Paroxysmal Sympathetic Hyperactivity (PSH) is commonly described in patients after Traumatic Brain Injury but it can present after an ischemic stroke following a complicated surgery. Usual clinical presentation are tachycardia, tachypnea, hypertension, sweating with seizures and dystonic posturing less likely initial signs. High grade fever and profuse sweating may allude sepsis or epilepsy leading to extraneous administration of anti-epileptics and antibiotics. Suspicion of pheochromocytoma arises in such situations which is ruled out by CECT. Clonidine (α-2 blocker), propranolol (β-blocker) and Baclofen (GABAb agonist) are agents used in treatment. Benzodiazepines like lorazepam are important part of treatment as discontinuing them can exacerbate PSH attacks. MRI brain with MR angiography usually reveals hyper-intensities on T2/FLAIR in subcortical areas (thalami, cerebellum, crus cerebri) and restricted diffusion with low ADC values. Recurrent PSH episodes can occur due to delay in diagnosing and treatment leading to contractures and difficulty weaning the patient off the ventilator with tracheostomy tube placed. This case presents a scenario in which the delay in diagnosing PSH led to extensive investigations, delay in specific treatment and a prolonged ICU stay of the patient. Mainstay of rehabilitation remains aggressive physiotherapy to improve contractures if any, medications for the autonomic fluctuations and regular follow up.

Introduction

Paroxysmal sympathetic hyperactivity (PSH) is a disorder of autonomic function regulation most commonly observed in patients with acute brain injury. It mostly occurs after traumatic brain injury, but it can also occur after non-traumatic brain diseases such as anoxic-ischemic coma after cardiac arrest, intracranial haemorrhage, and ischemic stroke-[1].

The core clinical features include - tachycardia, tachypnea, hypertension, sweating, hyperthermia and posturing-[2]. These episodes are mostly triggered by some external stimuli such as pain , movement , and urinary retention . PSH occurs due to diffuse or focal brain injuries that disconnect one or more cerebral centers from the caudal excitatory centers and the disconnection of descending inhibitory pathways causing spinal circuit excitation.

Tonic posturing during episodes can mimic tonic seizures, and the raised temperature can mimic infection, which can lead to unnecessary investigations, delays in proper management and prolonged ICU stays. Here, we are presenting one such case.

Case Presentation

A 32-year-old woman (gravida 2 and para1) had pain in the lower abdomen at week 31 of gestation for which she was hospitalized and emergency Lower Segment Cesarean Section was performed in view of fetal hypoxia. On the 2nd day post-delivery, she developed multiple episodes of seizures without regaining consciousness in between and was intubated for airway protection. Brain Magnetic Resonance Imaging(MRI) with angiography suggested posterior
circulation ischaemic stroke with bilateral narrow caliber of both vertebral arteries and a left fetal PCA. She was started on antiepileptic drugs and secondary prophylaxis for stroke. She was having high-grade fever and, multiple episodes of profuse sweating per day. Blood counts, cultures, and other markers of infection gave negative Results. She had episodes of tachycardia, tachypnea, hypertension, and sweating associated with fever.

The fever episodes created difficulty in weaning the patient from the ventilator. Therefore, she was started on clonidine and later on propranolol; however, these episodes continued to occur. Suspecting pheochromocytoma, abdominal contrast enhanced computed tomography (CECT) was performed, but the results were normal. The patient had recurrent episodes of increase in whole body tone (Fig 1) associated with bilateral lower limb tremors, abnormal posturing along with the above-mentioned episodes of fever. Initially, it was assumed that the rigors were those associated with fever, but later, speculating that the rigors were seizures, the dosage of her antiepileptic medications were increased, and she was administered triple antiepileptics (valproic acid, levetiracetam, and lorazepam). Her brain EEG showed no epileptiform discharges. Despite receiving triple antiepileptics, she continued to show seizure-like activity. Later, these episodes of fever, tachycardia, tachypnea, hypertension, sweating and increase in body tone occurred simultaneously, and the diagnosis of paroxysmal sympathetic hyperactivity was made.

The patient was administered clonidine, propranolol, and baclofen. Her episodes of sympathetic hyperactivity were controlled. Later, her empirical antibiotics were stopped as there was no evidence of infection and fever was explained as a part of sympathetic hyperactivity.

Antiepileptic medications were tapered off to only valproic acid. However, after discontinuing valproate and lorazepam she again started experiencing episodes of sympathetic hyperactivity. Therefore, lorazepam was reintroduced to control her symptoms. Finally, the patient was weaned off ventilator support and shifted to the ward.

**Investigations**

- Blood parameters: TLC - 10,000 /mm3, Hb - 12 g/dl, Platelets - 2.8 lac/mm3
- Blood culture: no pathogenic organism grown after 48 hrs of aerobic incubation.
- Urine culture: sterile
- Endotracheal aspirate culture: no pathogenic organism grown after 48 hrs of aerobic incubation.
- High vaginal swab culture: normal vaginal flora grown.
- Procalcitonin: 0.29 ng/ml
- CSF analysis: Acellular; sugar: 24 mg/dl; protein: 68 mg/dl; culture: sterile
- Dengue IGM: negative; ICT and peripheral smear for Malaria: negative, Typhidot IGM: negative; scrub typhus IGM: negative.
- Ultrasound abdomen: no significant abnormality.

**MRI Brain with MR Angiography**

The brain stem appeared bulky and showed T2/FLAIR hyperintense signal. Similar areas with T2/FLAIR hyperintense signal were also seen involving the thalami, right crus cerebri and bilateral inferior cerebellar peduncles. Many of these areas showed restricted diffusion with low Apparent Diffusion Coefficient (ADC) values. The bilateral (right > left) vertebral and distal basilar arteries appeared significantly attenuated in caliber. The right PCA appeared to be significantly attenuated in calibers compared with fetal origin of the left PCA.

- EEG: Generalized cerebral dysfunction; no epileptiform discharges.
- CECT abdomen: No significant abnormality.

**Outcome and Follow-up**

The episodes of sympathetic hyperactivity resolved, and the patient was shifted to the ward from the ICU with tracheostomy in room air. She developed contractures owing to persistent decerebrate posturing. Aggressive physiotherapy was administered. In the ward she regained her sensorium and was discharged with a GCS score of E4VTM6 for further follow-up in the OPD.

**Discussion**

A significant minority of patients who survive acquired
brain injury develop sympathetic hyperactivity, which includes episodes of periodic increase in heart rate and blood pressure, sweating, hyperthermia, and motor posturing, often in response to external stimuli, which can last for weeks or more months-[3]. Some studies argue that it is common but often unrecognized-[4]. Most studies have reported paroxysmal sympathetic hyperactivity after traumatic brain injury. Fewer cases have reported it to be a sequel of brain stroke following prolonged hospital stay. Though commonly seen in TBI, PSH has rarely been described in patients with brainstem strokes and anoxic brain injury-[5]. PSH may be mistaken for sepsis, which may lead to unnecessary treatment with antibiotics and prolonged hospital stays-[6]. Fever is relatively common among patients in the intensive care settings. Although the most obvious and concerning etiology is sepsis, PSH may be the underlying etiology-[7]. There are diencephalic structures analogous to the cerebral motor cortex that are capable of producing, when irritated, paroxysmal motor discharges similar to the focal discharges described as epilepsy-[8]. PSA may be camouflaged by epileptic seizures, leading to the unwarranted administration of antiepileptics to the patient. Obstetric patients can present with acute increases in heart rate, BP and the onset of HELLP syndrome mimicking PSH. Epigastralgia, hypertension, and tachycardia necessitate cesarean section, as in our case, with the subsequent development of HELLP syndrome mimicking PSH. An acute fluid shift from the splanchnic vasculature to the central vasculature may have occurred, causing HELLP syndrome as a result of vasospasm associated with sympathetic hyperactivity. Reporting such cases will facilitate in understanding if the reverse is true, that is, if PSH can mimic as HELLP syndrome-[9,10]. Pregnancy is a risk factor for paroxysmal sympathetic hyperactivity exacerbation, and delivery can result in resolution of the condition-[11]. PSH is reclassified as a sympathetic storm rather than an epileptic disorder because of its unresponsiveness to anti-epileptics and the absence of epileptic activity on EEG. [12] It is crucial for clinicians to distinguish this disorder from paroxysmal dystonias. [13,14] Sympathetic storms have been linked to dystonia-like posturing (e.g., PAID, i.e., "paroxysmal autonomic instability with dystonia") [15]. Antidopaminergic medications are best avoided to minimize the risk of neuroleptic malignant syndrome, which can potentially mimic PSH (dysautonomia). In contrast to delirium-associated persistent agitation and picking-like behaviours, PSH movements are episodic, tend to be provoked by touch, and are uniquely associated with increased sympathetic activity. In the management of this disorder opiates, γ-aminobutyric acid agents, dopaminergic agents, and β-blocker pharmacological agents have been studied. There is a lack of recommendations and comparisons of agents for the management of this disorder.

There is a paucity of recommendations and comparisons of agents for the management of this disorder. Monotherapy is usually ineffective for the management of paroxysmal sympathetic hyperactivity, and multiple agents with different mechanisms of action should be considered, as in our case, β-blockers have proven to be therapeutic (not as monotherapy) as in our case, α-agonists such as dexmedetomidine have reported therapeutic efficacy in many studies. However, clonidine another α-agonist has shown therapeutic efficacy, as in our case. The effectiveness of physiotherapy in PSH is rarely reported in medical journals; our case strives to provide an example of such therapeutic benefits.

**Conclusion**

Paroxysmal sympathetic hyperactivity is quite common in patients with brain insult. It can mimic seizures and/or sepsis (due to high grade fever) leading to unnecessary investigations, exposure to higher-grade antibiotics and antiepileptics. Benzodiazepines are beneficial in controlling sympathetic hyperactivity. Non-recognition of PSH can lead to difficulty in weaning patients off the ventilator and prolonged hospital stays. Recurrent episodes of sympathetic hyperactivity can lead to significant weight loss and contractures.

**References**

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