Acute visual loss in a patient with spinal cord injury

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Abstract

Introduction Patients with spinal cord injury (SCI), especially those with injury at and above T6, are prone to transient episodes of hypertension induced by noxious triggers below the level of SCI, known as autonomic dysreflexia (AD). An uncommonly reported presentation of AD is posterior reversible encephalopathy syndrome (PRES).

Case Presentation A 50-year-old male with the history of paraplegia from SCI presented with sepsis secondary to baclofen pump and urinary tract infections. On hospital day 4, he developed acute bilateral vision loss. The next morning he had a generalized-tonic-clonic seizure followed by cardiac arrest, with return of spontaneous circulation following resuscitation. Magnetic resonance imaging brain demonstrated multifocal areas of hyperintensity on T2 fluid-attenuated inversion recovery sequence, most pronounced in the occipital lobes. Systolic blood pressures (SBP) were under 180 mmHg throughout hospital stay but above his baseline (SBP 90 mmHg). PRES was diagnosed on the basis of clinical and radiologic evidence. With strict blood pressure (BP) control, there was resolution of visual abnormalities, headaches, encephalopathy, and seizures.

Discussion Although PRES has been most commonly described in malignant hypertension, it can be seen in patients with normotension or moderate hypertension who have low baseline BPs, such as patients with SCI. These patients are prone to AD due to imbalanced sympathetic outflow to vasculature below the level of injury caused by noxious stimulus. This results in massive regional vasoconstriction leading to an uncontrolled rise in BP above baseline. This episode of PRES could have been prevented by identifying patient's risk, recognizing early signs and potential triggers of AD, and implementing aggressive treatment of the underlying noxious stimuli.

Introduction

Patients with spinal cord injury (SCI), especially those with injury at and above T6, are prone to transient episodes of hypertension induced by noxious triggers below the level of SCI, known as autonomic dysreflexia (AD). Elevations in blood pressure (BP) of 20–40 mmHg above baseline often can have devastating neurologic and systemic manifestations in such patients. The common triggers for AD may include urinary catheter obstruction, constipation, pressure ulcer, and infection [1–4]. An uncommonly reported presentation of AD is posterior reversible encephalopathy

syndrome (PRES), a clinicroadiologic entity characterized by headache, encephalopathy, visual disturbances, seizures, and radiologic evidence of vasogenic edema predominantly localized in posterior cerebral hemispheres. The most common trigger for PRES is hypertensive crisis [2, 4–7].

Case presentation

A 50-year-old male with the history of Parkinson's disease, complete T3 paraplegia secondary to SCI (ASIA grade A (treated with intrathecal baclofen/morphine pump)), neurogenic bladder (suprapubic catheter in place), and neurogenic bowel who presented with sepsis secondary to baclofen pump infection and urinary tract infection (UTI). Blood cultures were positive for methicillin-sensitive *Staphylococcus aureus* and *Candida albicans*. Urine cultures were positive for *Pseudomonas aeruginosa* and *Candida parapsilosis*. The patient had been endorsing pounding occipital headaches for a few days prior to hospital admission, which worsened while the inpatient was refractory to standard

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Fig. 1 MRI brain showing multifocal areas of cortical and subcortical hyperintensity on T2-FLAIR (A), most pronounced in the posterior cerebral hemispheres and without corresponding restricted diffusion on diffusion-weighted imaging (B)

analgesics. He was taken to the operating room for removal of infected baclofen pump but he became encephalopathic post-op, which was attributed to delirium secondary to infection. On fourth day of hospital admission, he developed acute onset bilateral vision loss. A head computed tomography (CT) scan demonstrated hypoattenuation of the left and possibly also right occipital lobe with sulcal effacement that was concerning for stroke. Ophthalmology was consulted but no abnormalities were identified on Slit Lamp and Dilated Fundus Exam. The next morning he had a generalized tonic clonic seizure followed by cardiac arrest, requiring two rounds of CPR with return of spontaneous circulation. He was then intubated and transferred to the intensive care unit.

Neurology was consulted for findings on head CT. A CT angiogram was unremarkable. Magnetic resonance imaging (MRI) brain showed multifocal areas of cortical and subcortical hyperintensity on T2-fluid-attenuated inversion recovery sequence (FLAIR) sequence, most pronounced in the occipital lobes (Fig. 1A, B). Electrolytes were within normal limit. Patient was not on any immunosuppressive agents. Patient was connected to telemetry throughout the hospital stay with frequent monitoring of BPs. Patient's systolic blood pressures (SBP) had been under 180 mmHg throughout hospital stay but were above his baseline (around 90 mmHg). PRES was diagnosed on the basis of clinical and radiologic evidence. With strict BP control, there was resolution of visual abnormalities, headaches, encephalopathy, and seizures. After 10 days on the inpatient SCI rehabilitation unit, the patient was able to return home with only mild cognitive deficits.

Discussion

Patients with SCI, especially those with a cervical or high-thoracic injury (at and above T6), are vulnerable to lifelong abnormalities in systemic arterial pressure control and orthostatic intolerance. Their baseline BPs are generally lower than that of normal population. These patients are prone to transient episodes of hypertension induced by noxious and also non-noxious triggers below the level of SCI, known as AD. AD occurs more commonly in patients with SCI at or above T6 due to imbalanced sympathetic outflow to the vasculature below this level (especially the pelvic splanchnic vessels) caused by the noxious stimulus. This results in massive regional vasoconstriction leading to an uncontrolled rise in BP. The brainstem signaling to decrease the sympathetic outflow and allow vasodilation of the splanchnic beds to lower the BP is blocked by the SCI. Sustained elevations in BP of 20–40 mmHg above baseline often can have devastating neurologic and systemic manifestations, including intracerebral hemorrhaging, myocardial ischemia, and even sudden death [1–4].

The common triggers for AD may include urinary catheter obstruction, bladder over distension, UTI, severe constipation, routine procedures such as a bowel program and sepsis. The most common symptoms of AD include severe bilateral pounding headaches, shortness of breath, palpitations, anxiety [1, 2]. Common signs may include hypertension, bradycardia, tachycardia, and goose flesh.

An uncommonly reported presentation of AD is PRES, which refers to a clinicoradiologic entity characterized by headache, encephalopathy, visual disturbances (ranging from blurred vision to cortical blindness), generalized

seizures, and radiologic evidence of vasogenic edema predominantly localized in posterior cerebral hemispheres [2, 4–7]. MRI is the most sensitive imaging modality to detect vasogenic edema (T2-FLAIR sequence) and also to distinguish it from cytotoxic edema secondary to stroke (diffusion-weighted imaging (DWI)) [5, 6, 8]. The most common trigger for PRES is hypertensive crisis [4, 6, 7]. Although PRES has been most commonly described in malignant hypertension (SBP > 180), it can be seen in patients with normotension or mild to moderate hypertension (SBP < 180) who have low BPs at baseline [4, 8], such as patients with SCI (above T6).

Our patient's systolic BPs at baseline were around 90 mmHg. An open abdominal wound and infected intrathecal pump, subsequent bed rest, UTI, and sepsis were all possible contributions that lead to AD with sudden BP elevations to 140–160 mmHg. He demonstrated the early manifestations of AD by refractory pounding headaches in the setting of elevated BP above baseline, but when unrecognized, this lead to the classic clinical picture of PRES complicated by an episode of cardiac arrest. This was a diagnostic challenge for the medicine and neurology team, because although we were able to diagnose PRES (on the basis of clinical and radiologic evidence), we were initially not able to identify any trigger as the patient's BPs remained below 180 mmHg. It was the PM&R SCI Service, whom identified AD as the most likely etiology for this patient's episode of transient hypertension and educated the family and primary care teams on ways to recognize and prevent this in the future.

We believe that this episode of PRES could have been prevented by identifying his risk (T3 paraplegia), early signs and potential triggers of AD, knowing his baseline SBP and implementing aggressive treatment of the underlying noxious stimuli, and prompt BP control back to his baseline.

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Compliance with ethical standards

Conflict of interest The authors declare that they have no competing interests.

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