Posterior Reversible Encephalopathy Syndrome presenting with atypical findings: report of two cases

Cerrone P, Sucapane P, Mearelli S, Baldassarre M, Totaro R, Sacco S, Carolei A*, Marini C

Department of Neurology, University of L'Aquila, 67100 L'Aquila, Italy

****Chairman of the School of Neurology, University of L'Aquila***

Background

PRES is characterized by a variable association of symptoms including headache, consciousness impairment, visual disturbances, nausea/vomiting, seizures and focal neurological signs. Symptoms usually undergo partial or complete resolution within days or weeks identifying and treating the underlying cause. PRES is associated with a series of conditions such as exposure to toxic agents, hypertension, infection, preeclampsia/eclampsia, autoimmune diseases and renal failure. Brain MRI findings include hyperintensities on T2-weighted sequences and their reversibility on follow-up exams.

We describe two patients, one with atypical neuroimaging findings, not included among the most common patterns described in the literature and one with atypical clinical presentation.

Case 1

The first patient was a 42-year old woman, 34th week pregnant, admitted to hospital with premature rupture of membranes. Her past medical history was negative. On admission physical examination was unremarkable. CBC showed severe microcytic hypochromic anemia (Hb=6.9 mg/dL, Hct=25.2%, MCV=70.8 fL, MCHC=27.4 g/dL). Anemia was deemed chronic and was attributed to multiple uterine myomas. The patient was transfused and the following day she underwent cesarian section. On the third day after surgery she presented a generalized tonic-clonic seizure which was treated with diazepam IV. EEG showed only rapid low amplitude waves. Neurologic examination revealed a lethargic status, with a severe right hemiparesis. Brain MRI showed multiple areas with hyperintense signal in T2, DWI and Flair sequences involving not only posterior areas but also frontal lobes and right thalamus. ABP values were moderately high (highest value registered was 160/100 mmHg). Funduscopy examination revealed an acute retinal hemorrhage. The patient was treated with antihypertensives, antiepileptic (levetiracetam), and osmotic therapy. Three days after the reported seizure, the patient had improved and after twelve days she had only a mild right hemiparesis. After 19 days, a control brain CT showed complete resolution of brain alterations.

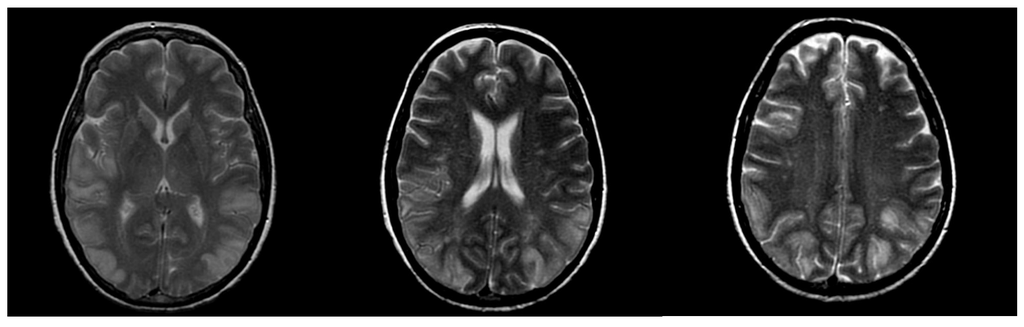

Figure 3: CASE 2. Brain MR showing hyperintensities on T2 weighted sequences

Case 2

A 59-year old woman, with a history of hypertension complicated with retinopathy and treated with antihypertensives was admitted to hospital complaining headache, confusion and postural instability. The following day the patient worsened with severe impairment of consciousness. Neurologic examination revealed lethargy, loss of verbal comprehension, poor and inadequate verbal production, dysphagia, and cortical blindness. Brain CT was normal. On admission the patient had metabolic abnormalities including hypoglycemia (49 mg/dL), hypokaliemia (3.0 mEq/L), hypocalcemia (6.3 mg/dL) and hypomagnesemia (1.3 mg/dL). Physical examination was normal. Treatment was aimed to correct metabolic imbalance. The day after admission the patient had two generalized tonic-clonic seizures. EEG showed widespread theta and delta sub-continuous activity. MRI showed hyperintensities in T2 and DWI sequences, localized in parietal and occipital lobes with mild cerebral edema. MR-Angiography did not show vascular abnormalities. CSF examination was normal. The patient was treated with osmotics and antiepileptics (levetiracetam). After 21 days from onset, the patient was awake, well oriented to space and people, though disoriented to time; she showed good verbal comprehension and answered appropriately to questions. 1 month after admission, Brain MRI showed a partial regression of signal alterations in T2 and Flair sequences.

Conclusion

In the first patient, the etiology of PRES could be linked to a pregnancy-related acute hypertensive episode, particularly to a late postpartum encephalopathy (LPE). The patient had not an history of hypertension but ABP values and funduscupy findings suggested the occurrence of an acute hypertensive episode. LPE frequently occurs in women without signs of a preeclamptic syndrome.

Imaging alterations are tipically bilateral and localized in the posterior white matter, but frontal and temporal lobes are often involved. Atypical localizations are cortical grey matter, basal ganglia, brainstem and cerebellum. In the first case there was an atypical neuroimaging pattern, with vasogenic edema involving white matter and cortex of all lobes and right thalamus with patchy distribution.

In the second patient PRES was probably linked to hypertension. The patient had a history of complicated hypertension. However during hospitalization ABP values were in the normal range. The patient had severe metabolic disorders like hypomagnesaemia, that has been associated with PRES. Clinical presentation was atypical with severe and prolonged impairment of consciousness without clinical improvement within the first weeks.

In conclusion, the cases we presented suggest that PRES may present with atypical imaging and clinical presentation and that high blood pressure episodes may be overlooked. PRES may mimic other neurological conditions. An important criterion to identify PRES is the reversibility of symptoms and radiological findings, but this may occur later than expected. Collecting carefully the medical history and searching possible signs of the associated conditions in suspected PRES may help in the diagnostic process.

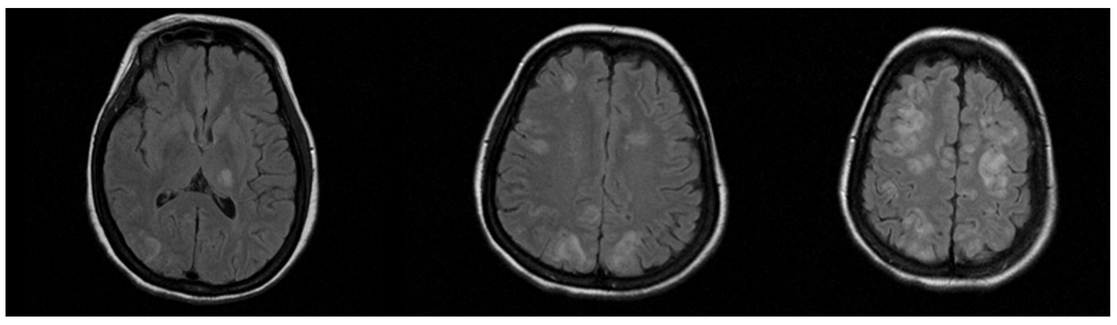

Figure 1: CASE 1. Brain MR showing hyperintensities on Flair sequences

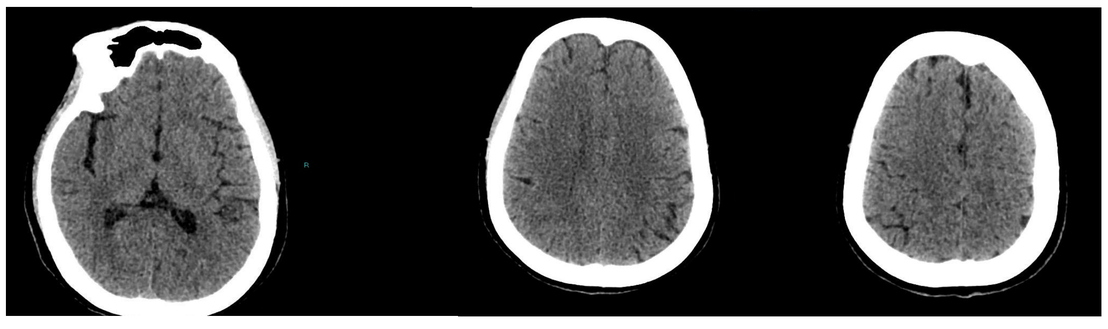

Figure 2: CASE 1. Brain TC performed after 19 days showing resolution of brain alterations

Bibliography

1. Legriel S, Pico F, Azoulay E. Understanding Posterior Reversible Encephalopathy Syndrome. *Annual Update in Intensive Care and Emergency Medicine*. 2011; Vincent J.L. (Ed.); volume 1; 631-653
2. Legriel S. et al. (Critically III Posterior Reversible Encephalopathy Syndrome Study Group). Determinants of Recovery from Severe Posterior Reversible Encephalopathy Syndrome. *Plos One* 2012; volume 7, issue 9
3. Bartynski WS. Posterior Reversible Encephalopathy Syndrome, Part 1: Fundamental Imaging and Clinical Features. *Am J Neuroradiol* 2008; 29:1036–42
4. Bartynski WS, Boardman JF. Distinct imaging patterns and lesion distribution in posterior reversible encephalopathy syndrome. 2007, *AJNR Am J Neuroradiol* 28: 1320–1327.
5. Hugonnet E. et al. Posterior reversible encephalopathy syndrome (PRES): Features on CT and MR imaging. 2013, *Diagnostic and Interventional Imaging* 94, 45—52
6. Farhana S., et al. Posterior Reversible Encephalopathy Syndrome (PRES) with postpartum Eclampsia: a case report. 2011, *Nepal Journal of Neuroscience* 1: 90-93