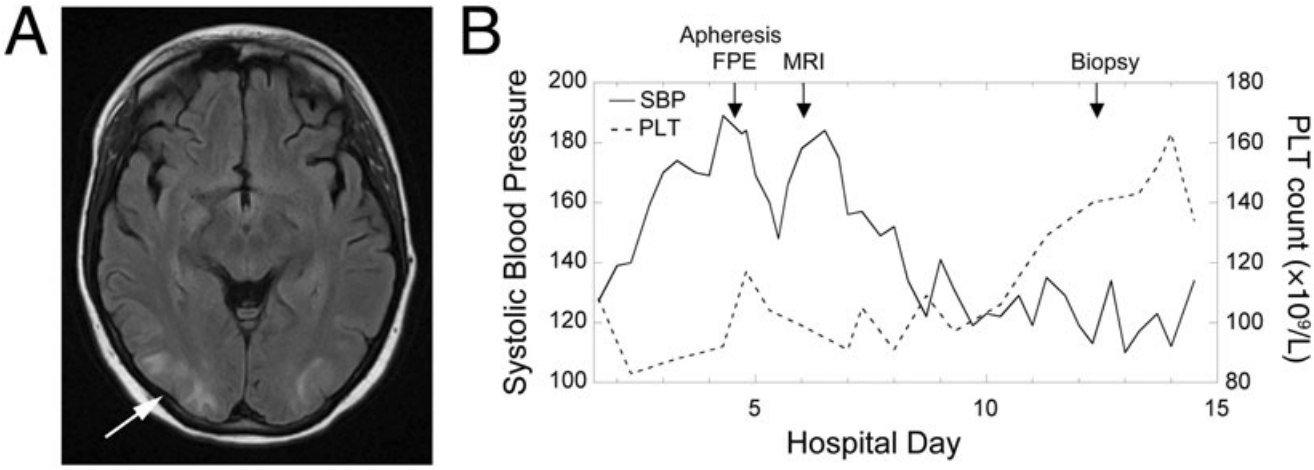

An imPRESsive mimic

Laura Cooling, Steven Gay, and Samuel Silver

A 20-year-old G1P1 woman presented to a local hospital with a 2-week history of headache, blurry vision, photophobia, edema, and easy bruising. She was found to be hypertensive (blood pressure, 180/90) with acute renal failure (Cr = 8.5), thrombocytopenia (platelet [PLT] count, 81 × 10^9/L), elevated lactate deydrogenase (LDH; (698 IU/L), and severe microcytic anemia (hemoglobin, 5.9 mg/dL; mean corpuscular volume, 74) with occasional schistocytes. An ADAMTS13 drawn on Day 2 was 59%. Her medical history was significant for migraines, chronic fatigue after a cesarean section 6 months previously, and a progesterone intrauterine device for birth control. She was diagnosed with thrombotic thrombocytopenic purpura (TTP)/hemolytic uremic syndrome (HUS) and treated with dialysis, plasma exchange, steroids, and rituximab with minimal clinical improvement. She was transferred to a university hospital after a documented episode of flash pulmonary edema (FPE) and possible seizure.

After transfer, the patient was diagnosed with possible HUS. Her laboratory results were significant for a PLT count of 108 × 10^9/L, an LDH level of 260 (range, 252-297), a haptoglobin level of 10 to 15 mg/dL, and ADAMTS13 activity of 78%. Shortly after admission, the patient developed a severe acute headache and underwent plasma exchange, which was complicated by FPE (see figure, B). A cardiac evaluation revealed left ventricular hypertrophy with mild ventricular systolic dysfunction and mitral regurgitation. On Day 5, the patient developed a visual field defect. An MRI showed increased T2 and FLAIR signal, without restricted diffusion or contrast enhancement, in the subcortical white matter of both occipital and parietal lobes, consistent with posterior reversible encephalopathy syndrome (PRES) (see figure, A; arrow). The patient's PLT counts, hallucinations, confusion, and vision subsequently recovered with aggressive blood pressure control and steroid taper. A renal biopsy showed a chronic ischemic vasculopathy due to hypertension with no

From the Departments of Pathology and Internal Medicine, University of Michigan, Ann Arbor, Michigan.

Address reprint requests to: Laura Cooling, MD, MS, Transfusion Medicine, University of Michigan Hospitals, 2F225 UH, Box 0054, 1500 E. Medical Center Drive, Ann Arbor, MI 48109-0054; e-mail: lcooling@med.umich.edu.

Received for publication April 22, 2009; revision received June 18, 2009; and accepted June 19, 2009.

doi: 10.1111/j.1537-2995.2009.02362.x

TRANSFUSION 2010;50:11-12.

evidence of HUS/TTP (see figure, B; PLT count (- - -) and systolic blood pressure (—) over the course of hospitalization. Arrows indicate time of FPE during a plasma exchange, left homonymous hemianopsia and MRI, and renal biopsy).

PRES is characterized by headache, confusion, seizures, and visual disturbances, including cortical blindness, due to vasogenic cerebral edema.[1] Moderate to severe hypertension is reported in 70% to 80% of patients. Clinical risk factors for developing PRES include preeclampsia, eclampsia, HELLP, allogenic transplant, cyclosporine, tacrolimus, autoimmune disease, sepsis, and chemotherapy.[1] Because PRES can occur in the settings of microangiopathic anemia, thrombocytopenia, and renal dysfunction, the onset of PRES and neurologic symptoms may be mistaken for TTP. Like malignant and severe hypertension,[2] the treatment for PRES is aggressive blood pressure control and treatment or removal of inciting risk factors.

CONFLICT OF INTEREST

The authors declare that there are no conflicts of interest relevant to the manuscript submitted to **TRANSFUSION**.

REFERENCES

1. Barynski WS. Posterior reversible encephalopathy syndrome, part 1: fundamental imaging and clinical features. AJNR Am J Neuroradiol 2008;29:1036-42.
2. Egan JA, Bandarenko N, Hay SN, Paradowski L, Goldberg R, Nickeleit V, Brecher ME. Differentiating thrombotic microangiopathies induced by severe hypertension from anemia and thrombocytopenia seen in thrombotic thrombocytopenia purpura. J Clin Apheresis 2004;19:125-9.