure was performed, demonstrating that the discharges were inhibited on eye opening.

The electroencephalographic findings were controlled after a combination therapy of Valproate (VPA), phenytoin and clobazam (CLB). In two subsequent control registers a diffuse slow activity was evident, without SSWDs. The patient was transferred to ward and was discharged the next day.

Retrospectively, all the interictal EEGs recorded in this patient in our department were re-evaluated; there were a total of 6 records done from 2009 to 2013. Interictally there was a left slowing of background activity and, two independent left foci: one over the centro-temporal area, and another in the temporo-occipital region with major activation during sleep. A record of February 2011, at the age of five, revealed for the first time left temporo-occipital runs of irregular spike-wake, when her eyes were closed, attenuating on eye opening, although at that time nobody in our laboratory was aware of this finding. Photosensitivity was not present.

Five days after discharge a follow-up video-EEG (Fig. 2 B, C), was obtained during wakefulness; she was seizure-free since the discharge, on treatment with VPA, LEV, and CLB. FOS was evaluated according to Panayiotopoulos´s suggestive technique (2): full darkness was achieved with dark goggles and elimination

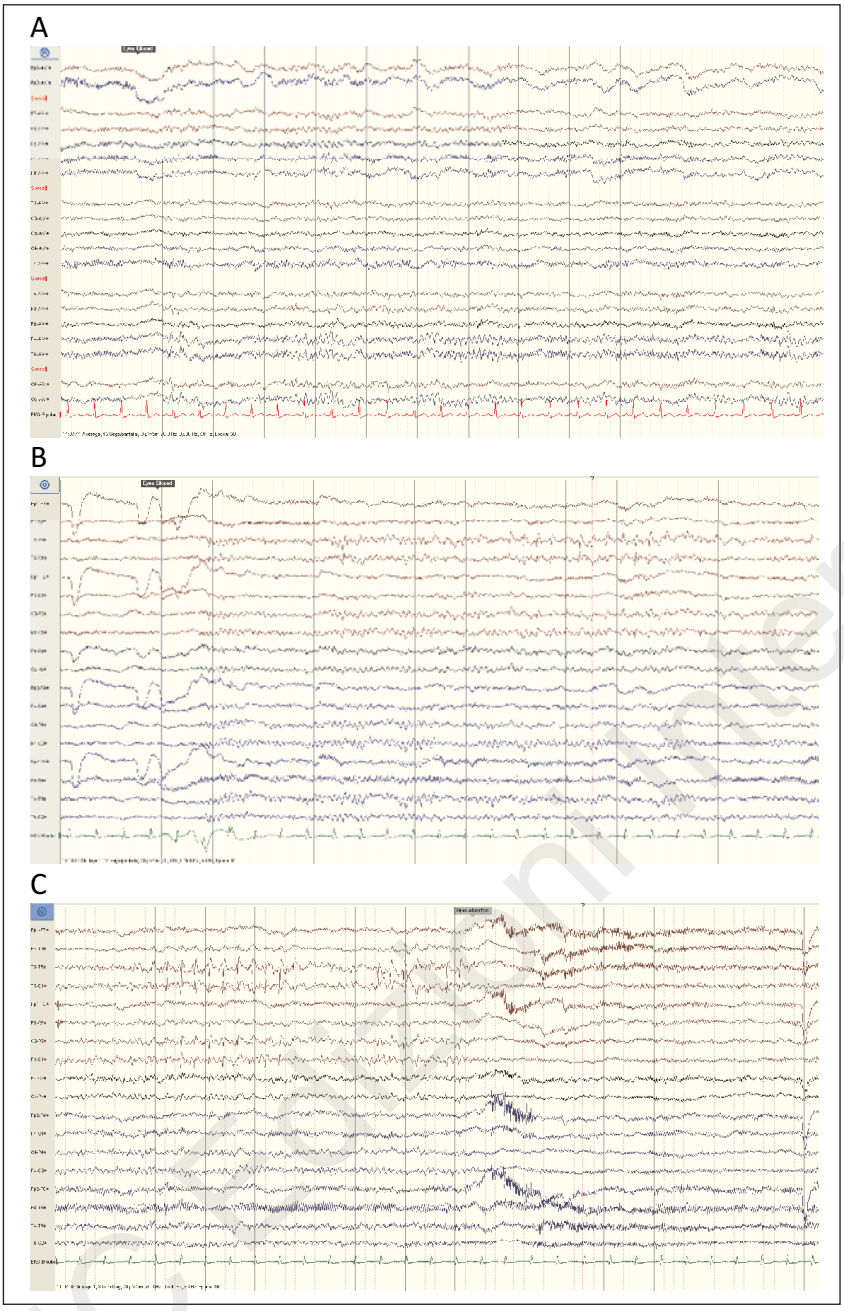

Figure 2 - A. Second day, with midazolam infusion there was a complete supression of the epileptiform discharges. B, C. Five days after the child was discharged. The interictal awake record shows sort runs of subcontinuous, repetitive spike-and waves brought on, with a delay of some seconds by eye closure and with the elimination of the central fixation with Ganzfeld stimulation, demonstrating the phenomenon of fixation-off sensitivity. Sensibility: 70 µV/cm, HFF: 70 Hz, LFF: 0.5, 15 seconds/epoc.

of central vision in an illuminated room was tested by the modified Ganzfeld stimulation (placing a sheet of white paper 20 cm in front of the patient, without visual clues). However we had some technical difficulties because she was not able to cooperate due to her learning difficulties.

Registration began with eyes opened showing only sporadic left temporo-occipital spike-waves. After a delay of 5-15 seconds, both active and passive eyelid closure induced short runs of repetitive spike-and-waves over the same region. When she opened her eyes, there was complete suppression of the discharges, but when her visual fixation was impeded by modified Ganzfeld stimulation, the activity did not respond to eyes opening. In total darkness opening and closing the eyes did not bring on any changes.

Discussion

FOS is an intriguing phenomenon, probably underdiagnosed (with an approximate incidence of 0.2%) among patients with seizures (1). FOS is a kind of reflex epilepsy of the visual system, where seizures are precipitated by visual stimuli, in this case loss of visual fixation.

In symptomatic epilepsies, FOS has been described in association with Sturge-Weber syndrome, occipital calcifications, celiac disease and malformations of cortical development, especially cortical dysplasia and the subcortical band heterotopia (1, 3, 6); our patients had a history of perinatal hypoxic and bilateral schizencephaly. We did not find any report of FOS in patients with schizencephaly.

Maher et al. (7) studied 31 children with a history of childhood epilepsy with occipital paroxysmal discharges suppressed by eye opening, with normal background activity, and found that 16% of them had a symptomatic epilepsy, suggesting that this pattern was not specific for a single epileptic syndrome, and it could be either idiopathic or symptomatic, even with normal background activity.

Most studies describing FOS in symptomatic epilepsies have been carried out in tertiary epilepsy center, where symptomatic drug-resistant epilepsies are more common. In one of these studies Kaul et al. (8), found 11 patients who exhibited FOS, and 8 of them (72.7%) had symptomatic focal epilepsy. Fattouch et al. (6), evaluated 15 epileptic patients with FOS persisting in adult life, and observed 11 (73.3%) cases with symptomatic epilepsy, concluding that the persistence of the FOS phenomenon in adulthood, unlike the childhood form, may represent the EEG expression of symptomatic epilepsy (6).

Several studies suggest the involvement of occipital hyperexcitability as the cause of FOS in symptomatic epilepsies, however the exact mechanism underlying FOS remains largely obscure (1, 3, 8).

FOS should be differentiated from epileptiform discharges triggered by eye closure, which appear only for a brief period after closing of the eyes (self-limited), are suppressed by completed darkness and typically are associated with photosensitivity (1, 3). FOS should also be differentiated from scotosensitivity, a condition that denotes EEG discharges or epileptic seizures elicited by complete elimination of retinal light stimulation (1-3, 5). Fixation-off sensitivity was documented in this patient, because every time visual fixation was compromised occipital discharges were induced immediately or with a few seconds delay after eye closure, which allowed us to rule out the possibility of eye-closure paroxysms or scotosensitive epilepsy.

FOS-related seizures have been described as myoclonic, absences, and absence status epilepticus (9). There are also cases who presented with transient cognitive impairment, however the overall intrinsic epileptogenic potential of FOS is presumed to be low (1, 3, 4). Apparently this patient did not have any clinical features during FOS, although we could not discard if she had visual hallucinations or any other symptoms due to the lack of collaboration for her mental retardation.

In this case FOS was initially unrecognized, and a misdiagnosis of simple partial status was done; however there was a complete suppression of the discharges after the therapy. In the follow-up video-EEG performed after the patient´s discharge, the occipital paroxysms had re-appeared, displaying some differences relating morphology, a more prolonged latency after eyes closure, and a discontinuous pattern. Maybe these differences could be related to antiepileptic therapy.

In conclusion, occipital paroxysms with FOS can manifest as continuous or repetitive discharges when central vision/fixation is compromised (i.e. eyes closed period), resembling an electrical status epileptic, that disappears upon visual fixation. To recognize this phenomenon, a detailed video-EEG monitoring should be considered in all children with continuous epileptiform discharges during the eye-closed state immediately subsiding with eye opening.

Conflicts of interest

The Authors have no conflicts of interest to declare.

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