

Case Report

Spectrum of Imaging Findings in Posterior Reversible Encephalopathy Syndrome (PRES) from Typical to Atypical

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ABSTRACT

Posterior reversible encephalopathy syndrome (PRES) is a clinico-radiological syndrome characterized by headache, consciousness impairment, seizures, and visual deficits and is associated with white matter changes predominantly affecting the posterior parietal and occipital lobes of the brain. Apart from the above-described typical location of the changes, the most common atypical location involves the brain stem/cerebellar hemispheres and basal ganglia. Additionally, PRES may be complicated in some cases by the presence of cytotoxic edema and haemorrhage.

Since magnetic resonance imaging (MRI) is more sensitive and specific imaging technique compared to computerized tomography, establishing the diagnosis and follow-up in patients with PRES is based mainly on MRI findings. It is particularly important not to exclude PRES as a possible diagnosis when we have the appropriate clinical presentation accompanied by the atypical radiological findings, since this clinical-radiological syndrome can often be manifested with an atypical MRI image.

Keywords: Posterior reversible encephalopathy syndrome, magnetic resonance, eclampsia, hypertension

INTRODUCTION

Posterior reversible encephalopathy syndrome (PRES) was first described by Hinchey, in 1996 ^[1], as a reversible syndrome manifested with an acute headache, consciousness impairment, seizures, and visual deficits, associated with white matter changes predominately affecting the posterior parietal and occipital lobes of the brain ^[2].

A multitude of conditions may lead to the development of PRES, with most common etiologies reported including moderate to severe hypertension, preeclampsia/eclampsia, infection with sepsis and shock, autoimmune disease such as systemic lupus erythematosus, multidrug chemotherapy regimens most often in the setting of hematopoietic malignancies, and in the setting of bone marrow and stem cell transplantation ^[1,3].

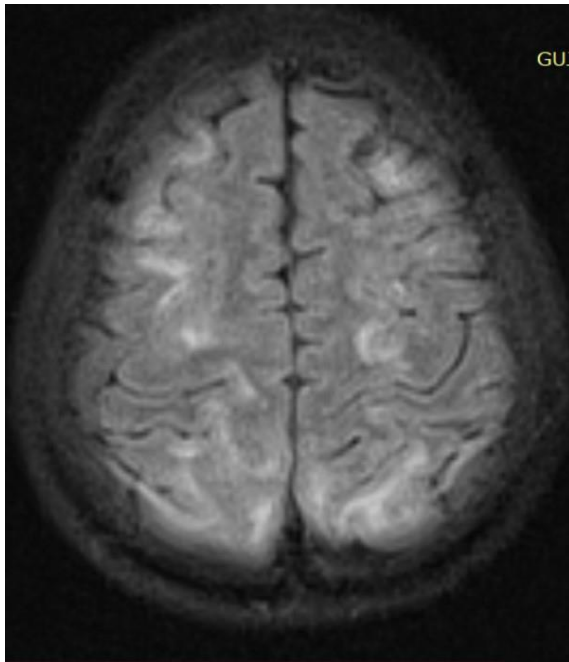
Posterior reversible encephalopathy syndrome (PRES) may be complicated by the presence of

haemorrhage, which utilized gradient echo T2* (GRE) images. Of note, haemorrhage was significantly more common in patients following bone marrow transplantation than in solid organ transplantation, potentially based on underlying coagulopathy, with similar increased incidence in those patients receiving systemic anticoagulation ^[4].

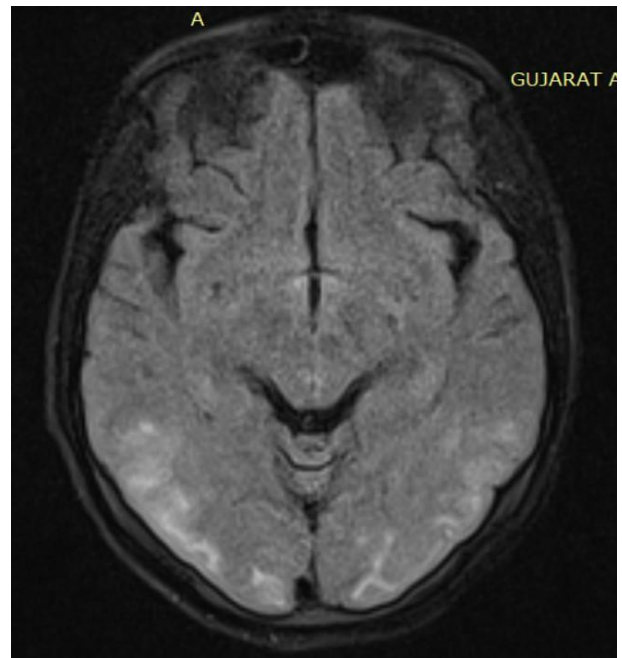
The mechanism of haemorrhage in PRES may be secondary to pial vessel rupture in the setting of severe hypertension or reperfusion injury in the setting of vasoconstriction ^[5].

CASES

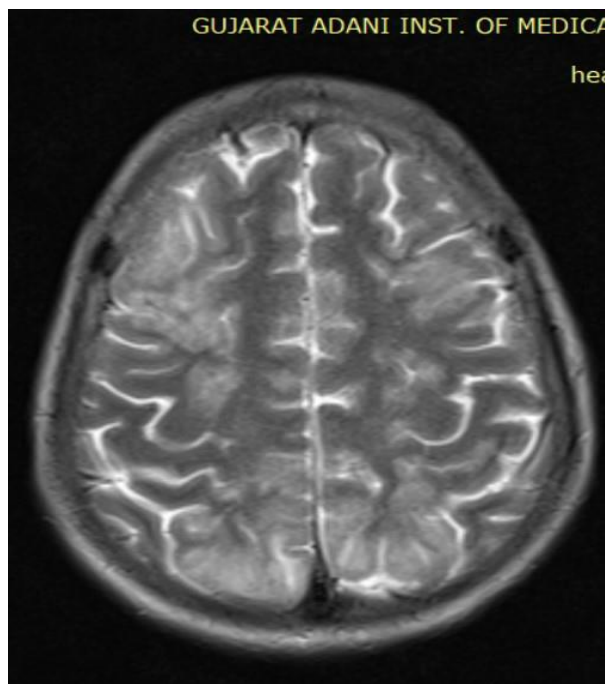
CASE 1: 21-year-old pregnant Female patient with a history of eclampsia presented to Emergency department after a seizure episode. Upon presentation the patient was disoriented and vitals revealed blood pressure of 160/100 mmHg.



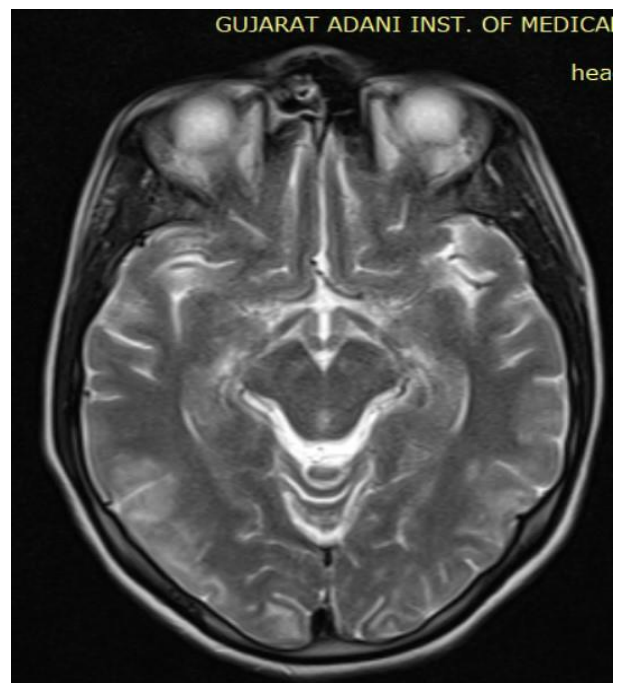
1A



1B



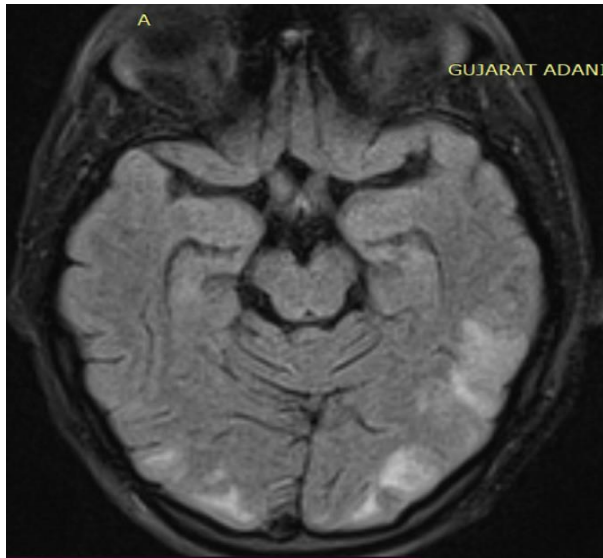
1C



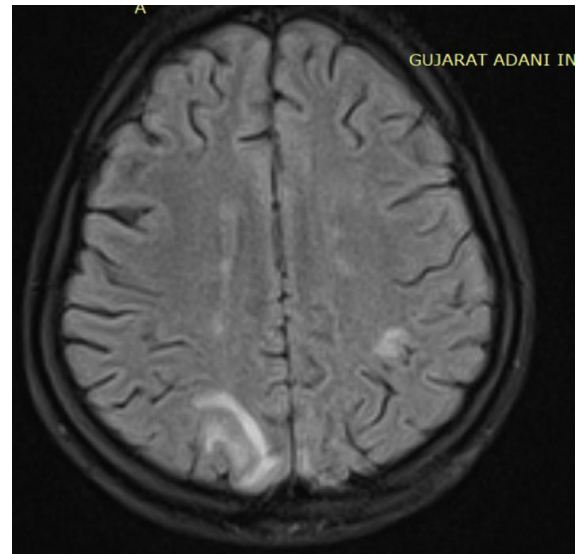
1D

Figure 1. Common presentation of posterior reversible encephalopathy syndrome. There are multiple symmetrical patchy areas of FLAIR (Image A and B) /T2 (Image C and D) hyperintensities seen in cortical, subcortical area of bilateral parieto-occipital and frontal region.

CASE 2: A 27-year-old postpartum female presented to us with history of generalised tonic -clonic seizures lasting 5 min duration associated with tongue bite and loss of consciousness preceded by headache since 2 days. Her antenatal period was uneventful. There was no history of diabetes or hypertension during pregnancy. Vitals on admission showed a blood pressure of 140/90mm Hg, heart rate of 82 per minute, saturating at 100% on room air.



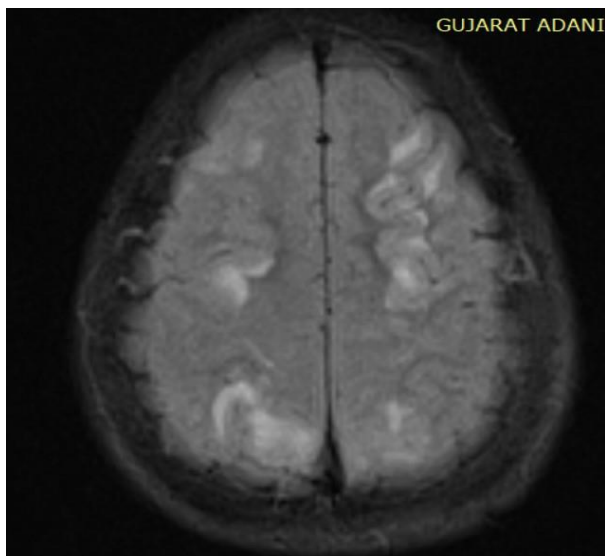
2A



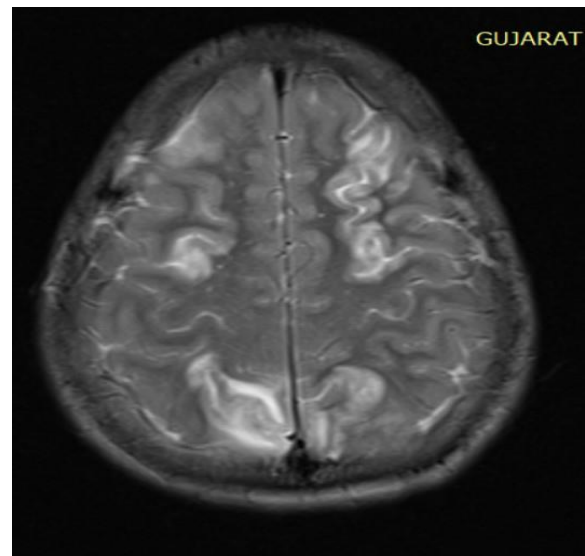
2B

Figure 2. Common presentation of posterior reversible encephalopathy syndrome. There are multiple patchy areas of FLAIR hyperintensities (Image A and B) seen in cortical, subcortical area of bilateral parieto -occipital region.

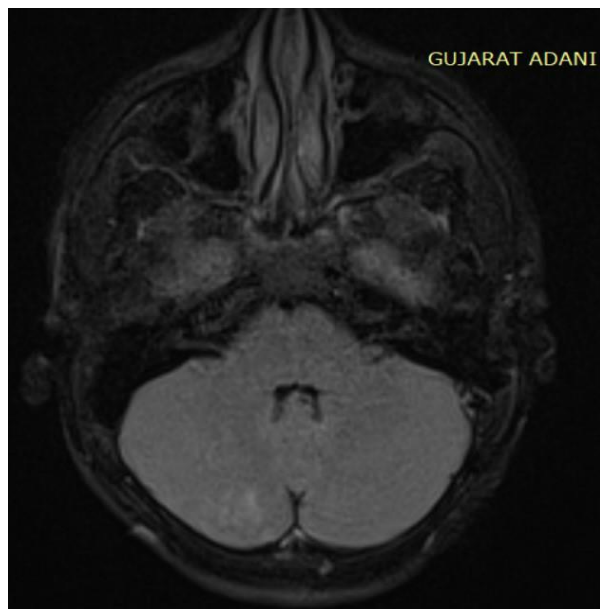
CASE 3: 31-year-old male patient with a history of HTN and CKD presented to emergency department after a seizure episode. Upon presentation, vitals revealed blood pressure of 180/110. The patient was then intubated for airway protection. Patient was started on labetalol drip for BP control. Laboratory workup showed no significant abnormalities besides worsening kidney function.



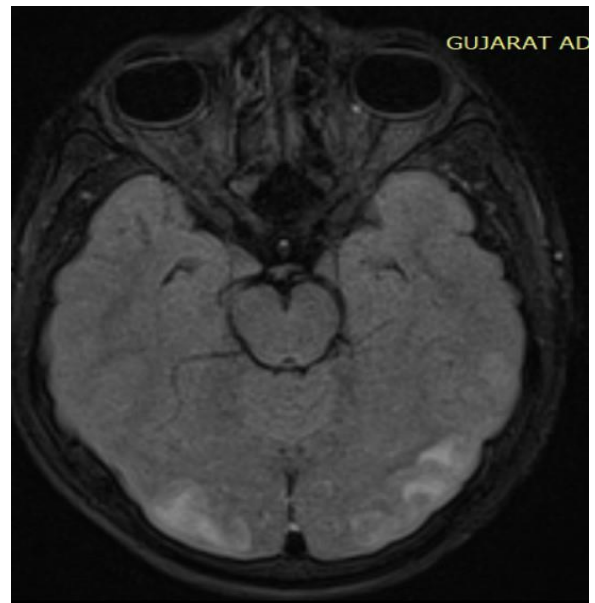
3A



3B



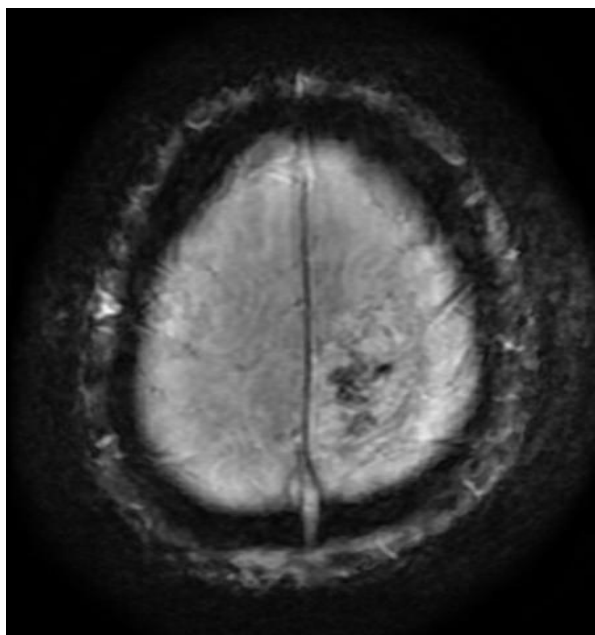
3C



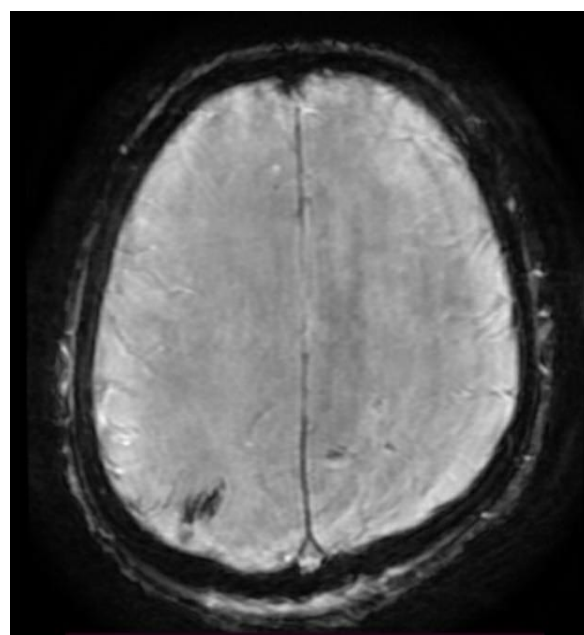
3D

Figure 3. There are multiple symmetrical patchy areas of T2/FLAIR hyperintensities seen in cortical, subcortical area of bilateral frontoparieto occipital region (Image A, B and D). Also, there is patchy areas of hyperintensities of right cerebellum (Image C), which is uncommon location of involvement in posterior reversible encephalopathy syndrome.

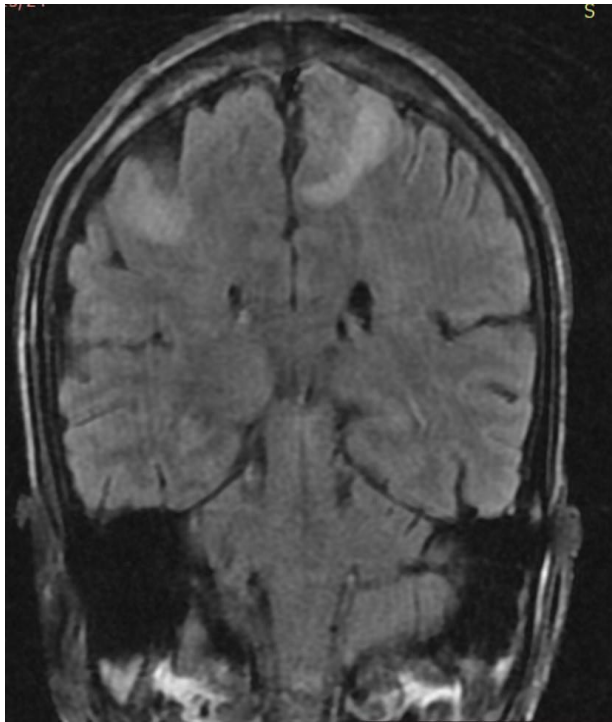
CASE 4: A 35-year-old male was brought to the emergency room with the history of three episode of generalised tonic-clonic seizures followed by loss of consciousness. Relatives of patient revealed that patient had underlying glomerulonephritis.



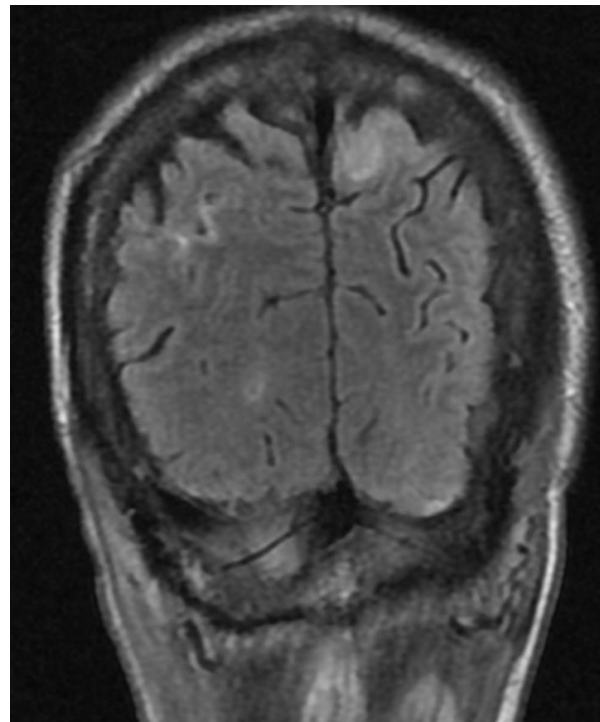
4A



4B



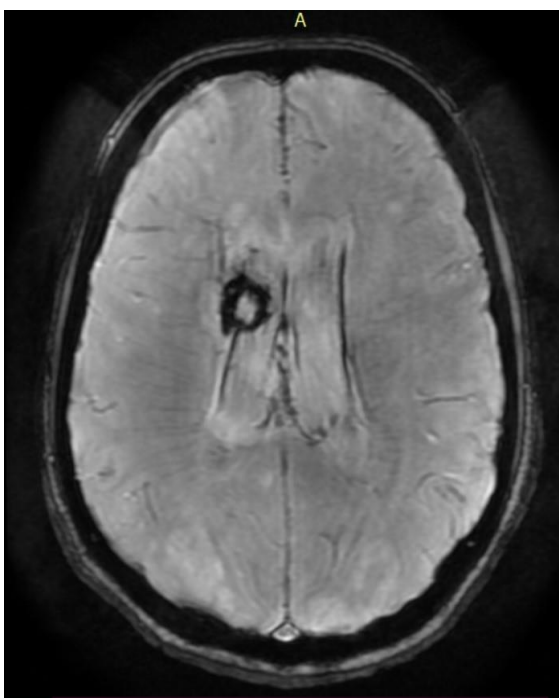
4C



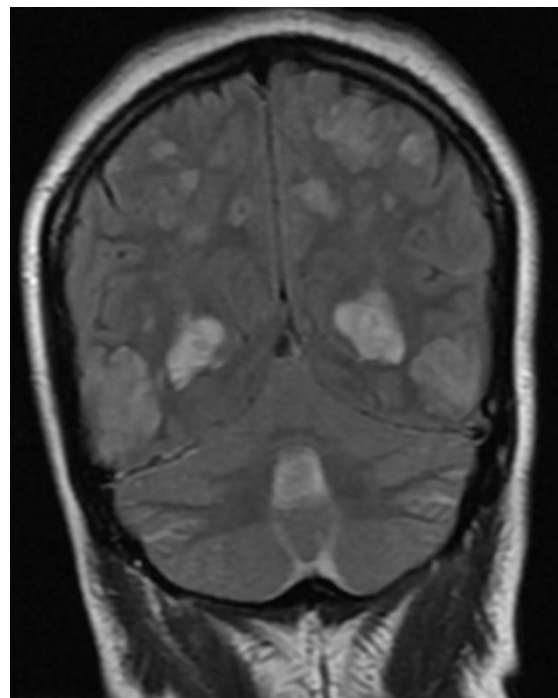
4D

Figure 4. Uncommon presentation of posterior reversible encephalopathy syndrome. There is areas of susceptibilities are seen in bilateral parietal lobes on gradient images (Image A and B) representing intraparenchymal haemorrhage. There are multiple symmetrical patchy areas of FLAIR hyperintensities seen in cortical, subcortical area of bilateral parieto-occipital region (Image C and D) in coronal images representing posterior reversible encephalopathy syndrome.

CASE 5: A 30-year-old postpartum female presented to us with history of generalised tonic -clonic seizures lasting 10 min duration associated with tongue bite and loss of consciousness. There was history of hypertension during pregnancy. Vitals on admission showed a blood pressure of 170/90 mm Hg.



5A



5B

Figure 5. Uncommon presentation of posterior reversible encephalopathy syndrome. There are areas of susceptibilities are seen in right lateral ventricle (Image A) on gradient images representing intraventricular haemorrhage. There are multiple symmetrical patchy areas of T2 hyperintensities seen in cortical, subcortical area of bilateral temporoparietal region (Image B) in coronal image representing posterior reversible encephalopathy syndrome.

DISCUSSION

The typical MRI findings of this syndrome are symmetric hyperintensity in bilateral parietooccipital cortical-subcortical white matter (98%) on FLAIR images. However, the frontal lobe, temporal lobe, cerebellum, basal ganglia, brain stem, and deep white matter can also be involved. The involvement of brain stem or basal ganglia with sparing of the subcortical regions was named as “central-variant” PRES (4%) and involvement of the splenium of the corpus callosum was distinctly reported in 10% of the patients [6].

Intracranial haemorrhage is known to occur in PRES and it could present as intraparenchymal hematoma, sulcal subarachnoid haemorrhage, and minute haemorrhages [4].

A significant fact related to PRES is the possible development of this disorder with no remarkable elevation of blood pressure. This is particularly the case during pregnancy, where there is a shift of cerebral autoregulation curve to the lower range of blood pressure. Since autoregulatory capability is completely attenuated if there is a severe endothelial injury, as it is the case in pre-eclampsia, even a moderate rise of blood pressure may lead to neurologic symptoms which culminate in eclamptic seizures [7].

Reversible cerebral vasoconstriction syndrome (RCVS) should also be mentioned as an important differential diagnosis. RCVS is a clinical-angiographic syndrome that occurs usually in women and is manifested by sudden attacks of severe headache, focal neurological defects, segmental narrowing and dilatation of large and medium cerebral arteries. There is a significant overlapping of clinical and radiological features between RCVS and PRES. Moreover, the two entities often occur simultaneously. RCVS may be idiopathic or occur after an administration of vasoactive drugs, migraine, in pregnancy, and puerperium. In RCVS patients, the major cerebral arteries are affected and stroke occurs, while in PRES patients the small blood vessels, arterioles, and capillaries are affected. PRES can be complicated by ischemic stroke, similar to the RCVS, which is presented with “watershed” distribution where subcortical mass is affected and the cortex is saved [8,9].

Regarding the pathogenesis, autoregulatory failure and endothelial injury have been proposed. Autoregulation is

the maintenance of a constant blood flow in the brain by arteriolar constriction and dilatation regulated by sympathetic innervation. Sudden elevation of systemic blood pressure could exceed the capacity of autoregulation and the arterioles dilate resulting in brain hyperperfusion, blood brain barrier breakdown, and extravasation of fluid and blood products. This could also explain the posterior predilection of the syndrome as the arteries of the posterior circulation have relatively poorer sympathetic innervation [1].

DIFFERENTIAL DIAGNOSIS

Differential diagnosis includes acute cerebral ischemia, cerebral venous thrombosis, transient cerebral hyperaemia, gliomatosis cerebri, infectious meningitis, encephalitis, post-infectious encephalomyelitis, vasculitis, epinephrine-induced toxic or metabolic encephalopathy, and demyelinating disorders.

CONCLUSION

It is of particular importance not to exclude PRES as a possible diagnosis when we have the appropriate clinical presentation which is not accompanied by the typical radiological findings since this clinical-radiological syndrome can often be manifested with atypical MRI findings. PRES can manifest as late postpartum eclampsia without prior evidence of preeclampsia and eclampsia during antenatal period. The recognition of atypical imaging manifestation of PRES is important to avoid delays in diagnosis and treatment, as is identification of complicating factors which may adversely affect patient prognosis.

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