Case Report

Posterior Reversible Encephalopathy Syndrome Presenting With Obstructive Hydrocephalus:

A Rare Presentation

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osterior reversible encephalopathy syndrome (PRES) is an acute clinical condition, often manifesting as a spectrum of findings including headache, encephalopathy, visual disturbances, and seizures. The disorder is believed to result from failure of cerebral autoregulation and endothelial dysfunction, resulting in vasogenic cerebral edema.1 As the name implies, the condition mostly involves the occipital and parietal regions. While infratentorial involvement can occur,2 PRES resulting in acute obstructive hydrocephalus is an exceedingly rare finding. Here, we present a case of PRES due to substance-induced hypertensive crisis resulting in obstructive hydrocephalus.

Case Report

A 28-year-old white man with a history of severe hypertension, hyperthyroidism, and cocaine and methamphetamine use presented to the emergency department (ED) with severe headache, nausea, dizziness, and bright spots in his left eye for 1 day. The patient reported using methamphetamine at the time of symptom onset. At that time, he measured his blood pressure and noted his systolic blood pressure was 279 mm Hg. Upon arrival to the ED, his initial blood pressure was noted to be 222/159 mm Hg. His neurological examination was noted to be unremarkable. A computed tomography (CT) scan showed diffuse edema in the cerebellum and brain

stem resulting in significant mass effect on the fourth ventricle and obstructive hydrocephalus (Figure 1). The patient was transferred to our facility with a suspicion of an infratentorial mass. Upon arrival, he was admitted to the neurocritical unit for further management. Magnetic resonance imaging with and without contrast showed vasogenic edema, most pronounced in the bilateral cerebellar hemispheres but also present in the supratentorial compartment. Periventricular transependymal edema was also noted. There was no evidence of any space-occupying lesion. His magnetic resonance angiogram was unremarkable. Based on a careful review of history and imaging studies, a diagnosis of PRES was established.

He was managed with gradual downtitration of his blood pressure. Neurosurgical interventions such as placement of an external ventricular drain were initially considered but withheld given improving symptoms and the reversible nature of PRES. There was a gradual resolution of his symptoms over the course of hospitalization. A repeat head CT at 72 hours showed significantly improving vasogenic edema and decreased hydrocephalus (Figure 1). The patient was discharged on the fourth day of hospitalization.

Discussion

The clinical presentation of PRES is highly variable, ranging from mild disorientation, headache, vomiting, and blurry vision^{3,4} to cortical

blindness, seizures, depressed consciousness^{5,6} coma, and death.⁷ Infratentorial involvement causing obstructive hydrocephalus is rare and may be misdiagnosed as an infiltrative tumor, infarction, or encephalitis, which could result in delayed treatment or unnecessary surgical interventions such as biopsy.8 In cases with progressive neurological deterioration, hydrocephalus may require CSF diversion with emergency external ventricular drain placement or endoscopic third ventriculostomy.6,8 Prompt recognition of PRES and its management can help in avoiding these interventions and possible complications.

The exact pathogenesis of PRES is still unknown. The 2 commonly reported mechanisms involve impaired cerebral autoregulation and endothelial dysfunction.3 Under normal circumstances, cerebral vasculature can maintain cerebral blood flow through changes in arteriolar diameter. However, in patients with extreme blood pressure elevation, these mechanisms fail and can result in extravasation across the blood-brain barrier. Such extravasation of fluid can also be seen with other causes of endothelial dysfunction such as autoimmune conditions or exposure to chemotherapeutic or immunosuppressive drugs.4

Our case highlights the association of acute elevation in blood pressure and use of vasoactive drugs such as methamphetamine resulting in PRES and a rare presentation with Figure 1.

Top Row: CT Head On Day 0 Showing Bilateral Parieto-Occipital Edema, Effaced Fourth Ventricle, and Third Ventricular Dilatation of 1.5 cm; Middle Row: CT Head on Day 3 With Decreasing Hydrocephalus, Fourth Ventricular Effacement, and Third Ventricle Diameter to 9.8 mm; Bottom Row: MRI FLAIR Changes With Obstructive Hydrocephalus, Transependymal Edema, and Vasogenic Edema in the Bilateral Cerebellar and Parieto-Occipital Regions Suggestive of PRES



Abbreviations: CT = computed tomography, FLAIR = fluid-attenuated inversion recovery, MRI = magnetic resonance imaging, PRES = posterior reversible encephalopathy syndrome.

obstructive hydrocephalus. Effective management of PRES focuses on early detection of symptoms with aggressive blood pressure control, which can prevent invasive procedures and its associated complications.

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