Posterior reversible encephalopathy (PRES) associated with HELLP syndrome: a case report

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INTRODUCTION

HELLP syndrome is a poorly understood multisystem pregnancy-related condition with a rapid onset and is typically seen in patients with severe pre-eclampsia. HELLP is associated with generalized vasospasm, microthrombi formation and coagulation defects.

The typical neurological symptoms of PRES include encephalopathy (50-80%), seizure (60-75%), headache (50%), and visual disturbance (35%).

Brain imaging in PRES usually reveals vasogenic edema in the bilateral parietal-occipital regions, and may include the frontal, temporal, basal ganglia, brainstem or cerebellum.

Most patients with PRES have a favorable outcome but mortality can be as high as 3-6%, persistent neurological sequelae in 10-20% patients, and recurrent PRES in 5-10%.

CASE REPORT

PATIENT PRESENTATION

History of presenting illness: a 36-year-old pregnant woman develops, at 35 weeks of pregnancy, pre-eclampsia. Admitted for an emergency C-section complicated with uncontrollable blood loss which led to a hysterectomy and bilateral salpingectomy. Immediate post operative complicated with multiple organ dysfunction (renal, hepatic, hemolysis and thrombocytopenia), associated with a hypertensive hemodynamic profile.

Past medical history: depression.

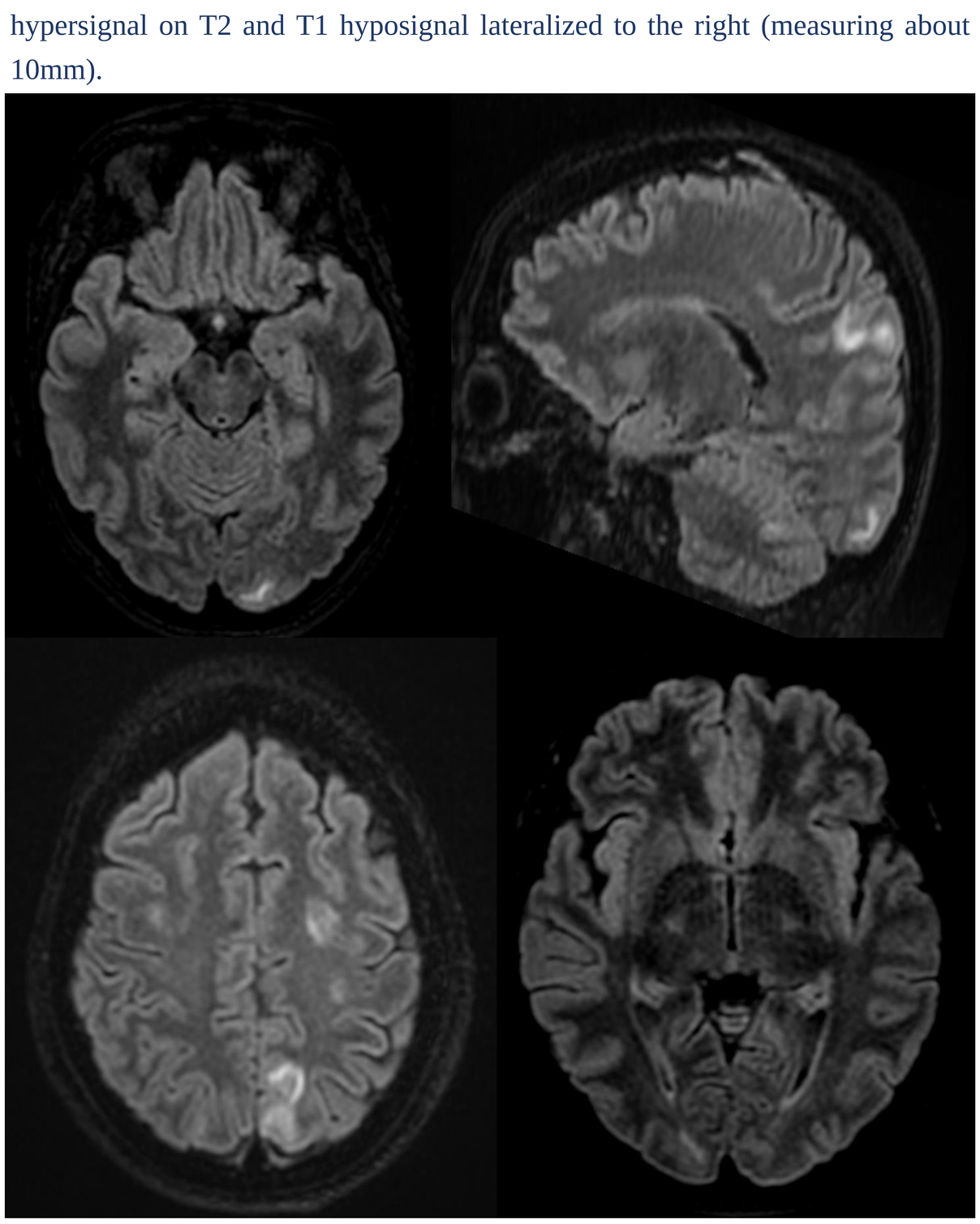
Home medications: sertraline.

Neurological exam: on admission, 14th June, blurred vision. On 20th June, focal seizure - amaurosis, anisocoria and right hemiparesis, concomitant with a hypertensive emergency.

Laboratory values: on admission, 14th June, hepatic and hematologic dysfunction with thrombocytopenia and anemia, hemolysis with moderate anisopoikilocytosis, elliptocytes and schistocytes; LDH 1702 U/L; renal dysfunction KDIGO 3 oliguric (Cr 3.53mg/dL); metabolic acidosis, hyperkalemia, nephrotic proteinuria and hypoalbuminemia (1,96 g/dL). Immunological testing revealed C3 and C4 consumption and IgG deficit. Genetic testing and ADAMTS13 were without alterations. Endocrinological testing (thyroid, corticotroph axis, prolactin, somatotrophic and gonadotropic lines) was normal.

Hospital course: Transferred to the ICU with the diagnosis of HELLP syndrome class I, and started a scheme of daily plasmapheresis. On the 6th day of hospitalization, the patient had a convulsive crisis along with maintenance of a resistant hypertensive profile. A cerebral MRI was performed with evidence of PRES and pituitary adenoma/apoplexy without pituitary hemorrhage. The patient initiated anti-convulsive and anti-hypertensive therapy, with rapid response and favorable evolution with resolution of all dysfunctions.

Imaging: T2/FLAIR hypersignal of subcortical/juxtacortical white matter of parietal regions (more evident/extensive on the left), occipital and posterior frontal, as well as on the posterior-superior aspect of the cerebellar hemispheres (also more evident on the left) and in the splenium of the corpus callosum on the left; small focus of diffusion restriction is admitted in the left parietal region. Adenohypophysis swelling, with a coarsely rounded area of heterogeneous hypersignal on T2 and T1 hyposignal lateralized to the right (measuring about 10mm).



CONCLUSIONS

HELLP syndrome is associated with severe clinical complications, which cause high maternal morbidity and mortality.

PRES pathophysiology is still largely unknown.

PRES should always be considered in women with acute hypertensive disorders and neurological symptoms during pregnancy and post partum.

MRI as a diagnostic gold standard.

Reversible with adequate pharmacological and life support.

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