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CE - MEDICAL ILLUSTRATION

Central PRES (posterior reversible encephalopathy syndrome) in HELLP syndrome

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Case presentation

A 24-year-old woman at 35-weeks of pregnancy with haemolysis, elevated liver enzymes and low platelets (HELLP syndrome), in the context of severe pre-eclampsia with high blood pressure (180/110 mmHg), suddenly complained of drowsiness and extreme weariness in the upper and lower limbs with gait difficulties. A computed tomography (CT) examination was performed, showing bilateral symmetric hypodensity of the basal ganglia, without contrast enhancement (Fig. 1a). Subsequent magnetic resonance imaging (MRI) showed hypointense areas on T1-weighted imaging (WI) (Fig. 1b) and high intensity on T2WI and FLAIR sequences (Fig. 1c, d), consistent with vasogenic oedema involving symmetrically the lenticular and caudate nuclei, and with extension to the internal and external capsule. A mild diffusion restriction indicating cytotoxic oedema was present in some areas (Fig. 1e, f). MRI angiography (not shown) showed permeability of the venous system. After pregnancy interruption, intensive antihypertensive and supportive treatment, and recovering from the HELLP syndrome and pre-eclampsia, the patient experienced a rapid and complete recovery of the neurological status. Follow-up MRI 2 weeks later showed complete resolution of the previous findings (Fig. 1g, h). Based on the clinical and radiological presentation, and on the evolution with complete clinical and imaging recovery, the final diagnosis was posterior reversible encephalopathy syndrome (PRES). Clinical, laboratory, and imaging follow-up ruled out alternative diagnoses.

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PRES is a radio-clinical entity occurring in predisposing clinical conditions such as hypertension, immunosuppressive agents, uraemia, autoimmune diseases, and neoplasms. The neurological symptoms of PRES are nonspecific. No specific tests to diagnose PRES or to predict the outcome are available, which renders clinical tasks complicated on the one side hand, and makes the imaging assessment critical on the other side. The pathophysiology of PRES is controversial, the dysfunction of the cerebrovascular regulation mainly being mainly incriminated [1].

In the acute stage, the typical imaging pattern is a vasogenic oedema in the subcortical parieto-occipital white matter, with radiologic improvement on follow-up imaging in 1–2 weeks. However, ischemic and haemorrhagic complications do occur in 10–20% of cases, especially if the condition is not promptly recognized and treated [1, 2].

The involvement of basal ganglia (central PRES) was is reported in around 10% of cases of PRES. The symmetric basal ganglia oedema with extensions to the internal and external capsules, associated with some mass effect, and surrounded by a more hyperintense rim delineating the lentiform nucleus on T2-WI, as in our case, was previously described as the "lentiform fork sign". This sign was has been reported in acute uremic encephalopathies (such as acute glomerulonephritis, hemolytic-uremic syndrome and thrombotic thrombocytopenic purpura) and in metabolic acidosis [3]. However, no uraemia or metabolic acidosis was present in our case, which makes these observations of particular interest.

The differential diagnosis of central PRES includes other metabolic processes such as hypoxic injuries and toxic encephalopathies (as methanol and carbon monoxide). However, these conditions usually are associated with restricted diffusion meaning ischemic complications, and a common involvement of the cortex. Symmetric vasculitis-like processes related to tuberculosis and fungal basal meningitis could can also bilaterally involve bilaterally the basal ganglia, but contrast enhancement at the skull base is

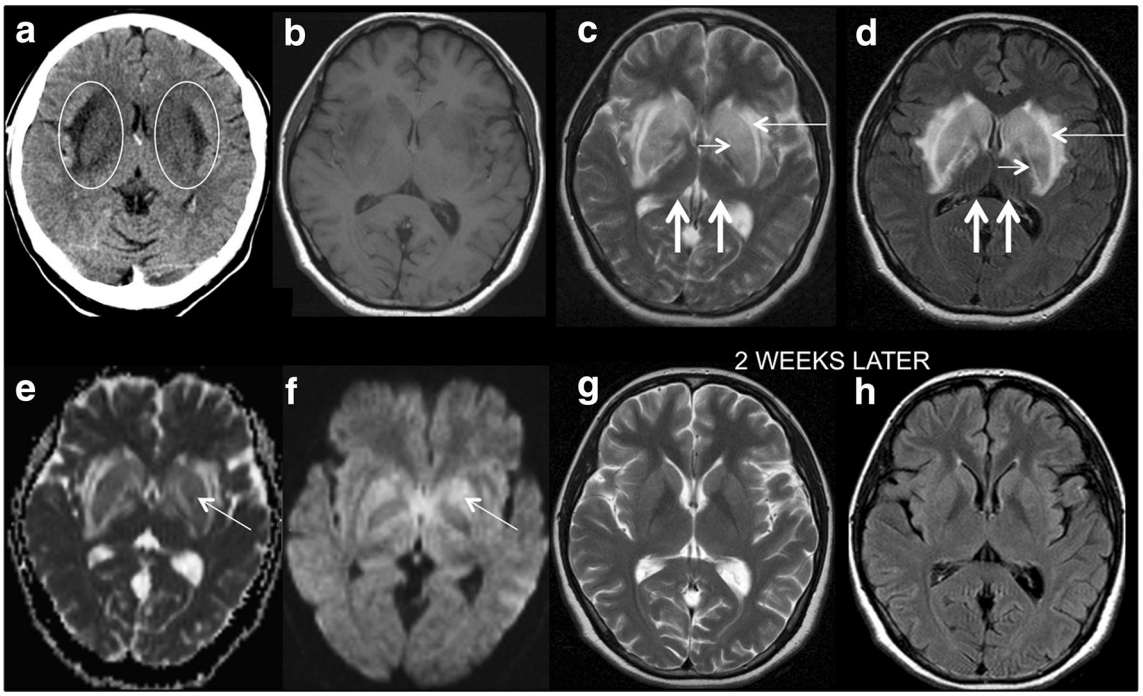

Fig. 1 CE-CT shows bilateral symmetric hypodensity of the basal ganglia, without contrast enhancement (circles in **a**). MRI exam (T1-WI in **b**; T2-WI in **c**; FLAIR sequence in **d**; ADC and DWI maps in **e**, **f**; follow-up T2-WI and FLAIR in **g**, **h**), in on the same day, demonstrates extensive involvement of the basal ganglia with extensions to the internal and external capsules, but sparing the thalami (thick arrows in **c**, **d**). A bright hyperintense rim delineates the boundaries of both putamina, laterally the external capsule (long arrows in **c**, **d**) and medially the external medullary lamina (short arrows in **c**, **d**), giving the appearance of the "lentiform fork sign". Slightly restricted diffusion was present in same areas on ADC-DWI maps (arrows, **e**, **f**). Follow-up MRI (**g**, **h**) shows complete resolution of abnormalities. *CE-CT* contrast-enhanced computed tomography, *MRI* magnetic resonance imaging, *T1-* and *T2-WI* T1- and T2-weighted imaging, *FLAIR* fluid-attenuated inversion recovery, *ADC* apparent diffusion coefficient, *DWI* diffusion weighted imaging

usually prominent, and small infarcts commonly occur in severe cases. Deep veins thrombosis is another differential, although the thalami are mainly involved. Other metabolic conditions such as dysmyelinating disorders should also be ruled out [1, 2, 4].

This case illustrates a central PRES in a HELLP syndrome, and shows a particular MRI pattern named the "lentiform fork sign". More importantly, the present report highlights the importance of need for a prompt diagnosis and treatment initiation in a patient with central PRES, for avoiding potential disabling complications, and for to permit the complete recovery of the patient.

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Compliance with ethical standards

Conflict of interest The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Statement of human and animal rights All procedures related to this report were in accordance with the ethical standards of the institutional and national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed consent The informed consent was obtained from the patient in order for her anonymized data to be used for publication.

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