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PANAYIOTOPOULOS SYNDROME A COMMON DIAGNOSIS SOMETIMES A DIAGNOSIC DIALEMMA TO PRACTITIONERS.

Pediatrics

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ABSTRACT

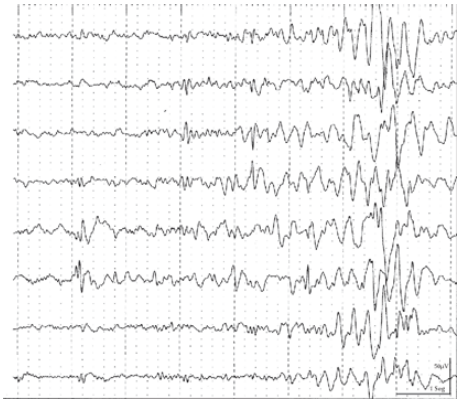
Panayiotopoulos Syndrome , also known as early onset occipital epilepsy, is a common childhood epilepsy syndrome with partial seizures. It emerges in mid-childhood usually between the ages of 3-10 years. Both boys and girls can develop this syndrome. We are reporting a case of 7 year old female child with focal seizures later became generalized and was associated with autonomous symptoms and brief loss of consciousness, The interictal EEG recording during sleep shows multifocal spikes and diffuse spike-and-wave discharges in the occipital region which is suggestive of Panayiotopoulos Syndrome.

KEYWORDS

Panayiotopoulos , EEG, Occipital Epilepsy, Diffuse Tensor Imaging

Panayiotopoulos syndrome (PS) is a common benign epileptic syndrome of childhood that affects 2-3/1000 children .(1) Panayiotopoulos syndrome is a common idiopathic, age and self-limited, common in childhood, benign condition with focal autonomic seizures and sometimes present as autonomic status epilepticus. It affects around 6% of children with aypical febrile seizures. Although it was described in 1989, in 2001 it was described as a electroclinical syndrome of childhood by the "International League against Epilepsy". (2) Because of atypical manifestations, it is not uncommon for PS to be diagnosed sometimes and even remain unrecognized , seizures manifest with a wide spectrum of autonomic manifestations mainly vomiting, syncope like symptoms , mydriasis, urine and fecal incontinence and cardiorespiratory irregularities. Half of the seizures last for more than 60 minutes, often 2 to 3 hours. There is marked variability of interictal EEG findings, from normal to multifocal spikes that can vary in serial EEGs. Ictal EEG onset is from the frontal or posterior regions.

We present a female child 7 years old, with clinical symptoms of fever, cold, cough, excessive sweating, vomiting and diarrhea with focal seizures of upper extremities later became generalized, later associated with brief loss of consciousness for 3 minutes, there was a similar history in her 3 months back at home which was not taken to the notice of health care provider, the other systemic examination was normal except for some dehydration. Complete blood count, RBS, Serum calcium and electrolytes were within normal limits, when electroencephalographic was taken the findings were consistent with Panayitopoulos syndrome. The interictal EEG recording during sleep shows multifocal spikes and diffuse spike-and-wave discharges in the occipital region. A normal MRI is the usual finding in patients with PS, abnormalities like gliosis, developmental anomalies and cortical malformations are sometimes picked up incidentally. Diffusion tensor imaging is more specific which shows attenuation of fibers over the temporal-occipital area and decreased thickness of the occipital lobe as shown in other studies(3),which are routinely done in the western setting, couldn't be done in our case. As there was no family history of epilepsy, infrequent nature of the disease and the prognosis being good, the parents were counselled and no antiepileptic was started.



DISCUSSION:

PS usually presents in the age group of 1–14 years, with 13% of the cases occurring between 3 and 6 years. The pathophysiology involves an epileptogenic activation of the low threshold central autonomic areas, but the signals are not strong enough to activate the cortical areas that usually manifest with motor or sensory symptoms. Autonomic epileptic seizures are the cardinal manifestations, with vomiting being the predominant symptom. Other autonomic symptoms which children can present with are mydriasis or miosis, pallor, cyanosis, flushing, cardiorespiratory variations like apnea with alterations in heart rate and thermoregulatory alterations, urinary with or without fecal incontinence, excessive salivation and sometimes altered intestinal motility (4). In 2006, an expert consensus defined PS as: "A benign age-related focal seizure disorder occurring in early and mid childhood. It is characterized by seizures often prolonged, with predominantly autonomic symptoms, and by an EEG that shows shifting multiple foci, often with occipital predominance. Benign occipital epilepsy of childhood, characterized by occipital paroxysms on EEG was first described by Gastaut(5). It is characterized by seizures that start with visual symptoms, which often are followed by hemi-clonic seizures and, in some cases migranous headaches. Prognosis of Panayiotopoulos syndrome is absolutely benign in terms of seizure frequency and subsequent evolution. Autonomic status epilepticus usually has no residual neurologic deficits.(6) The differential diagnoses to be considered in these cases are Migraine, Encephalitis, Gastro-esophageal reflux disease and Psychiatric manifestations. In case of frequent seizures, benzodiazepines can be used. Prophylactic therapy with antiepileptics is indicated when seizures are unusually frequent or otherwise significantly affecting the child's quality of life. Carbamazepine, sodium valproate, and Levetiracetam have been used in various studies with better results. (7) In photosensitive occipital seizures, avoidance of precipitating factors usually be sufficient(8,9)but if seizures persist, long term sodium valproate therapy was the drug of choice in few studies.(10) The risk of epilepsy in adult life is same as general population. Counselling of the parents about its benign nature and very good prognosis are important aspects of management.

Conclusion: Panayiotopolous syndrome is a clinically recognizable treatable epilepsy of early childhood. The typical presentation is that of autonomic seizures and ictal vomiting with or without EEG abnormalities. Counselling of parents should be done appropriately and the child should be advised to be on regular follow up. Physicians should know the varied presentations of this syndrome to ensure early diagnosis and proper management to prevent neurological and physical morbidities and when in doubt the expert opinion is taken at the earliest.

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