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Case Report

Cardiac failure in a child with tuberculous meningitis as a complication of Paroxysmal sympathetic hyperactivity

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ABSTRACT

Background. Paroxysmal sympathetic hyperactivity (PSH) is a disorder due to the loss of regulation of autonomic activity. The most common condition predisposing to the development of PSH is traumatic brain injury (TBI), followed by anoxic brain injury, stroke, tumors, and infections. Awareness about the condition and early recognition is important to avoid life threatening complications.

Case. We report a 4-year-old child with tuberculous meningitis with symptoms of PSH who developed cardiac failure. PSH episodes were treated with beta blocker, benzodiazepine, morphine, dexmedetomidine, baclofen, and tizanidine. Three weeks after readmission PSH episodes decreased and the patient was transferred to the general ward.

Conclusions. PSH assessment tool has benefits such as monitoring the patient, evaluating response to treatment and early diagnosing PSH patients.

Key words: paroxysmal sympathetic hyperactivity, tuberculous meningitis, child.

Paroxysmal sympathetic hyperactivity (PSH) is a disorder due to the loss of regulation of autonomic activity. PSH has previously been described as autonomic storms, hypothalamic dysregulation syndrome, dysautonomia, paroxysmal autonomic instability with dystonia, and diencephalic autonomic epilepsy. The most common condition predisposing to the development of PSH is traumatic brain injury (TBI), followed by anoxic brain injury, stroke, tumors, and infections. In 2014, an international panel was convened and published a consensus statement to define symptoms and diagnostic criteria of PSH.

Hereby we report a 4-year-old child with tuberculous meningitis with symptoms of PSH who developed cardiac failure.

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Case Report

A four-year old boy was admitted with fever, headache for three days, intermittent vomiting, and loss of consciousness. On his examination, neck stiffness has been determined, Glasgow coma scale (GCS) was 11. Vital signs were within the normal range. The diagnosis of tuberculous meningitis was made after head magnetic resonance imaging (MRI) and cerebrospinal fluid examination. Therapy of isoniazid, pyrazinamide, rifampin, and ethambutol has been initiated. On the 2nd day, he had a seizure, his computed tomography showed hydrocephalus. He was referred to our hospital after a ventriculoperitoneal (V/P) shunt placement and because of the need for tertiary intensive care follow up.

On admission to the pediatric intensive care unit (PICU), his heart rate was 110 beats per minute (bpm), blood pressure 105/60 mmHg, temperature 37.8 °C, GCS was 8. Physical examination revealed no abnormalities except

increased muscle tone. On the 2nd day in the PICU the patient deteriorated with a GCS of 5. Cerebral MRI showed progression of hydrocephalus and angio-MRI showed left anterior carotid artery (segment A_2) and posterior carotid artery (segment P_1, P_2) occlusion with embolic infarcts in the left cerebral hemisphere.

Tracheostomy was placed for long term ventilation and the patient has transferred to the general ward in a stable condition with home mechanical ventilatory support on the 24th day of admission. Cerebrospinal fluid culture detected *M.tuberculosis* with multiple drug resistance. His treatment was changed to linesolide, moxifloxacin, streptomycin, and ethambutol. Baclofen and tizanidine were continued for dystonia.

After 2 months of transfer to the general ward, he was readmitted to the PICU with symptoms of congestive heart failure. The heart rate was 190 bpm, blood pressure 190/110 mmHg, temperature 38 °C, and oxygen saturation 85% with fractional inspired oxygen of 50%. On his physical examination, his liver was palpable 4 cm under the right costal margin. On neurological examination, he had extensor posturing, mydriasis, and GCS of 4 points. Intravenous antihypertensive therapy (esmolol and glyceryl trinitrate) was immediately initiated. Transthoracic echocardiography revealed decreased systolic ventricular function with an ejection fraction (EF) of 30%.

On the 2nd day of readmission, antihypertensive treatment was weaned off gradually. During his observation in the PICU, he developed intermittent bradycardia and tachycardia episodes with high blood pressure, piloerection, mydriasis, dystonia, and fever. These episodes lasted approximately 30 minutes and improved after benzodiazepines. The electroencephalogram showed no epileptic discharge. During his stay in the PICU, sudden hypotension and hypertension periods with dystonia were observed. Serum cortisol level was within the normal range. Cardiac MRI revealed no abnormalities. Cultures of blood, urine, and tracheal aspirate were negative. A subsequent MRI showed progression of tuberculomas in the bilateral cerebral hemisphere and basal cisterns, and infarction in the basal ganglia.

The patient was considered as probable PSH according to the PSH assessment measure (PSH-AM). PSH episodes were treated with beta blocker, benzodiazepine, morphine, dexmedetomidine, baclofen, and tizanidine. Three weeks after readmission PSH episodes decreased and the patient was transferred to the general ward. Transthoracic echocardiography revealed improved systolic ventricular function with EF of 50%. Written informed consent was obtained from the parents for publication of the case.

Discussion

PSH is a disorder in the regulation of the sympathetic nervous systems which is mostly caused by severe traumatic brain injury (TBI). In 2014, an international panel was convened and published a consensus statement to define symptoms and diagnostic criteria.[1] The reported incidence of PSH ranges from 8% to 33% in adults and %13 in children following acquired brain injury.[2,3] In adult studies, most PSH cases have been reported to occur after TBI. Anoxic brain injury, stroke, tumor, infections, subarachnoid hemorrhage, hydrocephalus, and suprasellar cyst have been reported as causes of PSH.[4-6]

PSH shows a wide spectrum of clinical symptoms. Main clinical symptoms include tachycardia, hypertension, tachypnea, fever, sweating, and/or increased muscle tone with possible dystonic posturing. Patients may present with various combinations of these symptoms. These clinical features can manifest spontaneously or in response to slight-noxious stimuli such as aspiration of secretions, change of position, or physiotherapy. In a previous study, tachycardia has been reported as the

most common symptom.[7] The combination of hypertension, diaphoresis, and dystonia best predicted a diagnosis of pediatric PSH.[3] In acute settings patient treatment generally includes deep sedation and analgesia which may hide the symptoms of PSH. It is important to recognize PSH because, if untreated, PSH can persist and potentially result in serious complications such as dehydration, muscle loss, and contractures.

Clinical Features Scale and Diagnosis Likelihood Score has been determined when dominant symptoms are observed, and appropriate interventions were made according to the clinical situation. Kirk et al.[3] diagnosed 10% PSH in TBI in pediatric rehabilitation settings by clinical diagnosis of dysautonomia. However, recently Alofisan et al.[8] showed a prevalence of 20% PSH in severe TBI children by using the diagnostic tool. The tool has benefits of monitoring the patient, evaluating response to treatment and diagnosing PSH patients early. In this case, the PSH-AM score was 22 and the score dropped to 9 before the patient was transfer to the general ward.

In the hyperacute phase, most brain injury patients require deep sedation and analgesia during their follow up in intensive care units which makes it difficult to recognize PSH. In this case, the patient underwent invasive procedures and needed a longer duration of sedoanalgesia. During his stay in the general ward, noninvasive monitorization resulted in underdiagnosed hypertension-hypotension periods and medical treatment was continued with baclofen and tizanidine only. The patient was admitted to the intensive care unit with a life-threatening complication due to a delay in diagnosis.

Recent studies reported that older age in children and early tracheostomy was associated with an increased risk for developing PSH.[8,9] It is important to develop tools to define risk factors that may help recognize PSH patients.

Author contribution

The authors confirm contribution to the paper as follows: study conception and design: PYÖ, BK; data collection: SK; analysis and interpretation of results: PYÖ, EET, HFA; draft manuscript preparation: PYÖ, BK. All authors reviewed the results and approved the final version of the manuscript.

Conflict of interest

The authors declare that there is no conflict of interest.

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