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Posterior Reversible Encephalopathy Syndrome after Transfusion in Hb E-Beta Thalassemia

Posterior reversible encephalopathy syndrome (PRES), first described by Hinchey, et al. [1] in 1996, is characterized by headache, confusion, seizures, visual loss and sub-cortical edema on neuroimaging. Various etiologies like hypertension, infection, eclampsia and autoimmune disease are implicated. Post-transfusion PRES with pre-existing hematological disorder like sickle cell anemia has also been reported [2]. We report PRES after blood transfusion in a child with Hb E-beta thalassemia.

A 6-year-old girl, known case of Hb E-beta thalassemia, received two units of packed cells as her pre-transfusion hemoglobin was 4.8 g/dL. Two days later, she was admitted with several episodes of projectile vomiting, gradually increasing headache and brief loss of consciousness. There was no history of head trauma, fever, seizures, weakness or visual disturbances. Her vitals were stable with blood pressure of 92/60 mmHg. Glasgow Coma Scale (GCS) score was 12/15; Cranial nerves, superficial and deep tendon reflexes were normal; plantars were flexor. Meningeal signs were absent and hepatosplenomegaly was present.

Complete hemogram and blood biochemistry were normal. Hepatitis-B surface antigen and antibodies to HCV, HIV 1 and 2 were negative. CSF examination revealed 5 lymphocytes/mm³, sugar of 76 mg/dL and proteins 52 mg/dL. MRI of brain revealed multiple hyperintensities over subcortical white matter involving cerebral and cerebellar regions in T2 and Flair images (Fig 1). She recovered completely by second day with supportive management and was discharged on the fourth day. Repeat MRI done after six weeks showed near complete disappearance of hyperintensities.

Failure of cerebral auto-regulation to counteract the sudden rise in cerebral blood volume following transfusion is thought to cause vasogenic edema leading to PRES [3]. Hypoxic damage to endothelial cells secondary to anemia might also disrupt the integrity of the blood brain barrier [4]. Rapid rise of hemoglobin has also been implicated [5].

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References