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Case Report

Panayiotopoulos syndrome presenting with respiratory arrest: A case report and literature review

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ABSTRACT

We describe a child with Panayiotopoulos syndrome (PS) who presented with autonomic status epilepticus and developed respiratory arrest requiring intubation and mechanical ventilation. Because of that life-threatening episode and the risk of developing a similar event in subsequent seizures, we decided to initiate our patient on AED treatment. Such life-threatening complications were previously reported in only four children with PS. Although PS is considered to be a benign childhood epilepsy syndrome usually not requiring treatment with antiepileptic drugs, our case and the small number of similar cases in the literature show it is important to realize that it can rarely be associated with life-threatening complications. It is our opinion that children with PS who develop an episode of autonomic status epilepticus and those living in remote areas with no quick access to emergency departments should be initiated on AED therapy to minimize the risk of experiencing a subsequent potentially fatal seizure. We further suggest that the use of benzodiazepines in this syndrome should only be administered during the early stage of the seizure, since administration of this class of drugs during an established autonomic status epilepticus can result in further respiratory depression.

1. Introduction

Panayiotopoulos syndrome (PS), one of the benign childhood seizure susceptibility syndromes (BCSSS), is an idiopathic, probably genetically determined seizure disorder and recognized as an electroclinical syndrome of childhood by the ILAE Commission on Classification and Terminology [1]. Initially described in 1989 [2], this syndrome affects otherwise healthy children between the ages of 1 and 14 years with 76% of cases starting at 3–6 years of age [3]. Panayiotopoulos syndrome is a common syndrome affecting about 13% of children in the 3- to 6-year age group who present with one or more afebrile seizures [2]. The focal seizures, which mostly occur out of sleep, can be quite prolonged and are commonly characterized by prominent autonomic manifestations [4–7] with approximately 40%–50% of seizures ending with hemiconvulsions or secondary generalization [2,3,8–10]. Interictal EEG frequently shows focal or multifocal functional epileptiform discharges predominating in the posterior head region [3] and typically activated by sleep or only appearing during that state [4–6,11]. The seizures in PS are usually infrequent with a spontaneous resolution within one to two years of onset and an overall excellent prognosis [3,4,8–10]. Because of the benign nature of this syndrome and the infrequent seizures, treatment with an antiepileptic drug (AED) is usually not recommended [5].

We present a young girl diagnosed with PS who experienced respiratory arrest in the setting of an autonomic status epilepticus and required intubation and mechanical ventilation. Furthermore, we evaluate this rare complication of autonomic status, review the few previously reported cases of cardiorespiratory arrest in this syndrome, and discuss the treatment implications.

2. Case report

A previously healthy four-and-a-half-year-old girl with normal psychomotor development, born at term without perinatal complications to a nonconsanguineous couple, experienced her first seizure at the age of 4 years. Semiologically, the seizure was characterized by vomiting, unresponsiveness, right-sided eye deviation, and pallor. On arrival to our emergency department (ED) 15 min later, she was still unresponsive with an oxygen saturation of 89% that picked up to 98% following administration of oxygen by face mask. The seizure was aborted with intravenous diazepam following which the child gradually regained her baseline level of consciousness. Based on the semiology of the seizure and a negative workup including a normal epilepsy protocol brain MRI and a three-hour video-EEG recording, the child was diagnosed with PS. After discussing with and educating the parents about the diagnosis, it was decided not to initiate treatment with an AED.

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Five months later, while asleep in the car, the child woke up feeling sick, had retching, and vomited twice. She was pale, and although she was initially responsive, her parents noted deviation of her eyes to the right followed by a gradual deterioration in her level of consciousness associated with generalized flaccidity. On arrival to our ED, the child was totally unresponsive with a heart rate of 123 beats per minute and an oxygen saturation of 85%. Following suctioning of the airways and administration of oxygen by face mask, the oxygen saturation improved. An emergency head CT scan and electrolyte and blood glucose levels were normal. The patient was then noted to develop bradypnea without bradycardia. Arterial blood gases revealed a severe respiratory acidosis with a pH of 6.9 and a $PaCO_2$ of 122 mm Hg. The child was intubated, attached to a mechanical ventilator, and admitted to the pediatric intensive care unit. She woke up 3 h later at which time her arterial blood gases normalized with a pH of 7.4 and a $PaCO_2$ of 32 mm Hg. The child gradually recovered with no neurologic deficits and was extubated and discharged home the next day on sodium valproate.

3. Discussion

The diagnosis of PS in this child was made based on the characteristic semiology and duration of the seizures, age at seizure onset, normal brain MRI, and normal cognition and development.

The seizure in our patient originated out of sleep, a known characteristic of the majority of seizures in PS [8,10], and was characterized by retching and vomiting, which are the most striking ictal autonomic symptoms in PS, occurring in 74% to 86% of cases [3,7–10]. Subsequently, she became unresponsive and flaccid, an ictal feature of autonomic seizures referred to as ictal syncope and described in 1% to 71% of seizures in PS [3–5,9]. Other autonomic manifestations which may occur concomitantly or later in the course of the seizure include pallor, flushing, cyanosis, mydriasis, miosis, hypersalivation, bladder or bowel incontinence, and thermoregulatory alterations [3,4]. Although typical EEG findings consist of morphologically characteristic sharp waves with a rounded contour topographically involving the posterior head regions, 2–25% of children with PS consistently have a normal EEG [3, 8–10], as was the case in our patient.

The alarming manifestation in our case was the life-threatening bradypnea experienced by the young girl that resulted in a severe respiratory acidosis requiring intubation and ventilator support. Although ictal breathing abnormalities and cardiorespiratory arrest are mentioned under the autonomic seizure manifestations in PS [4], only four such cases were previously reported [4,12–14]. Panayiotopoulos described a child with PS who progressed to cardiopulmonary arrest with dilated unreactive pupils. He required external heart massage and mouth-to-nose resuscitation before his heartbeat picked up 2 min later [4]. Verotti et al. reported on a 6-year-old boy with PS who presented to the ED with autonomic status epilepticus with severe cardiorespiratory manifestations including apnea, absent pulse, and dilated unreactive pupils [12]. The child was resuscitated, intubated, mechanically ventilated, and transferred to the intensive care unit. He recovered 2 h later with no neurologic deficits. Grosso et al. described a 3.8-year-old healthy girl who presented with autonomic status epilepticus that lasted 8 h and was associated with cardiorespiratory arrest that necessitated admission to the intensive care unit [13]. Mujawar et al. reported the case of a previously healthy 3-year-old boy whose first seizure was initially characterized by retching and eye and neck deviation. In the ED, he became deeply cyanosed with absent respiratory movements and absent pulse. He required cardiopulmonary resuscitation for 8 min and two cycles of adrenaline before his cardiac rhythm and respiratory movements resumed. Although a number of diagnoses were entertained to explain his cardiopulmonary arrest, the diagnosis of PS was made after an EEG showed the typical epileptiform discharges [14].

Although no seizure-related death in PS is reported in the literature, it is important to note that the autonomic symptoms might not be recognized as seizure events and that children presenting with autonomic status are frequently misdiagnosed with encephalitis, atypical migraine, cardiogenic syncope, gastroenteritis, or cyclic vomiting syndrome and inappropriately treated [3–6]. While the risk of unexpected, unexplained death in children with idiopathic epilepsy was estimated not to exceed 65 per 100,000 child-years [15], it is likely that our patient and the four others described in the literature would not have survived were it not for the timely medical intervention.

It is difficult to assess the frequency of children with the PS who experience severe cardiorespiratory difficulties as part of their ictal manifestations. Some have estimated that cardiorespiratory arrest occurs in 1 out of 200 patients with PS [4]. Despite their relatively low frequency, such life-threatening events should make us reevaluate the need to treat some children with PS. In the absence of randomized clinical trials or official guidelines regarding the management of PS, we rely on expert opinions that recommend against initiating patients on AED treatment except for children with unusually frequent seizures [5]. For prolonged seizures, some have recommended the use of rectal or buccal benzodiazepines to abort the seizure [5]. While it is difficult to speculate, one can only wonder what would have happened to our patient if she was administered a benzodiazepine close to the time of her bradypnea, since this class of drugs can result in further respiratory depression. This concern was raised by Lacroix et al. [16] who reported five patients who developed severe respiratory depression following benzodiazepine administration for seizures with autonomic manifestations.

While it is unknown if children with PS who experienced a life-threatening event are at higher risk of developing a similar event in subsequent seizures, we decided to initiate our patient on AED treatment. We believe that it is safer to treat such a patient for one to two years rather than to take a chance on having this child experience another life-threatening seizure.

Our case and the previously reported cases establish that although PS is usually a benign syndrome, it is important to realize that some children can experience life-threatening events. For those children, as well as those diagnosed with PS and living in remote areas with no quick access to EDs, it is our opinion that treatment with an AED should be initiated to minimize the chance of experiencing a potentially fatal seizure.

Abbreviations

PS	Panayiotopoulos syndrome
AED	antiepileptic drug
BCSSS	benign childhood seizure susceptibility syndromes
ED	emergency department

Consent

Parental written informed consent was obtained for publication of this case report and any accompanying images.

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Competing interests

The authors have no interests which might be perceived as posing a conflict or bias.

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