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Case Report

Brain MRI findings of a 24-week pregnant woman with HELLP syndrome

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ABSTRACT

Introduction: HELLP syndrome (acronym comprised of hemolysis, elevated liver enzymes and low platelets) is seen in 0.1% of pregnant women. Posterior reversible encephalopathy syndrome (PRES) was reported to be 5% in patients with the HELLP syndrome. Atypical imaging appearances include contrast enhancement, hemorrhage and restricted diffusion on MRI.

Aim: We aimed to improve clinicians' perception about brain lesions in the HELLP syndrome with imaging findings.

Case study: Here, we present a case of an 18-year-old patient with a pregnancy of 24 weeks admitted with HELLP syndrome, with CT and MRI findings of PRES and intracerebral hematoma. MRI scan of the brain showed vasogenic edema in the occipital, frontal and parietal lobes bilaterally, basal ganglia and brainstem. An emergency cesarean section was successfully performed to end the pregnancy on the same day. Repeat MRI of the brain three days after initial admission showed partial improvement of the previous abnormalities with full clinical recovery.

Results and discussion: The patient in this report had a variant form of PRES where the pathologic process encompassed both the posterior and anterior circulations.

Conclusions: An appropriate multidisciplinary approach is the key for reducing the morbidity and mortality of PRES syndrome.

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1. Introduction

HELLP syndrome (acronym comprised of hemolysis, elevated liver enzymes and low platelets) is seen in 0.1% of pregnant women.[1] Nearly half of the neurological events related to pregnancy consist of intracerebral hemorrhage, venous thrombosis, subarachnoid hemorrhage and infarction, and the remainder are pregnancy related encephalopathy.[2] Here, we describe a patient with posterior reversible encephalopathy syndrome (PRES) and intracerebral hemorrhage that had occurred secondary to HELLP syndrome.

2. Aim

We aimed to improve clinicians' perception about brain lesions in the HELLP syndrome by demonstrating the imaging findings from our patient.

3. Case study

An 18-year-old woman (gravida 1; para 0) was transferred to our hospital from a local hospital at 24 weeks of gestation having blurred vision, somnolence, headache in the region of the back of the neck and head, mild loss of sensation in distal extremities and high blood pressure (160/110 mmHg). Her neurological examination did not provide additional significant findings. Her past medical history was unremarkable. Serum tests showed total bilirubin of 1.2 mg/dL, lactate dehydrogenase of 2224 U/L, serum aspartate aminotransferase of 187 U/L, serum alanine aminotransferase of 246 U/L and platelet count of $112 \times 10^3\ m^{-3}$. These findings suggested HELLP syndrome. Computed tomography (CT) revealed subarachnoid hemorrhage in the right temporal lobe and intraparenchymal hematoma in the right occipital lobe (Fig. 1). On magnetic resonance (MR), axial fluid attenuated inversion recovery (FLAIR) images demonstrated increased

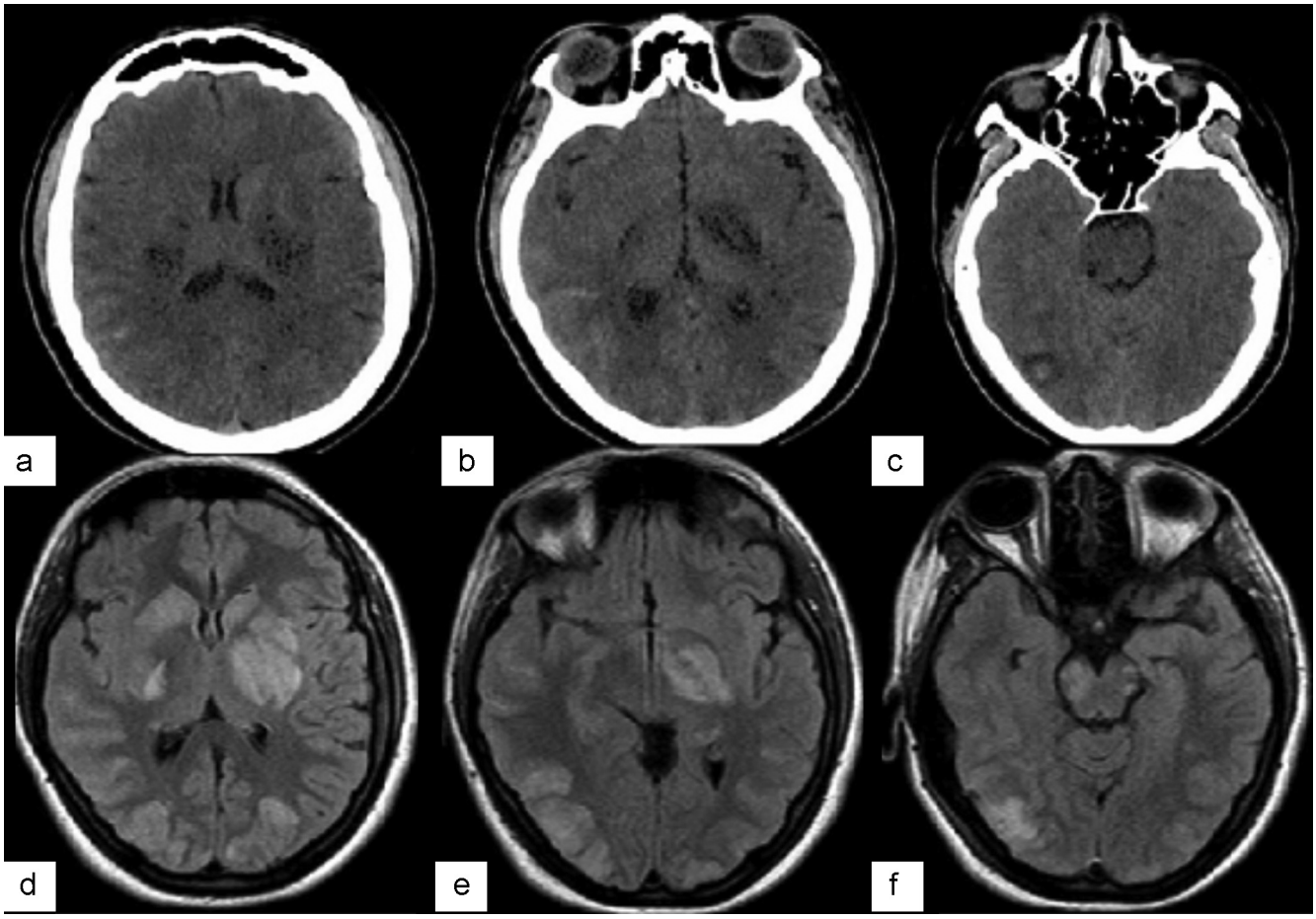

Fig. 1 – (a)–(c) Axial noncontrast CT images on the 1st day of admission reveal faint subarachnoid hemorrhage in the right temporal lobe and intraparenchymal hematoma in the right occipital lobe. (d)–(f) Axial FLAIR MR images demonstrate increased signal intensities in both of the cerebral pedincules, posterior limb of the bilateral internal capsule, left putamen, left globus pallidus extending partially to thalamus, bilateral occipital lobes, and predominantly on gray-white matter junction areas where it can be seen as low density areas on CT.

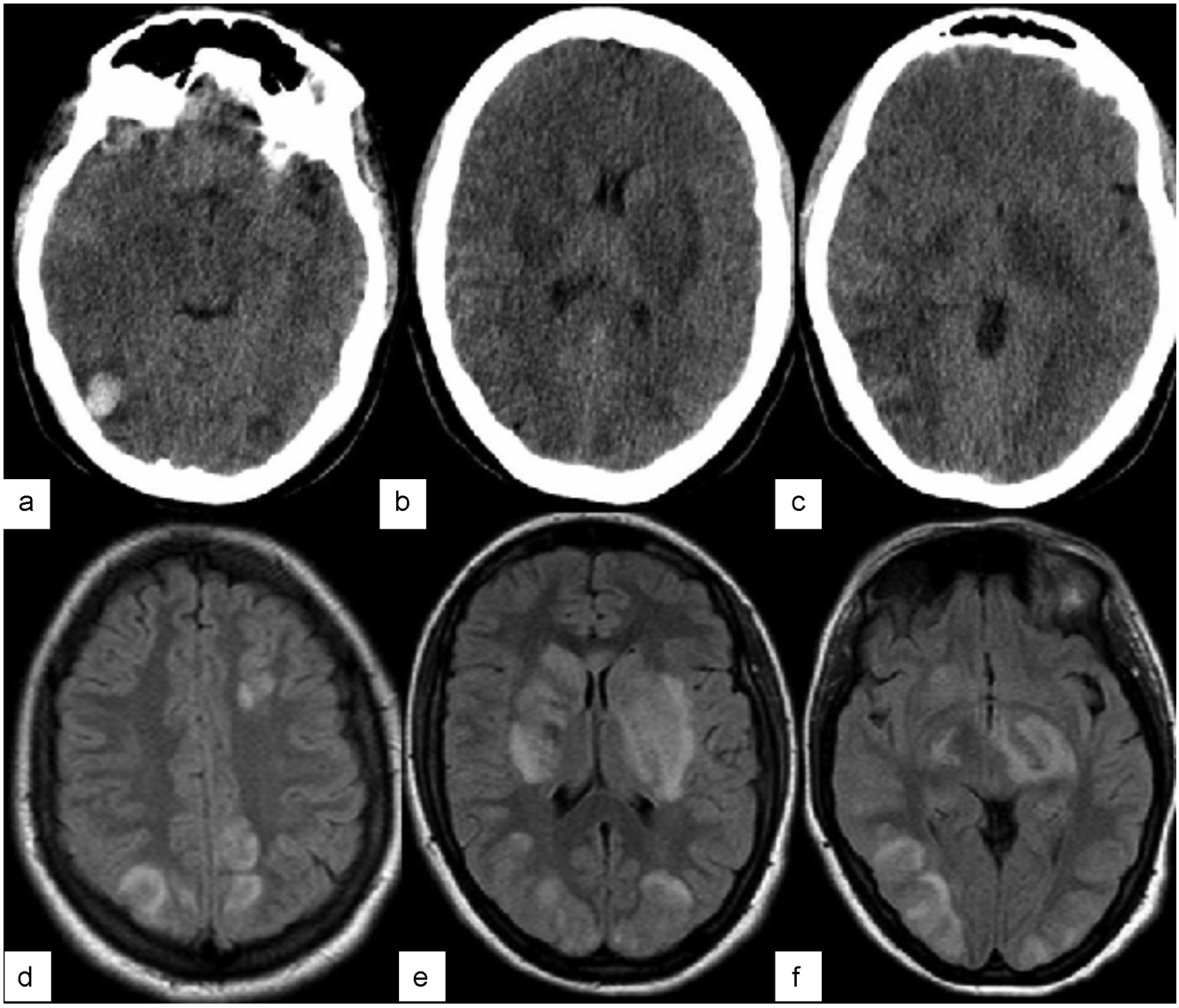

Fig. 2 – Axial CT (a)–(c) and MR (d)–(f) images on the 2nd day the hematoma in the right hemisphere, density and signal abnormalities expanded, and became more evident, contrarily subarachnoid hemorrhage disappeared.

signal intensities in both of the cerebral pedincules, posterior limb of the bilateral internal capsule, left putamen, left globus pallidus extending partially to the thalamus, bilateral occipital lobes, left frontal and right parietal lobes, predominantly in gray-white matter junction areas where it appeared as low density areas on CT (Fig. 1). These signal changes were iso-hyperintense on diffusion weighted imaging, showing no restricted diffusion and indicating vasogenic edema in some areas. Venous MR angiography revealed normal findings. She was aggressively managed with termination of pregnancy on the same day, 6 h after admission. She gave an ex-fetus birth with an APGAR score of 0. Doxazosin and $MgSO_4$ were administered to the patient in order to decrease blood pressure gradually. Her investigations in the postpartum period revealed platelet counts of $20 \times 10^3\ m^{-3}$ at 6 h, $51 \times 10^3\ m^{-3}$ at 24 h and $61 \times 10^3\ m^{-3}$ at 48 h; aspartate transaminase level was 81 IU/L, and alanine transaminase level was 77 IU/L at 6 h, and 42 and 35 IU/L at 48 h, respectively. Her blood pressure was 120/70 mmHg at 24 h. At the 2nd postpartum day control MR and CT scan showed that the hematoma in the right hemisphere, density and signal abnormalities expanded to the right part of the pons, and became more evident, whereas the subarachnoid hemorrhage disappeared (Fig. 2). On the 3rd day of admission, with the patient's full clinical recovery, imaging abnormalities were partially improved (Fig. 3). On the 10th day her clinical state became stable on observation and she was discharged home.

4. Results and discussion

PRES syndrome was first described by Hinchey et al. in 1996.[2] Symptoms of PRES in the general population comprise clinical seizures (87%), encephalopathy (92%), visual symptoms (39%),

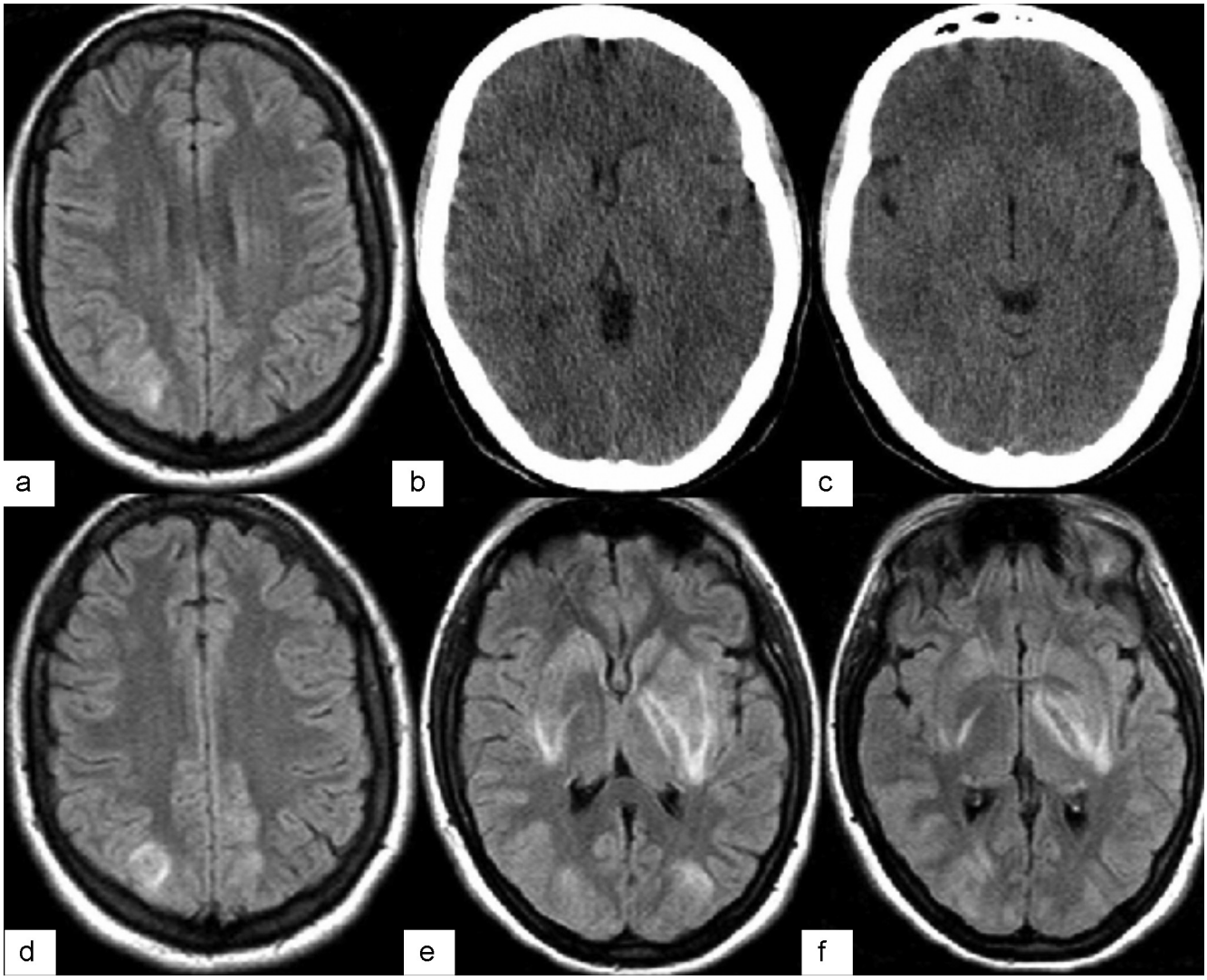

Fig. 3 – Axial CT (b) and (c) and MR (a) (d)–(f) images on the 3rd day of admission, with the patients full clinical recovery imaging abnormalities partially regressed.

and headache (%), resolving after a mean of 5.3 days.[1] The most common causes are diverse and include hypertension, eclampsia/pre-eclampsia, sepsis, immunosuppressive agents, chemotherapy, collagen vascular disease and renal failure.[3]

The exact pathogenesis of PRES remains controversial but the most probable mechanism is vasogenic edema secondary to an acute increase in arterial blood pressure, which overwhelms the autoregulatory capacity of the cerebral vasculature, causing arteriolar vasodilatation and endothelial dysfunction. In addition to this, increased activation of the coagulation system is proposed to play a contributing role in the cause of intracerebral hemorrhage and the increased concentrations of oxyhemoglobin derived from hemolysis and thrombocytopenia in patients with HELLP might exaggerate this vascular response.[4,5] As the name suggests, there is a predisposition to vasogenic edema in PRES for the posterior white matter, because the anterior cerebral circulation has higher sympathetic innervations than the vertebrobasilar system, which is protective against damaging hypertension.[1]

Two types of PRES have been described: classical and variant. The classical form is characterized by clinical symptoms that are associated with bilateral and symmetric vasogenic edema in the parietal and occipital lobes, while the variant form demonstrates predominant involvement of the brain stem and cerebellum.[6,7] A new definition, termed ''central variant,'' describes the form involving the central brainstem and basal ganglia but lacking cortical and subcortical oedema.[8]

The patient in this report had a variant form where the pathologic process encompassed both the posterior and anterior circulations. In addition to the PRES syndrome, the HELLP syndrome frequently involves the cerebellum, brainstem, thalamus and basal ganglia, where less collateral circulation exists.[1]

It was demonstrated that the involvement of the brain stem is not influential in the prognosis, and the influential factor in prognosis is the reversibility of lesions and hemorrhage.[7] Typical imaging findings are demonstrated as hyperintense

areas on FLAIR images in the parietooccipital, posterior frontal cortical and subcortical white matter, while less commonly the brainstem, basal ganglia and cerebellum are involved. Atypical imaging appearances include contrast enhancement, hemorrhage and restricted diffusion on MRI that our patient had intraparenchymal and subarachnoid hemorrhages.[9] She had cerebral edema findings in some regions of the posterior circulation, including both the mesencephalon and pons, and the anterior circulation, especially in the areas of gray and white matter junctions.

The frequency of hemorrhage in PRES was reported to be 5% in patients with eclampsia/delayed eclampsia.[10] Subarachnoid hemorrhage (SAH) is the most common type and the second most common is parenchymal hemorrhage in patients with PRES.[7,9] Our patient had both types of hemorrhage in the right temporal lobe. Findings of PRES on angiography include focal or diffuse vasoconstriction, vasodilation or a "string-of-beads" appearance. This vessel irregularity can also be seen on arterial CT/MR angiography. It may be helpful but is not essential for reaching a correct diagnosis that we did not prefer to perform such an imaging method. These findings may be confused with other diagnoses, such as vasospasm or vasculitis.[3] In a retrospective study by Legrie et al., among 70 patients with PRES, imaging abnormalities resolved in 88%, ischemic and/or hemorrhagic complications occurred in 14% and the mortality rate was 16%.[11]

Our patient demonstrated both PRES and intracerebral hemorrhage findings. Fortunately, she was treated without any sequela. Several years after a pregnancy, formerly pre-eclamptic women have cerebral white matter lesions more often and more severely than control women with normotensive pregnancies.[12] So these patients should be on close neurologic follow-up for the remainder of their lives. We recommend MRI for symptomatic patients with suspected PRES as well as for asymptomatic patients with severe pregnancy induced hypertension to determine whether cerebral edema exists so that the patient can undergo urgent delivery to prevent devastating neurological consequences. In treatment of PRES, the most important considerations are elimination or reduction of the causative drug, aggressive management of blood pressure in patients with hypertension, treatment of seizures, and urgent delivery by cesarean section for patients who exhibit refractory symptoms.[6]

5. Conclusions

An appropriate multidisciplinary approach is the key for reducing the morbidity and mortality of PRES syndrome especially in pregnant women.

Conflict of interest

None declared.

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