

Case Report

Ictal Cardiorespiratory Arrest Associated with Status Epilepticus in Panayiotopoulos Syndrome

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A healthy 10-year-old boy vomited during sleep and later complained of abdominal pain; he became drowsy and uncommunicative. At the nearby hospital E.R., he deteriorated rapidly, and his respiratory movements were absent with cardiac arrest. He was immediately resuscitated. Brain MRI showed no abnormalities. EEG revealed an abnormal pattern with recurrent multifocal epileptiform activity over the bilateral occipital and frontal regions during sleep. Based on the clinical/radiological findings we diagnosed Panayiotopoulos syndrome (PS), a benign form of early-onset pediatric epilepsy characterized by autonomic symptoms. Life-threatening cardiopulmonary arrest is rare in PS, but long seizure duration of PS may associate with apnea and bradycardia.

Key words: pediatric epilepsy, autonomic symptom, cardiac arrest, afebrile seizure, EEG

Chrysostomos Panayiotopoulos first described the condition named Panayiotopoulos syndrome, defined by a trio of clinical symptoms: nocturnal seizures, tonic eye deviation, and vomiting [1]. The majority of the seizures in this syndrome start with autonomic signs that occur during sleep, either individually or in combination with behavioral symptoms such as loss of consciousness [2]. In general, 50% of the seizures progress to secondary generalized seizures. Although Panayiotopoulos syndrome is a common condition affecting about 13% of children in the 3- to 6-year-old age group who present with one or more afebrile seizures, it is often misdiagnosed despite its lengthy and dramatic manifestations [3].

Here, we describe the case of a 10-year-old boy with

Panayiotopoulos syndrome who presented with ictal cardiorespiratory arrest and status epilepticus needing cardiopulmonary resuscitation for revival. Autonomic status epilepticus has been reported in patients with Panayiotopoulos syndrome, but ictal cardiorespiratory arrest is very rare in this syndrome [4]. Sharing our experience may help emergency physicians accurately diagnose this syndrome and initiate the correct treatment/follow-up plan with consultation from pediatric neurologists.

Presentation of the Case

The patient was a 10-year-old Japanese boy with normal psychomotor activity in school and at home. His birth and early life history were unremarkable

(height, 133 cm; body weight 27.2 kg). His past medical history included an episode of syncope 6 months prior to his admission to our emergency room (E.R.); the syncope was concluded to be of unknown etiology based on radiological and electroencephalogram examinations. Since then, no epilepsy was observed. On the night before his E.R. admission, he had vomited at 4 a.m. He fell asleep again, but woke up 2 h later complaining of nausea and abdominal pain. Soon after, he became drowsy and uncommunicative, and his parents called an ambulance. In the ambulance, his oxygen saturation was observed to be 70%, and high-flow oxygen was then initiated by an emergency medical technician.

Upon arrival at our E.R. 10 min later, the patient was still unresponsive with 78% oxygen saturation under the administration of oxygen by face mask. On examination, he was bradycardic with labored respiration. His Glasgow Coma Scale score was 6 (E1V1M4). When passively opened, his eyes deviated to the right and were flaccid; he had feces and urine incontinence. His glucose levels, calcium, electrolytes, and magnesium were within normal ranges. The analysis of blood gas revealed severe respiratory acidosis; pH 7.077,

pCO₂ 103.4 mmHg, pO₂ 58.4 mmHg, and Base Excess 3.1 mmol/L. He deteriorated rapidly in the E.R., and absent breathing and a lack of carotid pulse in the presence of cardiac electric activity was noted (cardiorespiratory arrest status in pulseless electrical activity).

After successful resuscitation based on basic life support by chest compression, intubation, and mechanical ventilation, the patient was transferred to the intensive care unit with the diagnosis of autonomic status epilepticus. The electrocardiogram (ECG) 15 min after the recovery of spontaneous circulation and a brain magnetic resonance imaging (MRI) scan showed no abnormalities. Cerebrospinal fluid tests were normal. Eight h later, the patient had completely recovered and was extubated without abnormal neurological findings.

The electroencephalogram (EEG) performed 2 days later showed an abnormal pattern with recurrent multifocal epileptiform activity during sleep over the bilateral occipital and frontal regions (Fig. 1). Based on the clinical/radiological findings, we made the diagnosis of Panayiotopoulos syndrome. Daily valproic acid (500 mg/day) was administered. The patient continued to remain healthy without seizures or neurological deficits at his last visit during a 10-month follow-up.

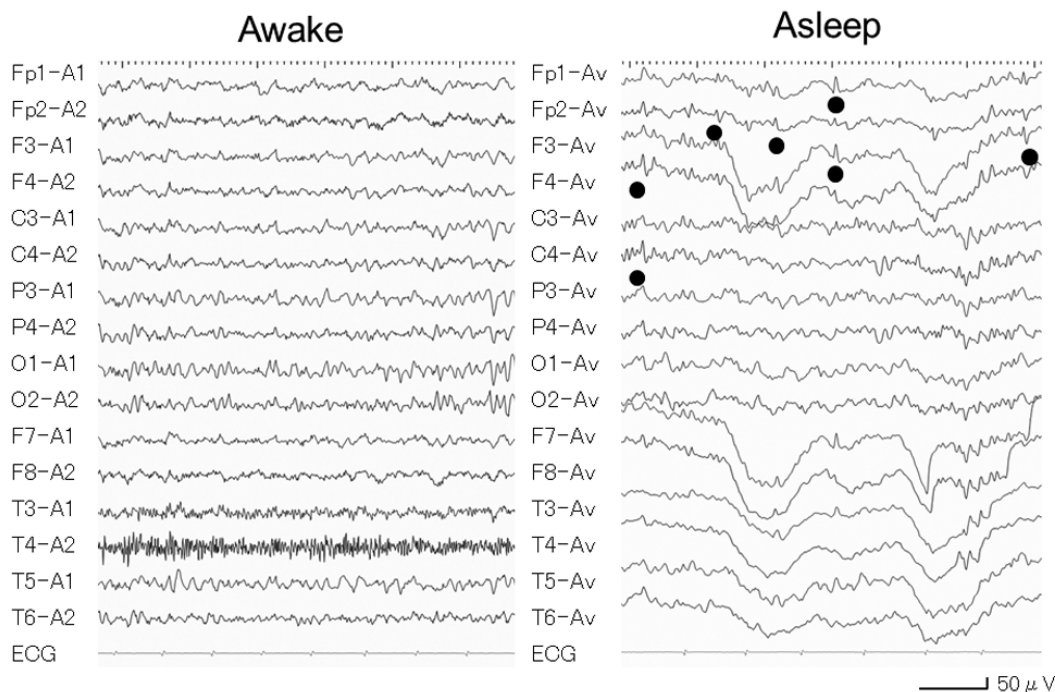


Fig. 1 The EEG demonstrated an abnormal pattern with recurrent multifocal epileptiform activity during sleep over the bilateral occipital and frontal regions.

Discussion

Panayiotopoulos syndrome is a common type of benign childhood early-onset epilepsy characterized by a variety of autonomic symptoms. Panayiotopoulos syndrome typically starts at the peak age of 3-6 years old (range 1-14 years old) with no apparent gender or race prevalence [5]. Panayiotopoulos syndrome is specific to childhood; it does not occur in adults [6]. Seizures in Panayiotopoulos syndrome occur predominantly during sleep, particularly the early part of sleep. This can be both nocturnal sleep and daytime naps [7]. The pathophysiology of autonomic seizures in Panayiotopoulos syndrome is unclear.

Vomiting, one of the autonomic symptoms, is a common manifestation of several organic and functional disorders and has received little attention in epilepsy. Vomiting combined with seizures raises the suspicion of increased intracranial pressure. If no structural pathology is found, vomiting occurring during an epileptic fit may be considered evidence of a coexisting migraine or as seizure activity consequent to migraine. Clinicians should be aware of the varying manifestations of this syndrome (particularly the autonomic characteristics), since these manifestations are often incorrectly diagnosed as motion sickness, cyclical vomiting syndrome, encephalitis, acute gastroenteritis, migraine, and syncope. Misdiagnosis can lead to prolonged hospitalization and incorrect treatment with greater cost burden. Our patient initially presented with cardiorespiratory arrest, but subsequently experienced typical seizures. Our initial differential diagnoses were a choking spell, encephalitis, and epilepsy. This case highlights the importance of EEG recordings for focal seizures with autonomic symptoms, which may contribute to the diagnosis of Panayiotopoulos syndrome. Moreover, clinicians should recognize the possibility of the rare but potentially life-threatening occurrence of clinically relevant ictal cardiorespiratory arrest in Panayiotopoulos syndrome.

Panayiotopoulos syndrome has been diagnosed based on the criteria of at least one afebrile seizure with autonomic manifestations, with or without ictal clinical manifestations including motor phenomena or eye deviation, followed by impaired consciousness with secondary generalization. Unremarkable brain imaging, normal neurological development, and occipital and extra-occipital spikes at interictal EEG are typical find-

ings of this syndrome.

Seizures in Panayiotopoulos syndrome are typically long. Nearly half (44%) of the seizures last > 30 min [8]. Panayiotopoulos syndrome is the most common specific cause of afebrile seizures lasting > 30 min or recurrent seizures lasting a total of > 30 min [9]. Cardiorespiratory arrest is rare; life-threatening cardiorespiratory arrest occurs in up to 1 per 200 individuals with Panayiotopoulos syndrome [6].

Sudden unexplained death in epilepsy is a nontraumatic unwitnessed death that occurs in epileptic patients previously found to be relatively healthy, with no cause founded even after autopsy. Since the recognition of sudden unexplained death in epilepsy, seizure-related cardiorespiratory abnormalities have been a research focus [10]. The risks of ictal apnea or ictal bradycardia are increased if the patient had a longer duration of seizure, has desaturation, or takes more antiepileptic drugs [11]. Ictal asystole, an often-un-sought autonomic phenomenon, occurs most commonly with temporal followed by frontal lobe seizures. Ictal asystole is more common compared to ictal atrioventricular block and ictal bradyarrhythmias.

In our patient, bradypnea resulted in severe respiratory acidosis, requiring intubation and ventilator support. Although ictal breathing abnormalities and cardiorespiratory arrest are recognized with autonomic seizure manifestations in Panayiotopoulos syndrome, only a few such cases have been reported [4, 12].

Panayiotopoulos syndrome is remarkably benign in terms of seizure frequency and evolution [6]. One-third (27%) have a single seizure only, another half (47%) have 2-5 seizures. Only 5% have > 10 seizures [8]. Ninety percent of patients go into complete remission within 1-2 years of onset [5]. The risk of epilepsy in adult life seems to be no higher than that in the general population [6]. Prophylactic treatment with antiepileptic medication may not be needed for most patients, and aggressive treatment should be avoided due to the risk of drug-induced complications. However, considering the potential risk of cardiorespiratory arrest in Panayiotopoulos syndrome, further reports and studies are needed.

In conclusion, we have described the case of a 10-year-old boy with Panayiotopoulos syndrome presenting with status epilepticus and ictal cardiorespiratory arrest. Cardiac arrest can be associated with Panayiotopoulos syndrome; EEG and ECG monitoring

are critical. Although rare, ictal cardiorespiratory arrest can be potentially life-threatening in children with Panayiotopoulos syndrome and should be noted.

References

1. Panayiotopoulos CP: Vomiting as an ictal manifestation of epileptic seizures and syndromes. *J Neurol Neurosurg Psychiatry* (1988) 51: 1448–1451.
2. Panayiotopoulos CP, Michael M, Sanders S, Valeta T and Koutroumanidis M: Benign childhood focal epilepsies: Assessment of established and newly recognized syndromes. *Brain* (2008) 131: 2264–2286.
3. Panayiotopoulos CP: Benign childhood epilepsy with occipital paroxysms: A 15-year prospective study. *Ann Neurol* (1989) 26: 51–56.
4. Mujawar QM, Sen S, Ali MD, Balakrishnan P and Patil S: Panayiotopoulos syndrome presenting with status epilepticus and cardiorespiratory arrest: A case report. *Pediatr Emerg Care* (2011) 27: 754–757.
5. Michael M, Tsatsou K and Ferrie CD. Panayiotopoulos syndrome: An important childhood autonomic epilepsy to be differentiated from occipital epilepsy and acute non-epileptic disorders. *Brain Dev* (2010) 32: 4–9.
6. Covanis A: Panayiotopoulos syndrome: A benign childhood autonomic epilepsy frequently imitating encephalitis, syncope, migraine, sleep disorder, or gastroenteritis. *Pediatrics* (2006) 118: e1237–1243.
7. Ferrie C, Caraballo R, Covanis A, Demirbilek V, Dervent A, Kivity S, Koutroumanidis M, Martinovic Z, Oguni H, Verrotti A, Vigevano F, Watanabe K, Yalcin D and Yoshinaga H: Panayiotopoulos syndrome: a consensus view. *Dev Med Child Neurol* (2006) 48: 236–240.
8. Panayiotopoulos CP: Autonomic seizures and autonomic status epilepticus peculiar to childhood: diagnosis and management. *Epilepsy Behav* (2004) 5: 286–295.
9. Okanishi T, Maegaki Y, Ohno K and Togari H: Underlying neurologic disorders and recurrence rates of status epilepticus in childhood. *Brain Dev* (2008) 30: 624–628.
10. Goldman AM: Mechanisms of sudden unexplained death in epilepsy. *Curr Opin Neurol* (2015) 28: 166–174.
11. Singh K, Katz ES, Zarowski M, Loddenkemper T, Llewellyn N, Manganaro S, Gregas M, Pavlova M and Kothara SV: Cardiopulmonary complications during pediatric seizures: a prelude to understanding SUDEP. *Epilepsia* (2013) 54: 1083–1091.
12. Verrotti A, Salladini C, Trotta D, di Corcia G and Chiarelli F: Ictal cardiorespiratory arrest in panayiotopoulos syndrome. *Neurology* (2005) 64: 1816–1817.