Ictal miosis – a rare symptomatology of focal autonomic seizures.

Napadowe zwężenie źrenic – rzadka symptomatologia napadów ogniskowych.

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ABSTRACT

Epileptic seizures, manifested by pupils' size alteration without impairment of consciousness, may remain unnoticed or be mistaken with other nonepileptic events. There is a lack of comprehensive descriptions and evidence in the literature regarding seizures with pupillary autonomic manifestation as the isolated phenomena. This fact indicates the need of reporting the cases of focal autonomic seizures, clinically manifesting with pupillary changes, which should be discussed in order to remain aware of this rare seizure symptomatology and asses their actual frequency in a longer perspective.

Therefore, we present clinical and diagnostic implications regarding a 3,5-year-old boy, diagnosed with focal seizures with autonomic component. There were no pregnancy and delivery complications and the psychomotor development was also normal. Interictal EEG revealed sharp waves in occipital and parieto-occipital regions, pronounced over the right hemisphere. MRI of the brain was normal. Hospital observation and video recording confirmed focal seizures with autonomic manifestation. He was successfully treated with carbamazepine and remains seizure free.

Our findings provide the evidence that focal epileptic seizures are elusive and difficult to recognize. Capturing the seizures on video monitoring isessential to the diagnosis. Careful observation, proper diagnosis and early implementation of antiepileptic therapy may provide satisfactory and long-lasting improvement in managing the seizures.

Key words: autonomic seizures, pupillary changes, ictal miosis, blurry vision, ictal spinning.

STRESZCZENIE

Napady padaczkowe, manifestujące się pod postacią zmiany szerokości źrenic, bez zaburzeń świadomości, mogą pozostać niezauważone lub niewłaściwie rozpoznane jako incydenty niepadaczkowe. W piśmiennictwie niewiele jest prac dotyczących napadów manifestujących się w postaci izolowanych zaburzeń autonomicznych, w tym napadowych zmian szerokości źrenic. Fakt ten wskazuje na utrzymującą się potrzebę dyskusji i uzupełniania literatury o doniesienia na temat napadów ogniskowych z objawami autonomicznymi, manifestujących się w postaci napadowych zmian szerokości źrenic, celem oszacowania rzeczywistej częstości występowania tego typu napadów.

Mając na uwadze powyższe, przedstawiamy obraz kliniczny oraz implikacje diagnostyczne, dotyczące przypadku 3,5-letniego chłopca, bez obciążeń w wywiadzie ciążowo-okołoporodowym, rozwijającego się prawidłowo, z rozpoznaną padaczką z napadami ogniskowymi, manifestującymi się w postaci zaburzeń autonomicznych. W międzynapadowym zapisie EEG u chłopca stwierdzono fale ostre w okolicach ciemieniowo-potylicznych oraz potylicznych, głównie znad prawej półkuli mózgu. MRI mózgowia był prawidłowy. Obserwacja kliniczna oraz nagrania video potwierdziły występowanie napadów ogniskowych z objawami autonomicznymi, bez zaburzeń świadomości. Włączono terapię karbamazepiną, uzyskując ustąpienie napadów padaczkowych.

Przedstawione przez nas dane kliniczne oraz wyniki badań ukazują, że semiologia występujących u chłopca ogniskowych napadów z komponentą autonomiczną możne stanowić duże wyzwanie diagnostyczne/budzić duże trudności diagnostyczne. Nagranie na video napadów o tej rzadkiej manifestacji klinicznej może stanowić podstawę ich rozpoznania i zaklasyfikowania. Uważna obserwacja kliniczna oraz właściwa diagnoza pozwala na szybkie rozpoczęcie właściwej i skutecznej terapii przeciwpadaczkowej.

Słowa kluczowe: napady autonomiczne, zmiany szerokości źrenic, napadowe zwężenie źrenic, zamazane widzenie, napadowe uczucie wirowania.

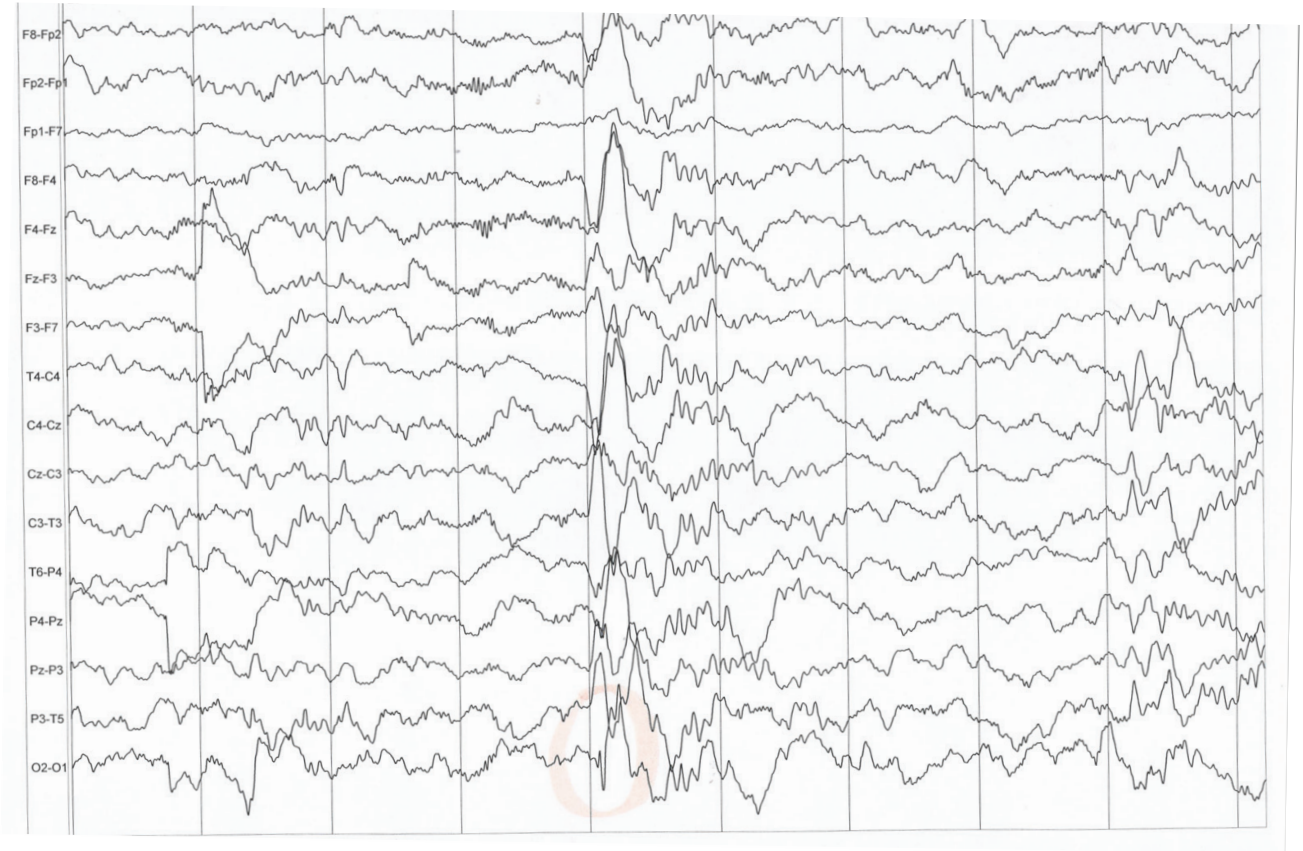
According to the ILAE glossary, autonomic seizures are defined as an objectively documented and distinct alteration of autonomic nervous system function involving cardiovascular, pupillary, gastrointestinal, sudomotor, vasomotor and thermoregulatory functions [1, 2]. Although autonomic signs or symptoms accompany a great number of focal seizures, they rarely occur as an isolated phenomenon without impairment of consciousness. Usually, they appear in a various combination with other neurological symptoms, but can also occur solely as we would present. Pure autonomic seizures, regarding sudden alterations of size of pupils are a rare situation and remain a great challenge to recognize as the unusual manifestation of epilepsy, both in adults and children [2, 3].

Only a few reports have described ictal isolated pupil miosis without impairment of awareness in a child [4, 5]. Because of the unusual and uncertain symptomatology the seizures may be masked and wrongly diagnosed as a nonepileptic event. For this reason we would like to share our clinical and diagnostic findings regarding this rare manife-

station of focal seizures. We report a case of a child who experienced ictal events, manifested by sudden isolated miosis of pupils, which consistently led us to the diagnosis of focal autonomic seizures.

Three-and-a-half-year-old boy was referred to the Department of Developmental Neurology due to the complaints of a sudden blurry vision and a feeling of spinning. These incidents started about five months before with unstable frequency, from once a month at the beginning, up to every day later, and lasting to 10–15 seconds. Parents noticed that during events the pupils were narrow. The boy remained in a logical contact, responded to the simple questions correctly, precisely indicating the beginning and the end of an incident, corresponding temporarily to the pupil contraction and a final physiological dilatation, adequate to the external lumination. The incidents occurred only in awaking state during a day. Medical history revealed uncomplicated pregnancy and delivery, as well as a correct psychomotor development. Family history did not indicate any neurological disease, including seizures. The boy was previously healthy and did not suffer from any other diseases or illnesses. The last ictal incident occurred about a week prior to hospitalization. On admission to the clinic, the neurological and developmental status was normal and adequate to the age. Pediatric examination also did not reveal any pathological signs or symptoms. Clinical observation and video recording revealed a focal aware seizures with autonomic component, corresponding to the medical history (a video recording in attached file). Interictal EEG revealed bilateral sharp waves in occipital and parieto-occipital regions, more pronounced on the right side, with a slight tendency to synchrony (Fig. 1). Attempts to record ictal EEG were unsuccessful despite prolonged recording. Brain MRI did not reveal any structural abnormalities. On the base of the clinical and EEG findings, a carbamazepine therapy was introduced. The boy was discharged home and advised for a review to assess the initial response. On a following medical evaluation 9 months later he remained seizure free.

The clinical manifestation of focal autonomic seizures refers mostly to cardiovascular, respiratory, gastrointestinal, urinary, vasomotor, pilomotor, secretory or pupillary changes. Alterations of autonomic functions occur frequently in the course of many types of epileptic seizures and stand for these seizures' component. However, the autonomic symptomatology sometimes subtle and equivocal, may be the sole manifestation of seizures. On the one hand, the essential task is to distinguish these accompanying and secondary variations of autonomic system in a course of many seizure types from congruent autonomic seizures [2, 4, 6]. This matter specially refers to temporal lobe epilepsies in which autonomic features are often the initial and apparent component of a seizure (ictal vomiting, hypersalivation) [2, 7–9]. Autonomic symptoms in focal seizures are also thought to have localizing value, however this thesis requires further investigations [10, 11]. Other epilepsies with prominent autonomic features refer to various types of neonatal seizures, focal epilepsies of infancy, Dravet



syndrome or different epilepsies associated with specific genetic mutations [2, 3]. Nevertheless, the other viewpoint is that isolated autonomic phenomena standing for epileptic seizures, may remain unnoticed, especially in small children. The same may refer also to adults who may consider this seizure with a full awareness as the experience of strange transient feeling or impression. It is also essential for the physicians to be conscious of this infrequent manifestation of epilepsy, especially that a symptomatic etiology need to be excluded. Our patient presents interictal EEG discharges in occipital and parieto-occipital regions. Autonomic symptomatology in occipital epilepsies is widely described in aspect of benign childhood occipital seizures (BCOS), such as Panayiotopoulos or Gastaut syndrome [3, 12]. Idiopathic photosensitive occipital seizures are also well documented [13, 14]. Similarly, symptomatology regarding visual hallucinations or ictal nonepileptic events imitating occipital semiology (such as migraine attacks with vomiting and headache) are widely known [12, 15]. However, reports in literature regarding other symptomatology, especially ictal miosis, even accompanied by other occipital seizure manifestation (e.g. feeling of spinning) have been still scarse [3, 16, 17]. There are some reports regarding ictal mydriasis as the autonomic manifestation of seizures as a result of sympathetic neurons excitation [2, 18]. Little is known about pupillary construction mechanisms during epileptic seizure. Sadek *et al.* described a patient with ictal miosis due to the focal cortical dysplasia and claimed that epileptogenic zone in this kind of seizures may be located in the left middle parietal gyrus, which was proved in electrocorticography [4]. Other authors reported a few cases with epileptic seizures manifested by pupillary constriction, however, due to the small number of patients, the results regarding the mechanism and precise localisation are not coherent [4, 5]. These scarse reports confirm the necessity of heedful observation and reporting this rare manifestation of epilepsy.

Although the presence of correlating seizures and interictal discharges in EEG contributed to the final conclusions, it would be much more difficult to diagnose the disease without clinical monitoring or capturing the seizure on video recording. Modern video techniques and their accessibility allow to document ictal events which relevantly improved our knowledge of epileptic seizures semiology. Overall, careful observation, accurate diagnosis and early implementation of antiepileptic therapy may provide satisfactory outcome in managing the seizures.

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