

Panayiotopoulos syndrome in a child masquerading as septic shock

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Abstract

Panayiotopoulos syndrome (PS) is a benign childhood epilepsy with predominant autonomic symptoms. The syndrome can have varied presentations resulting in diagnostic dilemma. We herein describe a 3-year-old boy with PS, who had manifestations similar to septic shock. His investigations were normal and had a complete recovery. Through this case, we wish to highlight the unusual presentation of PS as septic shock. Physicians should be aware of the different ways in which this syndrome can present to ensure its early diagnosis and treatment.

Keywords: Autonomic symptoms, febrile convulsion, occipital epilepsy, septic shock

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Introduction

Panayiotopoulos syndrome (PS) is a benign, idiopathic, and probably genetically determined seizure susceptibility syndrome.^[1,2] Although it was initially described in 1989, it was formally recognized in 2001 as a distinct electroclinical syndrome of childhood by the "International League against Epilepsy."^[3] However, more than a decade later, this syndrome still remains an underrecognized entity resulting in diagnostic and therapeutic dilemmas. Several atypical presentations of PS such as respiratory arrest, priapism, and syncopal attacks have been reported.^[4-6] Because of these atypical manifestations, it is not uncommon for PS to be diagnosed late or even remain unrecognized. We herein describe a child with an unusual presentation of PS mimicking as septic shock.

Case Report

A 3-year-old boy was admitted with complaints of cough, cold, fever, and poor oral intake for 2 days. He had two episodes of febrile convulsions in the past 1 year.

The episodes would begin with sudden onset vomiting accompanied by cyanosis and moderate to high-grade fever. After the vomiting, he would develop deviation of eyes to one side lasting for approximately 30 min. This was followed by loss of consciousness of variable duration, ranging from 1 h to 12 h. After the second episode, electroencephalogram (EEG), cerebrospinal fluid studies, and neuroimaging were done which were normal. He was not started on any prophylactic antiepileptic medications, and a diagnosis of atypical febrile convulsions was made. His birth and developmental history was normal. There was no family history of epilepsy. On admission, he was febrile with mild congestion in the throat. Rest of the general and systemic examination was normal. He was started on symptomatic treatment after admission. At 0400 h in the night, he developed high-grade fever and vomiting. This was followed by bluish discoloration of the extremities, tachypnea, and deviation of both eyes to the right side. The episode lasted for about 10–15 min. He

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was unresponsive during the entire episode. He passed a large quantity of loose stools, had the second episode of vomiting, and became drowsy after the episode. His temperature was 39.9°C, oxygen saturation 90%, blood pressure 72/30 mmHg, respiratory rate 39/min, and heart rate 220/min. Fluid resuscitation was done, and high flow oxygen was started. Loading dose of phenytoin (20 mg/kg) was administered. His electrocardiogram showed sinus tachycardia. He was shifted to Intensive Care Unit for observation and monitoring. Within 2 h, he started regaining consciousness and became hemodynamically stable. Within 5 h of the episode, he was fully conscious with a normal neurological examination. After 24 h, he was shifted out to the pediatric ward. Complete blood count, serum electrolytes, blood gases, serum calcium, serum magnesium, and glucose levels were within normal ranges. Blood and urine culture were normal. C-reactive protein was 0.2 mg/L (normal <5). He was discharged after 3 days from the hospital. Phenytoin was continued for 3 days after discharge and stopped. The manifestation during the episode was similar to that seen in septic shock. However, the rapid recovery, normal investigations, and previous history of similar episodes ruled out septic shock. On follow-up after 2 months, he was asymptomatic and had a normal neurological examination. Neuroimaging and EEG were normal.

Discussion

PS occurs in the age group of 1–14 years, with 13% of the cases occurring between 3 and 6 years.^[7] Autonomic epileptic seizures are the cardinal manifestations, with emesis being the predominant symptom. Other autonomic symptoms reported are mydriasis/miosis, pallor, cyanosis, flushing, cardiorespiratory (apnea/changes in heart rate) and thermoregulatory alterations, urinary and/fecal incontinence, hypersalivation, and altered intestinal motility.^[1,4,7] Behavioral disturbances, headache, or other nonpainful cephalic sensations are commonly observed at onset.^[7,8] This is usually followed by more conventional manifestations of seizures with the child becoming confused or unresponsive. Eyes may deviate to one side (60%) or there may be staring look. Half of the seizures last for more than 30 min and can end with hemi or generalized convulsions. Other, less frequent ictal features described include aphasia, hemifacial spasms, visual hallucinations, and oropharyngeal movements.^[1,4,7,8] Cardiorespiratory arrest has also been described in a few cases.^[1,4] Fever is a common trigger for focal autonomic seizures in PS as seen in our case.^[9]

Two-thirds of the seizures occur during sleep or brief daytime naps.^[8] PS is often misdiagnosed if the autonomic

symptoms are not recognized as seizure events. Diagnosis can be confused with other nonepileptic conditions such as atypical migraine, motion sickness, gastroenteritis, encephalitis, or syncope.^[1,7,8] Table 1 shows the various atypical presentations of PS reported in the literature. The autonomic manifestations seen in our case were vomiting, diarrhea, tachycardia, and hypotension. EEG typically shows shifting and/or multiple foci often with occipital predominance, suggesting the possibility that the site of seizure onset is usually occipital.^[1] However, normal EEG recordings may occur in 25% of children as seen in our case.^[8] At least five of the following criteria need to be present to make a diagnosis of PS: Infrequent seizures, prolonged seizures 5 min, ictal vomiting, eye deviation, autonomic manifestations, behavioral and altered consciousness.^[7] Our case fulfilled six of the above criteria.

The pathogenesis of PS involves an epileptogenic activation of the low threshold central autonomic areas. However, the signals are not strong enough to activate the cortical areas that usually bring about motor or sensory manifestations. Hence, most of the seizures have purely autonomic features. However, in some cases, the epileptogenic potential gradually becomes stronger and activates cortical center resulting in secondary generalizations with motor activity.^[1] Prognosis is usually good with remission usually occurring 1–2 years after onset. Around 25% have frequent seizures, but the outcome is still favorable.^[1,7,8] Prolonged seizures do not appear to result in residual deficits or have adverse prognostic significance. Around 20% of cases may progress to rolandic epilepsy which usually remits. The risk of epilepsy in adult life appears to be no higher than in the general population.^[1,7,8] Education and counseling of the parents/caregivers about its benign nature and excellent prognosis are important aspects of management. In case of frequent seizures, rescue therapy with benzodiazepines can be advocated at home/school. Prophylactic therapy with antiepileptic drugs is indicated when seizures are unusually frequent, distressing, or otherwise significantly affecting the child's quality of life. Carbamazepine, sodium valproate, and levetiracetam have been used in various studies with good results.^[1,7,8]

Table 1: Atypical presentations of panayiotopoulos syndrome reported in literature

Case report	Manifestation
Dirani <i>et al.</i> ^[4]	Respiratory arrest
Brabec <i>et al.</i> ^[5]	Priapism
Nishikura <i>et al.</i> ^[10]	Encephalopathy
Koutroumanidis <i>et al.</i> ^[6]	Syncopal attacks
Mujawar <i>et al.</i> ^[1]	Cardiorespiratory arrest
Ozkar <i>et al.</i> ^[11]	Intractable vomiting

Conclusion

We highlight this unusual presentation of PS as septic shock. Physicians should be aware of the varied presentations of this syndrome to ensure its early diagnosis and proper management.

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Conflicts of interest

There are no conflicts of interest.

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