Unusual focal ictal symptoms in Jeavons syndrome
Crise focale au cours d'un syndrome de Jeavons

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Abstract:

We report the case of a 16-year-old female with Jeavons syndrome, who presented focal manifestations that could suggest clinically a partial seizure. Jeavons syndrome is typically characterized by eyelid myoclonia with or without absences and bursts of generalized 3-6Hz polyspike-waves complexes precipitated by eye closure and photic stimulation. Generalized seizures, head and Upper limbs jerks can also be associated, but to our knowledge, lateralized jerks, as seen in our patient, have never been described. We underline the importance of video EEG in the diagnosis of these cases.
Key words: Eyelid myoclonia- Absence- Jeavons.

Résumé:
Nous rapportons le cas d'une patiente de 16 ans chez qui le diagnostic de syndrome de Jeavons est retenu et qui a présenté au cours d'une enregistrement vidéoEEG des symptomes focaux évoquant une crise partielle. Le syndrome de Jeavons est caractérisé par des myoclonies palpébrales plus ou moins associées à des absences avec à l'EEG des décharges de polypointes ondes généralisées à la fermeture des yeux, aggravées par la stimulation lumineuse intermittente. Des crises généralisées, mais aussi des myoclonies affectant la tête et les membres peuvent s'observer, mais à notre connaissance, il n'a jamais été rapporté de signes focaux. Nous soulignons à travers cette observation l'intérêt de la vidéo EEG dans le diagnostic de ce type de crises.
Mots clés: Myoclonies palpébrales- Absence- Jeavons.

Introduction:

Eyelid myoclonia with or without absences was first described by Jeavons. It typically associates eyelid myoclonia to brief absences and short bursts of generalized 3-6 Hz polyspike and wave complexes immediately after eye closure. Though it was first described in 1977 [1], it is still not recognized as a distinctive condition. The hallmark of Jeavons syndrome is eyelid myoclonia but many authors reported associated atypical signs. We report the case of a patient with Jeavons syndrome who presented focal ictal signs. We highlight through this case the importance of Video-electroencephalogram (video EEG) before starting therapy in clinically focal seizures.

Case study:

A 16-year-old female complained of involuntary episodes of looking up and blinking occurring up to 10 times a day. The episodes had begun at the age of 14 and were specially triggered by looking at the sun. The patient was aware of the involuntary eye movements but complained sometimes of brief lapses in concentration. Neurological examination was normal. Her past medical history was unremarkable and there was no family history of epilepsy.
Video EEG showed a normal background activity and generalized, high amplitude 3Hz discharges of polyspikes and waves activity immediately after eye closure, which were enhanced during photic stimulation. During these discharges, the patient sometimes presented brief absences. She also presented during photic stimulation an episode of right deviation of the eyes followed by right deviation of the head, then jerks involving the head and the right upper arm (Figures 1 and 2). The patient remained responsive during this episode.

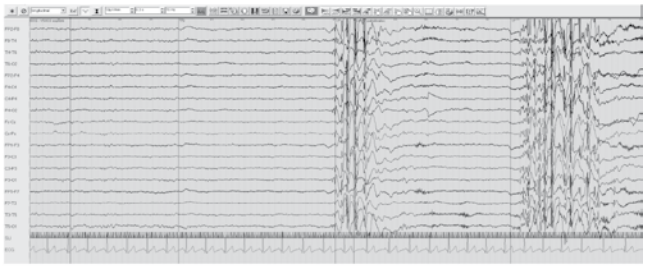

Figure 1 : EEG with intermittent photic stimulation showing normal background with generalized 3Hz discharges of polyspikes and waves complexes on eye closure.

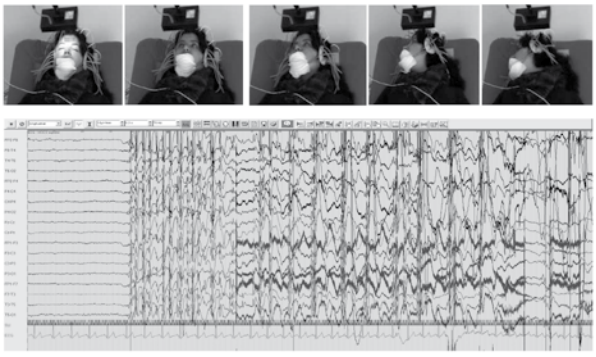

Figure 2 : Sequences of video EEG showing a seizure after eye closure : right deviation of the eye balls, then right deviation of the head followed by jerks of the right arm and the head. EEG : generalized 3Hz polyspikes and waves complexes.

Discussion:

This focal episode, made of right deviation of the eyes then the head with jerks of the head and right arm, could have suggested "clinically" a partial seizure. However, the diagnosis of Jeavons syndrome was made on the basis of clinical and EEG features: eyelid myoclonia with or without absences (EMA) and bursts of 3Hz generalized polyspike-wave complexes precipitated by eye closure and photic stimulation.

Though many authors reported cases and series of Jeavons syndrome, there are still controversies about whether or not it should be recognized as an independent epileptic syndrome or only a type of seizures that might be seen in many epileptic syndromes [1] .

In a study of 63 cases of Jeavons syndrome, Caraballo and al. suggest that there might be 2 groups of Jeavons syndrome: a first group where EMA could be considered as a photosensitive idiopathic epileptic syndrome (including a subgroup of early-onset of EMA associated or not with GTCS and mental retardation). A second group where EMA may correspond to a type of seizures in idiopathic generalized epilepsies [2].

In Jeavons syndrome, eye jerks are typically associated with upward déviation of the eyeballs. Head jerks may also be seen [3]. Myoclonic jerks occur rarely in EMA and are usually restricted to the upper extremities [4], associated development delay was also reported [5], but to our knowledge, lateralized symptoms, as seen in our patient, have never been reported.

We underline the importance of video EEG in the diagnosis of such conditions and ruling out partial seizures. In our case, clinical features suggesting a partial seizure could have led to start Carbamazepine or Oxcarbamazepine which are known to aggravate seizures in this syndrome [6].

Sodium Valproate was started in our patient. The outcome was favorable, but as the patient gained weight. We switched to Levetiracetam which resulted in poor response, then to Lamotrigin with good outcome.

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