PRES Syndrome: a frequently missed clinical entity?

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Clinical Case:

22-year-old Afro-American female with a past medical history of SLE, lupus nephritis, crescentic GN, ESRD on hemodialysis, seizure disorder and hypertension non-compliant on medications misses two sessions of hemodialysis and gets admitted to our hospital following two episodes of tonic-clonic seizures. The seizures were separated by a duration of 15 minutes with each lasting 1-3 minutes, accompanied by stool and urinary incontinence. The patient regained consciousness in between the two episodes. Seizure controlled with intravenous lorazepam. Aura preceded seizures for a few minutes. The patient's medications included hydralazine, levetiracetam, methyldopa, metoprolol succinate, mycophenolate mofetil and torsemide. Admission vitals were significant for high BP 227/117 mm Hg and heart rate 108, respiratory rate 21. No motor or sensory deficit was found. MRI brain showed mild patchy increased FLAIR signal involving bilateral posterior occipital subcortical white matter. Mildly increased signal involving bilateral posterior superior frontal and parietal subcortical white matter were seen. The patient was diagnosed to have PRES secondary to malignant hypertension. The patient was given hemodialysis; BP was controlled and subsequently discharged with antiepileptic and antihypertensive medications, with outpatient follow-up.

Discussion:

PRES or Posterior reversible encephalopathy syndrome is defined as a clinical syndrome characterized by insidious onset headache, confusion or altered mentation, visual changes and seizures, associated with characteristic neuroimaging findings of posterior cerebral white matter edema. PRES associated with hypertensive encephalopathy, eclampsia, and the use of cytotoxic and immunosuppressant drugs are the most common. Pathogenesis of PRES remains incompletely understood. It has been related to disordered cerebral autoregulation or endothelial dysfunction. Commonly associated conditions are hypertensive encephalopathy, acute renal injury, sepsis and multi-organ failure, eclampsia, autoimmune disease, use of immunosuppressive drugs (like tacrolimus, cyclosporine, chemotherapy, recreational drugs (like cocaine), organ transplantation, chronic hypertension or chronic kidney disease. PRES evolves over a matter of hours. The most common presentations are seizures, disturbed vision, headache, nausea and vomiting, altered mental status. More than 70% of patients with PRES are hypertensive, though a significant proportion has normal or only mildly raised blood pressure. The main differential diagnosis includes cerebral venous sinus thrombosis, intracranial hemorrhage, occipital lobe ischemic stroke, primary central nervous system vasculitis among vascular causes. The non-vascular causes include Infective encephalitis, autoimmune encephalitis, metabolic or toxic encephalopathy.

A rapid withdrawal of the trigger appears to hasten recovery and to avoid complications in PRES. Removal of the offending drugs (if any), aggressive blood pressure management, delivery of the baby in case of eclampsia, and antiepileptic medicines for treating seizures. Anesthesia and ventilation may be necessary for airway protection in obtunded patients. Corticosteroids theoretically improve vasogenic edema, but till now, there is no evidence for their use in PRES.

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