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CASE REPORT

Paroxysmal sympathetic hyperactivity syndrome after recurrent stroke: A case report

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ABSTRACT

Paroxysmal sympathetic hyperactivity (PSH) is a state of autonomic dysfunction characterized by symptoms such as tachypnea, tachycardia, hypertension, hyperthermia, sweating, and dystonia. It can occur after traumatic brain injury, hypoxic-ischemic encephalopathy, and diseases such as stroke. Hypoxia, extensive axonal damage, and young age are believed to predispose to the development of PSH. These patients may be diagnosed with pulmonary embolism, septicemia, or epileptic seizures. Delays in diagnosis prolong hospital stay. Here, we present an 81-year-old man who developed PSH after a recurrent stroke and our management.

Keywords: Paroxysmal sympathetic hyperactivity, Ischemic stroke, Autonomic dysfunction

INTRODUCTION

Paroxysmal sympathetic hyperactivity (PSH) is an autonomic dysfunction characterized by tachypnea, tachycardia, hypertension, hyperthermia, sweating, and motor features such as dystonia (1). PSH may occur after brain damage such as traumatic brain injury (79%), hypoxic-ischemic encephalopathy (10%), and stroke (2,3). Hypoxia, extensive axonal damage, and young age are believed to predispose to the development of PSH (2). The exact pathophysiology of PSH is unknown (3). According to previous studies, PSH has not been associated with increased mortality or poor prognosis (3). Diagnosis can be delayed due to its rarity and confusion with some other diseases. This delay results in patients receiving incorrect treatments and prolongs their hospital stay. Here we present an elderly patient who developed PSH after a recurrent ischemic stroke.

CASE REPORT

An 81-year-old male patient presented to the emergency department with complaints of speech disorder the day before. He had a history of Parkinson's disease and epilepsy.

He had been bedridden due to a stroke 19 years ago. He was regularly using acetylsalicylic acid 100 milligrams once a day and phenytoin 100 milligrams (mg) 3 times

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a day. According to the neurological examination, the patient's consciousness was inclined to sleep. The cranial nerve examination was normal. He had newly developed motor aphasia and right sequela hemiparesis. Diffusion magnetic resonance imaging revealed limited diffusion consistent with an ischemic infarction in the left insular cortex (Figure 1). The patient was admitted to the neurology service with the diagnosis of acute ischemic infarction. Antithrombotic therapy was started. During the follow-up of the patient, saccadic eye movements and construction in the whole body appeared. He also exhibited high blood pressure, tachycardia, tachypnea, and flushing. Diazepam was administered to the patient considering epileptic seizure. Then, levetiracetam was added to his treatment. The patient responded to the treatment. Meanwhile, no abnormality was observed in the EEG taken twice.

In the follow-up radiological examinations, no pathological finding that could be related to the clinical findings was found.

After being transferred to the intensive care unit, the patient was intubated, and a barbiturate (pentobarbital) infusion was administered for 72 hours.

During the follow-up, the sympathetic episodes recurred after 72 hours. Benzodiazepine (midazolam) infusion was given followed by dexmedetomidine. When the treatment was discontinued, the attacks recurred. We considered PSH because there was no response to epilepsy treatment, typical epileptic attacks were not observed, and accompanying sympathetic discharge findings were present. We diagnosed PSH 7 days after ICU admission.

The PSH test, proposed by Baguley et al. (1), was used for the diagnosis, and the PSH index score was 15 points in our patient (Table 1). We considered a possible diagnosis of PSH according to these criteria.

The patient was managed using beta-blockers propranolol (80 mg daily), trazodone (100 mg daily), and gabapentin (1800 mg daily); so, the frequency and

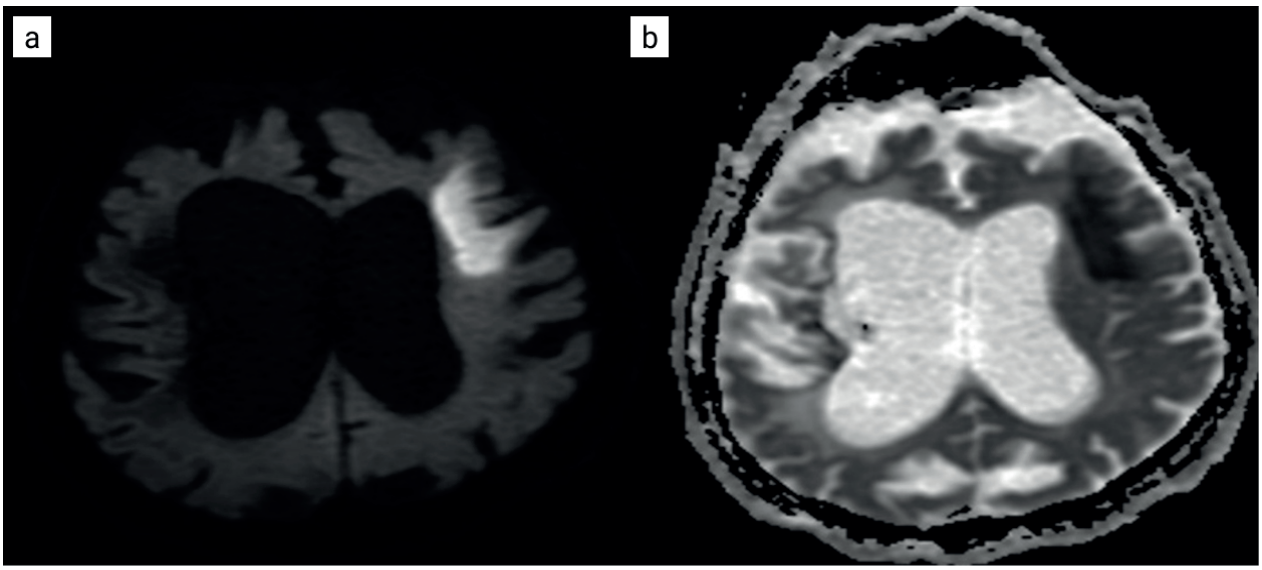

Figure 1. The DWI scan shows a left-sided hyperintensity in the frontal lobe (a), which can be appreciated as a hypointensity on the ADC trace map (b).

Table 1. The patient's baguley score measured during paroxysmal sympathetic hyperactivity

	0	1	2	3	Score
Heart rate (BPM)	<100	100 to 119	120 to 139	≥140	3
Respiratory rate (breaths/minute)	<18	18 to 23	24 to 29	≥30	3
Systolic blood pressure (mmHg)	<140	140 to 159	160 to 179	≥180	2
Temperature (°C)	<37	37 to 37.9	38 to 38.9	≥39	1
Sweating	Nil (none)	Mild (moist skin)	Moderate (beads of sweat)	Severe (profuse generalized sweating)	1
Posturing during episodes	Nil (none)	Mild (increased tone but not requiring treatment)	Moderate (increased tone requiring treatment)	Severe (very increased tone refractory to treatment)	0
				CFS subtotal =	10
Likelihood Tool (DLT)					
Score as "1" if present					**Score**
Clinical features occur simultaneously					
Episodes are paroxysmal in nature					1
Over-reactivity to normally nonpainful stimuli					
Features persist ≥3 consecutive days					1
Features persist ≥2 weeks post-brain injury					
Features persist despite treatment of differential diagnoses					1
Medication administered to decrease sympathetic features					1
≥2 episodes daily					1
Absence of parasympathetic features during episodes					
Absence of other presumed cause of features					
Antecedent acquired brain injury					
				DLT subtotal =	5
				Combined total (CFS + DLT) =	15

severity of attacks decreased. Episodes completely disappeared after a while. The patient later died due to metabolic disorders and respiratory failure.

DISCUSSION

Paroxysmal sympathetic hyperactivity (PSH) is an autonomic dysfunction characterized by tachypnea, tachycardia, hypertension, hyperthermia, sweating, and motor features such as dystonia. PSH is more commonly observed in young patients (2).

Paroxysmal sympathetic hyperactivity may occur after brain damage such as traumatic brain injury (79%), hypoxic-ischemic encephalopathy (10%), and stroke (3).

The incidence of PSH is particularly high in patients with axonal damage and deep brain damage (4).

Hydrocephalus (2.6%), brain tumors, central nervous system infections, and hypoglycemia are rare causes of PSH (3). Previous studies have shown that PSH does not raise morbidity or worse clinical outcomes (3).

Septicemia, epilepsy, pulmonary embolism, and some serious diseases may have symptoms similar to PSH. In the presence of these diseases, PSH may not be considered in the foreground.

Such as, while hyperthermia primarily suggests septicemia, pulmonary embolism is suspected if hyperthermia accompanies tachypnea. Dystonic posture may suggest an epileptic seizure diagnosis (5). Our patient was elderly and had many comorbid conditions. We thought the epileptic seizure was due to the patient's posture. We considered PSH because of its resistance to antiepileptics and accompanying autonomic findings.

The exact pathophysiology of PSH is unknown. Different theories have been put forward. The first is the disconnection theory, which is explained by the damage to the connection between the sympathetic nerves, centered in the hypothalamus and brainstem, and the higher cerebral cortex (3,6).

Another theory postulates an imbalance between excitation and inhibition, causing mismatched dendritic arborization and stimulation of spinal cord circuits (7).

Additionally, the insula may play a role in the hyperactivity of the sympathetic nervous system (8).

Bilateral insula, supramarginal gyrus, and amygdala are associated with the sympathetic nervous system (9). Our patient had an insular infarction. Sympathetic nerve hyperexcitation leads to PSH by increasing catecholamine release.

Studies have shown that sympathetic hyperactivity in PSH causes an increase in catecholamine release, with elevated levels of adrenocorticotropin, epinephrine, norepinephrine, and dopamine during attacks (10).

The goal of treatments is to reduce excessive sympathetic nerve activity. There is no definitive protocol for PSH treatment management. The treatment aims to distinguish which symptoms are urgent and the need for priority (11). Many combinations of drugs acting through different pathways can be used in treatment. Since the pathology of the disease is not clear, symptom control comes to the fore in treatment (12,13). It has been reported that opioids (morphine and fentanyl), intravenous anesthetics (propofol), neuromodulators (gabapentin, bromocriptine, baclofen, benzodiazepines (diazepam, midazolam, Lorazepam and clonazepam), alfa-2 agonists (clonidine, dexmedetomidine), peripherally acting muscle relaxants (dantrolene) and beta-adrenergic blockers (propranolol, labetalol, metoprolol) can be used (7). The ideal treatment is to use short-acting drugs, choose the appropriate regimen and avoid uncontrolled drugs.

In addition, it is recommended to treat hyperthermia with antipyretics, agitation with sedatives, and hypertension with antihypertensive agents (12,13). In our patient, epilepsy and cardiac-pulmonary symptoms were primarily considered. On the other hand, autonomic findings at follow-up and inadequate response to primary treatment led us to the diagnosis of PSH. The disease could then be brought under control with treatments performed on this axis.

CONCLUSIONS

Paroxysmal sympathetic hyperactivity is rare in patients with stroke. Typical symptoms can occur in many other diseases, so it is possible to experience delays in diagnosis. The treatment is possible with drugs affecting different mechanisms. Early diagnosis of PSH can shorten the hospital stay and prevent wrong treatments. In this case report, we wanted to draw attention to this rare disease and raise awareness.

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Ethical approval

Written informed consent was obtained from the participants.

Author contribution

Concept: FB, SY; Design: FB, ŞAT; Data Collection or Processing: FB, SE; Analysis or Interpretation: SY; Literature Search: FB, ŞAT, SE; Writing: FB, SE, ŞAT. All authors reviewed the results and approved the final version of the article.

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Conflict of interest

The authors declare that there is no conflict of interest.

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