



## Unusual presentation of reversible encephalopathy syndrome with classic and atypical imaging findings

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### Abstract

Posterior reversible encephalopathy syndrome is common manifestation of acute hypertensive encephalopathy usually seen in peak age of 20-40 years commonly associated with eclampsia seen presenting with symptoms such as seizures, headache and visual disturbances and showing characteristic parieto occipital predominant FLAIR hyper intensity on MR imaging. We herein report unusual combination of classic atypical CT and MRI findings in a case of PRES syndrome in a 55y male patient who is a known hypertensive, a case of resolving post streptococcal glomerulonephritis and was diagnosed based on clinico radiological correlation

**Keywords:** hypertensive encephalopathy, parieto occipital, magnetic resonance imaging, computed tomography, eclampsia, seizures

### Introduction

Posterior reversible encephalopathy syndrome (PRES) describes a usually reversible neurologic syndrome with a variety of presenting symptoms ranging from seizures to altered mental status. The term describes a potentially reversible imaging appearance and symptomatology that can be caused by hypertension, eclampsia and preeclampsia, immunosuppressive medications such as cyclosporine, various antineoplastic agents, severe hypercalcemia, thrombocytopenic syndromes, Henoch-Schönlein purpura, hemolytic uremic syndrome, amyloid angiopathy, systemic lupus erythematosus, and various causes of renal failure. Etiopathogenesis is thought to be related to a hyperperfusion state, with blood-brain barrier breakthrough, resulting cortical or subcortical edema. In this case report, we present a case of PRES syndrome in adult with a combination of classic and atypical findings and resolution of imaging findings on treatment.

### Case History

A 55 year old male presented with 1 episode of tonic clonic seizure and on checking his vitals, his blood pressure was found to be 170/110 mm hg. Due to low GCS patient was intubated. Patient was first admitted on 10/08/2020 with puffiness of face and swelling of both feet since 10 days. Patient was asymptomatic 10 days back, then he developed puffiness of face gradual in onset, nonprogressive associated with swelling of feet. He is a known hypertensive not on any medication. Patient was evaluated for proteinuria and hematuria. Nephrology consultation revealed diagnosis of post streptococcal glomerulonephritis (resolving) with ASO titre -800 C3 -20 C4-21.

CNS: No focal neurological deficits

Patient was advised CT brain plain on the same day, which revealed ill-defined hypo densities in bilateral parieto occipital lobes predominantly in subcortical region and in high frontal lobes. Acute Subarachnoid bleed in bilateral parieto occipital sulci [figure 1]. For further characterization patient was advised MRI brain which revealed T2/ FLAIR hyper intensities involving bilateral cortical and sub cortical regions of parieto occipital lobes and high frontal and parietal lobes in watershed regions [figure 2] T2/FLAIR hyper intensity with blooming on SWI in subarachnoid space in bilateral parieto occipital sulci - S/O Acute subarachnoid haemorrhage [Figures 3, and 4] Small foci of diffusion restriction in bilateral high parietal lobes [figure 5] MR angiography showed no vascular abnormalities [figure 5] Therefore syndrome.

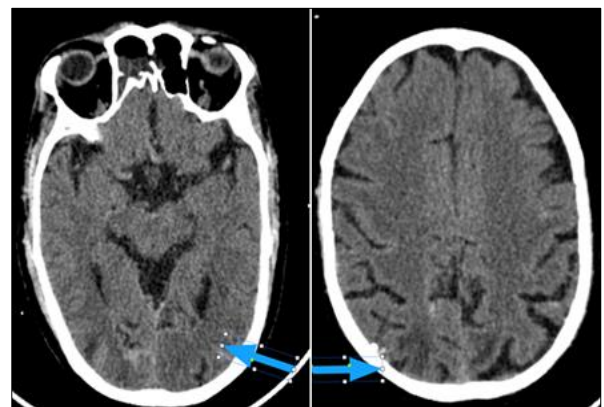


Fig 1

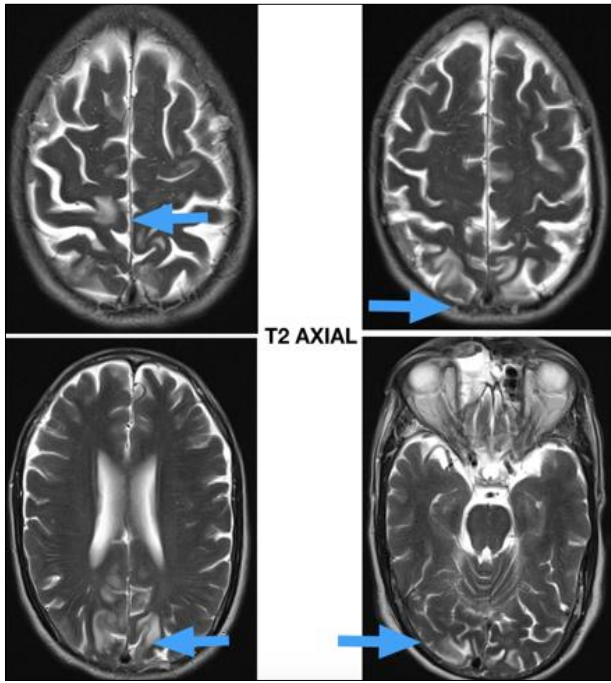


Fig 2

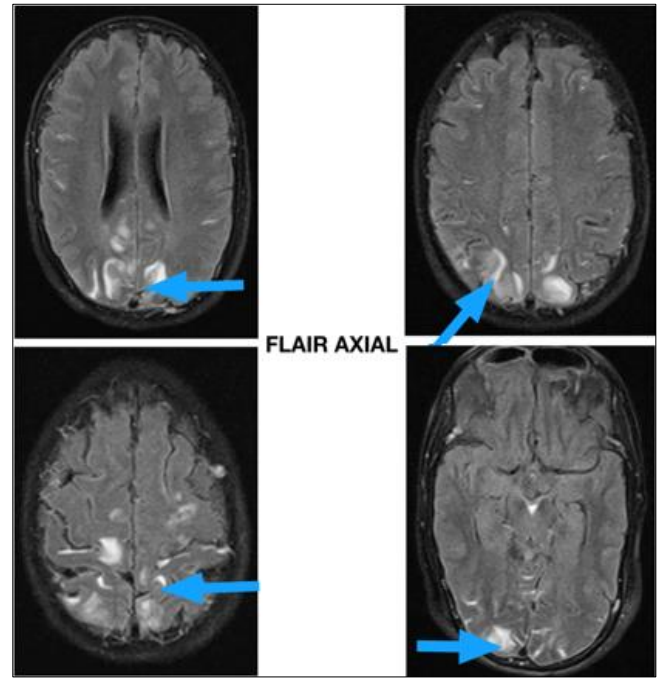


Fig 3

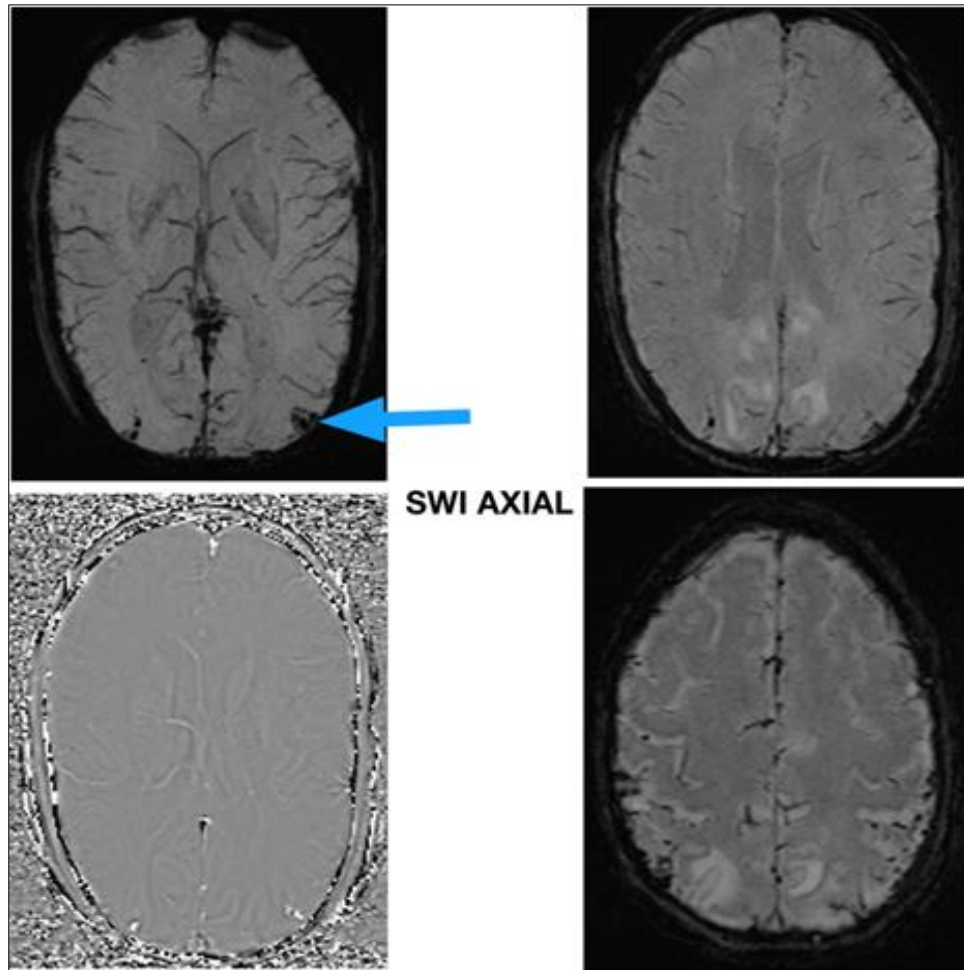


Fig 4

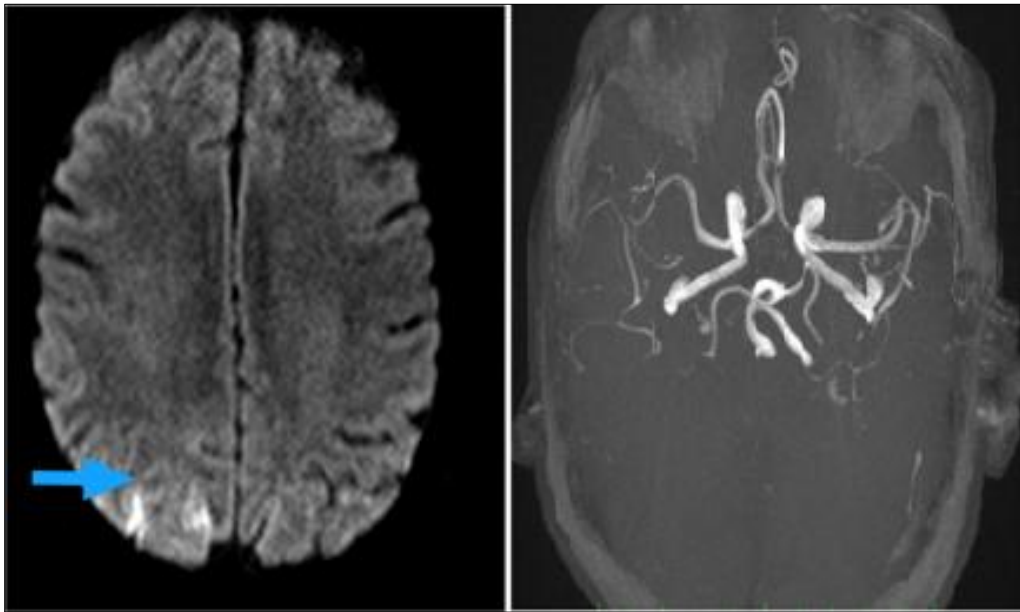


Fig 5

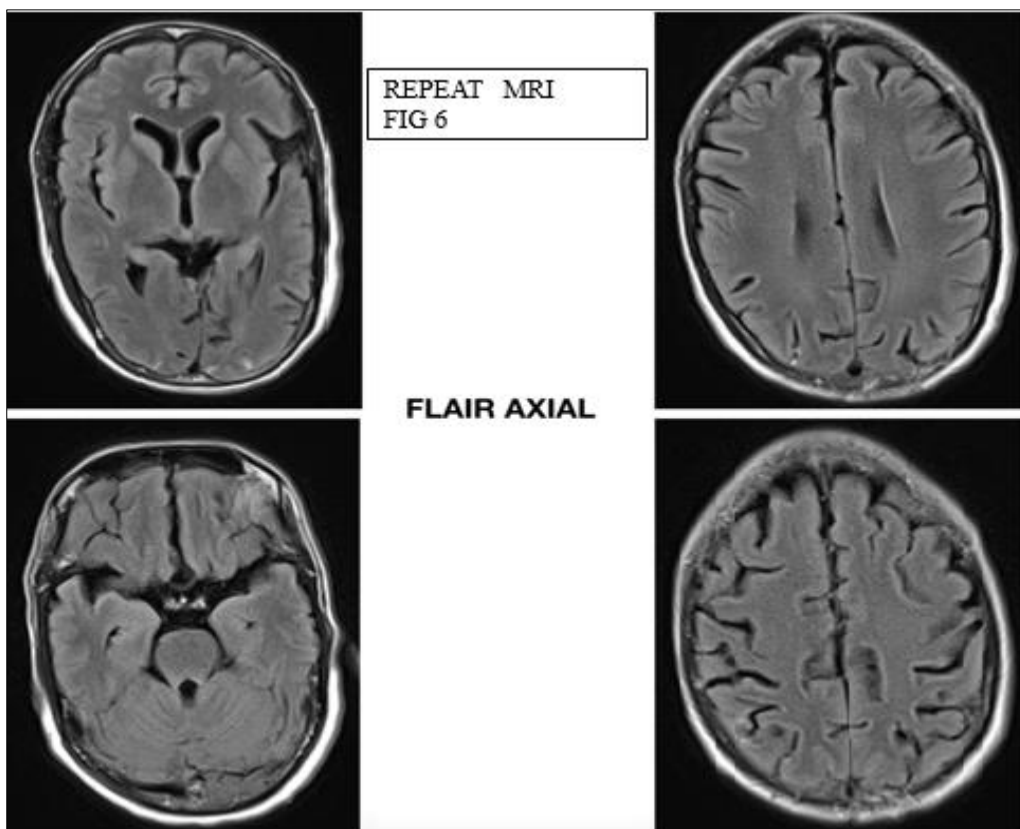


Fig 6

### Case Discussion

Posterior reversible encephalopathy syndrome (PRES) is a common presentation of acute hypertensive encephalopathy. It is identified in patients with pre eclampsia and eclampsia and in patients who underwent organ transplantation. PRES has also been seen in patients with other systemic conditions such as Wegener granulomatosis, hypertension, systemic lupus

erythematosus (SLE), nonspecific renal inflammatory conditions (glomerulonephritis, hepatorenal syndrome), and postchemotherapy<sup>[1, 2]</sup>. Patients typically present with signs such as headache, altered alertness and behavior ranging from drowsiness to stupor, seizures, vomiting, mental abnormalities including confusion, and abnormalities of visual perception. Seizures may begin focally but usually become generalized.

Lethargy and somnolence are among the first signs noted. Patients can present with hemianopia, frank blindness and visual neglect [3]. The term PRES focuses on the similarity in appearance on imaging, in particular, the common location being parietal-occipital lobe or “posterior” location of the lesions [1].

The Etiopathogenesis being: two hypothesis are considered

- Severe HTN leads to vasodilatation, failed cerebral autoregulation and breakthrough hyper perfusion.
- Excessive circulating cytokine → to injury of the micro vascular endothelium, increasing vascular permeability.
- Hydrostatic leakage and extravasation or transudation of fluid and macromolecules through damaged arteriolar walls into the adjacent brain interstitium.
- Vasoconstriction and hypoperfusion leads to brain ischemia and subsequent vasogenic edema [4, 5].

CT/MR imaging, the brain typically demonstrates focal regions of symmetric edema in bilateral cerebral hemispheres. The parietal and occipital lobes are most commonly affected, which is followed by the frontal lobes, the inferior temporal-occipital junction, and the cerebellum [2]. Lesion confluence may develop as the extent of edema increases. MR diffusion-weighted imaging (DWI) was the most important in establishing that the abnormality was vasogenic edema. The edema usually completely reverses. The classic and atypical patterns normally seen are; Dominant parietal pattern [classic] Superior frontal sulcus pattern (involving the mid and posterior aspects of superior frontal sulcus) Holohemispheric watershed pattern [1, 6]. Focal or confluent vasogenic edema was present in the classic “posterior” parietal or occipital lobe region most consistently [2]. W.S. Bartynski and J.F. Boardman *et al* said Vasogenic edema that is typically seen linearly along the superior frontal sulcus seems to define the junction between the anterior cerebral territory and middle cerebral territory, which suggests a distribution between the medial (ACA, PCA) and lateral cerebral branches (MCA) Uncommon locations include Involvement of the temporal lobe basal ganglia (14%), brain stem (13%), and deep white matter (18%), particularly when they are associated with the hemispheric features [1]. Distinct involvement of the splenium (10%) was clearly observed [1]. Most cases of PRES are caused by vasogenic edema, so DWI is usually negative (not cytotoxic).

PRES with restricted diffusion occurs in 15-30% of cases and is usually seen as small foci of restricted diffusion within larger regions of nonrestricting vasogenic edema.

PRES-associated intracranial hemorrhage is uncommon, seen in only 5-15% of cases

1. Focal parenchymal hematoma
2. Multifocal haemorrhage
3. Convexity subarachnoid haemorrhage [7, 8, 9].

Restricted diffusion changes are seen to be potentially reversible. Benziada-Boudour *et al* demonstrated a case of PRES with foci of restricted diffusion that resolved on follow-up imaging with no abnormal imaging sequelae [9]. Catheter angiography can show focal or diffuse vasoconstriction, vasodilation or a “string-of-beads” appearance, which is also seen on CT angiography or MR

angiography. These findings can be confused for other diagnoses such as vasospasm or vasculitis [9, 10]. The intent of this study being to show both the typical and atypical distributions and manifestations of PRES using a variety of CT and MRI sequences. In our study, reversible vasogenic edema was almost always present in the cortical or subcortical white matter of the parietooccipital region, the atypical findings being the involvement of the superior frontal sulcus and high frontal and parietal watershed zones another atypical feature being convexity subarachnoid haemorrhage and foci of diffusion restriction.

## Conclusion

In conclusion, the case we presented suggests that PRES may present with atypical imaging findings and clinical presentation and that associated conditions such as glomerulonephritis gives a clue to diagnosing PRES and that high blood pressure is not always present in a case of PRES. An important criterion to identify PRES is the reversibility of symptoms and radiological findings, but this may occur later than expected.

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