A case of post-streptococcal glomerulonephritis leading to posterior reversible encephalopathy syndrome

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Abstract

Posterior reversible encephalopathy syndrome (PRES) is a clinical and radiographic syndrome characterized by headache, confusion, seizures, visual disturbance, and characteristic lesions on neuroimaging of the brain. A disorder of the cerebrovascular autoregulation system, it is generally described in adult patients and commonly associated with hypertension, eclampsia and immunosuppressive agents. PRES is a rare but potentially serious complication of acute post-streptococcal glomerulonephritis (PGSN) in children. A treatable diagnosis, it is crucial to consider PRES in a pediatric patient who presents with headache, seizure or confusion in order to improve outcome and prevent permanent neurologic damage.

Hospital Course

Upon arrival to the ED, blood pressure was 134/82. An hour later, he had another episode of unresponsiveness with right gaze deviation and clonic movements in the right arm. Neurology was consulted.

At the time of neurology’s assessment, vitals were notable for a blood pressure of 179/11, heart rate of 92 and temperature of 37.4. Neurologic examination revealed inattention, flattening of the right nasolabial fold, right pronator drift but otherwise full strength, and asymmetric reflexes with increased reflexes throughout the right upper and lower extremity. He had bilateral nonsustained clonus at the ankles and negative Babinski.

He was admitted to the PICU for further work up and management. He was started on a nicardipine drip for persistently elevated high blood pressure and Keppra with no seizure recurrence. Lumbar puncture was unremarkable. MRI head revealed bilateral, symmetric predominantly supratentorial border zone cortical T2 and T2 FLAIR hyperintensity with enhancement, which was suggestive of PRES. EEG was consistent with focal neocortical dysfunction in the occipital region.

Work up of his hypertension was notable for nephrotic range proteinuria with hematuria, normal BUN and Cr, a positive ASO titer, positive antistreptolysin antibody with a low C3, negative ANA and positive ANCA at 1:160. Thyroid function and urine catecholamines were normal. He was ultimately diagnosed with acute post-streptococcal glomerulonephritis.

He was discharged on day 5 of his hospitalization on isradipine and Keppra. Approximately 6 weeks after discharge, his blood pressures normalized and he was able to remain off of blood pressure medications and antiepileptics. He continued to have hematuria but no proteinuria.

Discussion

Posterior reversible encephalopathy syndrome (PRES) is a clinicoradiologic condition first described by Hinchey in 1996. Generally described in adults, PRES is gradually being recognized in the pediatric population. PRES is characterized by headaches, altered mental status, visual disturbances, seizures, and characteristic neuroimaging findings. The mechanism of PRES is unknown, though three theories have been proposed: (1) hypertension-induced cerebral autoregulation loss, (2) cerebrovascular endothelial dysfunction, and (3) vasospasm and hypoperfusion resulting in vasogenic edema. It is thought that the posterior brain is more commonly affected due to a lack of sympathetic innervation.

The most common etiologies of PRES in children are acute elevation in blood pressure, renal failure, fluid retention, and immunosuppressive treatment. The prevalence of PRES associated with acute post-streptococcal glomerulonephritis is unknown, though it is estimated that 5-10% of children hospitalized with acute glomerulonephritis develop PRES. Prompt recognition and treatment of PRES is crucial as delayed diagnosis may result in irreversible cerebral infarction, irreversible cytotoxic edema, neurodevelopmental delay, epilepsy, and vision abnormalities.

The main treatment of PRES is blood pressure control. Often, an antiepileptic is started to minimize recurrence of seizures. Treatment of hypertension leads to resolution of PRES induced neurologic symptoms within 5-7 days.

References


