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ORIGIN OF FRONTAL AND TEMPORAL LOBE SPIKES IN EARLY-ONSET BENIGN OCCIPITAL LOBE EPILEPSY - PANAYIOTOPOULOS SYNDROME

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Abstract

Panayiotopoulos syndrome is a common idiopathic childhood-specific seizure disorder formally recognized by the International League Against Epilepsy. Panayiotopoulos syndrome is defined 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 and/or multiple foci, often with occipital predominance. This is a case report of an early adolescent girl came with autonomic complaints and was diagnosed as Panayiotopoulos syndrome and was started on antiepileptics.

Keywords: Panayiotopoulos Syndrome (PS), Focal Seizures, Self-limited Focal Epilepsies of Childhood (SLEC), Benign Childhood Seizure Susceptibility Syndrome (BCSSS), Genetic Predisposition, SCN1A Gene Mutations, Autonomic Manifestations, Afebrile Nonconvulsive Status Epilepticus,Epileptiform Discharges, Frontotemporal Spikes.

INTRODUCTION

Panayiotopoulos syndrome (PS) has been documented in medical literature since 1984, with Chrysostomos Panayiotopoulos's seminal work in 1989 defining it as a distinct epileptic syndrome , [1]. Formal recognition by the International League Against Epilepsy (ILAE) came in 2001, designating PS as "early-onset benign childhood occipital epilepsy" [2,3]. Over the past two decades, numerous retrospective and prospective studies have enhanced our comprehension of this syndrome [4-7]. In the 2017 ILAE Classification of Epilepsies, PS was classified among the self-limited focal epilepsies of childhood (SLEC) [8]. More recently, the Nosology and Definitions ILAE Task Force 2021 proposed the term "self-limited epilepsy with autonomic seizures", yet for consistency with Dr.Panayiotopoulos's pioneering contributions, we utilize the term PS in this study .

PS is believed to be part of a broader benign childhood seizure susceptibility syndrome (BCSSS), influenced by genetic factors and characterized by age-related and age-limited features. It shares connections with self-limited epilepsy with centro-temporal spikes (SLECTS) and late-onset self-limited childhood occipital epilepsy Gastaut type (SLE-G) [9,10]. Despite considerable advancements, the exact etiology of PS remains elusive. While a familial history of epilepsy or febrile seizures suggests a genetic

predisposition, the mode of inheritance appears multifaceted. Some patients have been found to carry mutations in the SCN1A gene [11,12].

Clinically, PS is typified by seizures that are often prolonged, infrequent, and predominantly nocturnal, presenting with focal onset and primarily autonomic symptoms. Pallor, nausea, and vomiting are common autonomic manifestations, often accompanied by eye and head deviation and generalized hypotonia. While at seizure onset consciousness is usually preserved, partial or complete unresponsiveness ensues in the majority of patients [12,13]. Approximately 75% of children with PS experience emetic symptoms and pallor alongside additional autonomic features, frequently leading to misdiagnosis. Nearly half of the seizures extend beyond 30 minutes, with PS constituting a significant cause of afebrile nonconvulsive status epilepticus in childhood [14,15].

CASE PRESENTATION

A young adolescent girl presented to the outpatient department accompanied by her parents, reporting recurrent episodes of vomiting and headache spanning over a period of four years. She described experiencing intermittent headaches localized in the frontal and temporal regions and exhibited sensitivity to loud sounds. Notably, there were no instances of loss of consciousness, involuntary micturition, or tongue biting during these episodes. Furthermore, the episodes did not display any diurnal variation, and there were no apparent psychosocial stressors or signs of anxiety in between episodes. The patient had been experiencing these recurrent episodes since early childhood and had no prior history of medical or psychiatric illnesses, nor was there a family history of such conditions. Investigations revealed normal results from basic blood tests and upper gastrointestinal endoscopy. However, a computed tomography (CT) scan of the abdomen showed bilateral polycystic ovaries. Electroencephalography (EEG) findings indicated generalized epileptiform discharges with frontotemporal spikes (figure *1*), while magnetic resonance imaging (MRI) of the brain yielded normal results.

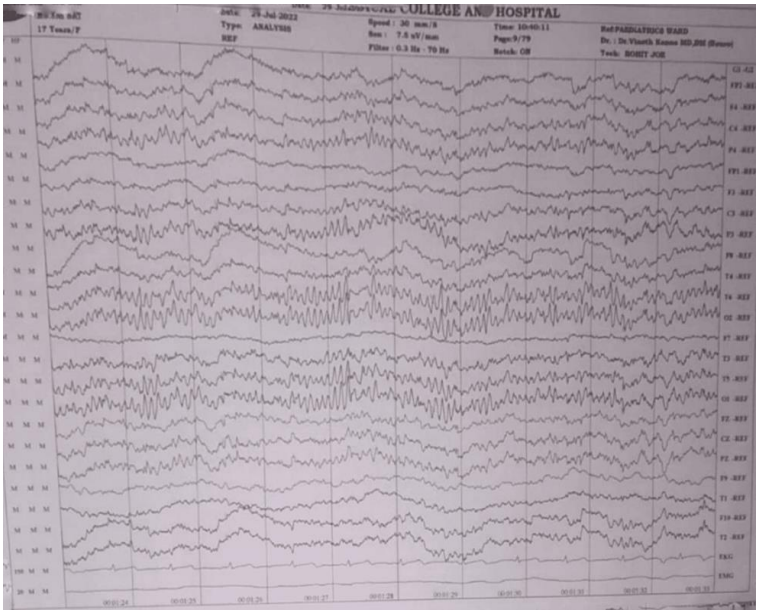

Figure 1: EEG PICTURE showing generalized epileptiform changes in frontotemporal regions

Initially diagnosed with migraine, the patient received treatment with propranolol and flunarizine, which proved ineffective. Subsequently, a provisional diagnosis of cyclical vomiting was considered, leading to treatment with antiemetics, pantoprazole, and antispasmodics, also without success. Based on the EEG findings, the patient was started on Levetiracetam, resulting in a reduction in symptom frequency. Following a six-month follow-up, the patient remained free from further episodes. It was emphasized that education about Panayiotopoulos syndrome is essential for its management, and prophylactic treatment with antiepileptics may not be necessary for most patients.

Differential Diagnosis

Diagnosis may be easily missed - mild and brief ictal autonomic symptoms in presence of clear consciousness suggest non epileptic conditions such as gastroenteritis, syncope, atypical migraine; while prolonged severe attacks may simulate life threatening insults such as encephalitis.

DISCUSSION

Panayiotopoulos syndrome, is a rare form of childhood epilepsy characterized by focal seizures, which originate from one area of the brain [1,2]. These seizures often involve complex visual hallucinations, autonomic symptoms like vomiting or sweating, and altered consciousness. Panayiotopoulos syndrome typically occurs in children between the ages of 3 and 10 years old, and the seizures may be prolonged, sometimes lasting for several minutes. They can occur during sleep or upon awakening and may progress to status epilepticus in some cases. The exact cause of Panayiotopoulos syndrome is not fully understood, but it is thought to be related to abnormal electrical activity in the brain. Diagnosis is based on clinical history, observation of seizure characteristics, and EEG findings.Various unusual clinical ictal features have been reported in PS, including pure autonomic status epilepticus, coughing, fever, respiratory or cardiorespiratory arrest, and other manifestations. While occipital spikes were predominant in the interictal EEG of PS, extra-occipital spikes, such as temporal and frontal spikes, were also commonly observed, further complicating diagnosis [16].

Although Panayiotopoulos syndrome is characterized by occipital spikes Leal AJ et al have studied that frontal spikes in these patients represent secondary activation triggered by occipital inter ictal discharges and do not represent an independent focus [17]. Hogarth et al in their study reports that only 10% of PS has temporal inter ictal electrical activity. This is in line with our findings where frontal temporal spikes were noted [18]. Treatment usually involves antiepileptic medications to control seizures, and most children with Panayiotopoulos syndrome have a good prognosis, with many outgrowing the condition by adolescence. However, close monitoring and management by healthcare professionals are important to ensure optimal outcomes and minimize the impact of seizures on the child's life.

CONCLUSIONS

Panayiotopoulos syndrome is remarkably benign. Remission usually occurs within two years from onset. A third of these children have a single seizure, and only 5-10% have more than 10 seizures that may be very frequent sometimes but the outcome is still

favourable. One fifth of children with this syndrome may develop other types of infrequent, usually Rolandic seizures, but these also remit before the age of 16 years.

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