

Posterior Reversible Encephalopathy Syndrome- Distinct Imaging Features on MRI

Ishwar Agarwal¹, D B Dahiphale², Vikas Shinde³

¹Resident, Department of Radiology, MGM Medical College Aurangabad (MS) India.

²Associate Professor and HOD, Department of Radiology, MGM Medical College Aurangabad (MS) India.

³Assistant Professor, Department of Radiology, MGM Medical College Aurangabad (MS) India.

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ABSTRACT

Background: Posterior Reversible Encephalopathy Syndrome (PRES) is rare but severe neurotoxic syndrome of cerebral vaso-regulation. It has varied clinical presentations and diverse patterns of expressions which make diagnosis difficult. Atypical features hereby must be recognized in order to more completely understand this condition in which early treatment can successfully revert the disease process and make a path to good clinical progress. Aim and objective: To identify and evaluate different imaging patterns of PRES on MRI. Common clinical presentation and association with different diseases were also evaluated. **Methods:** The study was done over a period of one year from August 2018 to August 2019 and total 20 patients were included in the study. Patient included in the study were patient who were referred to the department of radiology, MGM medical college and hospital Aurangabad for MRI scan and were diagnosed as PRES on imaging study. Proper history, clinical details, chief complaints and follow up clinical outcome after treatment of the patient were taken. The data were tabulated and observed and conclusion were made. **Results:** In PRES there is symmetrical involvement of parieto-occipital lobes, but asymmetrical involvement of bilateral cerebral hemisphere was also seen in 26% of the patients. Other areas were also involved with involvement of basal ganglia and brain stem were rare. The most common cause for PRES was found to be acute onset hypertension and post-partum epilepsy/ Eclampsia. The most common age group involved is 10-30 years age group. Typical location was involved in 12 patients (63%) of the patient as compared to 7 patients with distribution of vasogenic oedema in atypical location (36.8%). Out of total 19 patients, 4 patients (26%) showed restriction on DW images. Out of total 19 patients only 2 patient showed presence of intraparenchymal hematoma / subarachnoid haemorrhage. **Conclusions:** Though Posterior reversible encephalopathy syndrome presents with classical imaging manifestations of symmetrical parieto-occipital subcortical white matter hyper intensities, radiologists must be aware of atypical sites and presentation so as to be able to properly diagnose this condition.

Keywords: Posterior reversible encephalopathy syndrome, Atypical presentation, Imaging findings, Restricted Diffusion.

1

INTRODUCTION

Posterior reversible encephalopathy syndrome (PRES) was first described by Hinchey and his colleagues in 1996 describes a neurological syndrome characterized by variety of symptoms, including headache, altered mental status, visual disturbances and seizures.^[1,2] The term posterior is usually referred to the most common location of the imaging finding that is parieto-occipital lobe. The term reversible is used because the syndrome has a reversible imaging and clinical findings.^[3] In it there are different unique patterns of vasogenic edema seen on imaging study due to neurotoxicity in this syndrome. Identifying these various patterns of imaging findings is of utmost important for the

The exact mechanism for development of PRES is still not fully understood. There is no single hypothesis to explain the cause vasogenic edema in different clinical settings. There are different hypotheses proposed which together explain the cause of vasogenic edema. The vascular theory of PRES is the most widely acceptable. The posterior circulation has a relatively sparser sympathetic innervations thus weaker auto regulation. Therefore, in cases with acute hypertension, there is increased blood flow in posterior circulation due to poor auto regulation with leakage protein and fluid from the vascular wall leading to vasogenic edema. Even though this theory proposes cerebral hyper perfusion as the cause of imaging abnormalities, some PET studies have actually shown cerebral hypo perfusion in PRES.^[4] Another theory is related to endothelial damage (as in cases of eclampsia) which leads to vasogenic edema.^[5,6] Also increasing blood pressure leads to vasospasm and subsequent infarction leading to vasogenic edema.^[7,8]

PRES may present as Arterial hypertension (this is the classic setting and it was the first factor to be

Name & Address of Corresponding Author

Dr Ishwar Agarwal
Resident,
Department Of Radiology,
MGM Medical College Aurangabad (MS)
India.
radiologist to diagnose it.

described. Moderately to severely elevated blood pressure is reported in 75% of patients) or eclampsia or pre-eclampsia. IN many cases PRES is found to be associated with various chemotherapy regimens, septicemia, renal failure and autoimmune diseases such as systemic lupus erythematosus, scleroderma and Wegner’s granulomatosis.

In pediatric population PRES is found to be associated with risk factors such as, renovascular diseases, immunosuppressive therapy, Haemato-oncologic patients associated with cytostatic drugs and Systemic diseases such as leukemia, aplastic anemia, solid tumors and autoimmune diseases. The common presenting complaints include Generalized seizures, Headaches, confusion, Nausea and vomiting, focal neurological deficit, such as cortical blindness, cerebellar syndrome and Hemiparesis.

We undertook this study to analyze imaging findings in cases of PRES.

MATERIALS & METHODS

This was a prospective study conducted in the department of radiology of a tertiary care medical college. Patients referred to our department for MRI and diagnosed to be having Posterior Reversible Encephalopathy Syndrome were included in this study on the basis of a predefined inclusion and exclusion criteria.

Proper history, clinical details, chief complaints and follow up clinical outcome after treatment of the patient were taken. The diagnosis of PRES was made by same radiologist using the standard radiological criteria for PRES.

The MRI images were studied with particular emphasis on finding out the anatomical site of involvement. Diffusion weighted imaging was done to find out the restricted diffusion pattern on MRI. The statistical analysis was done using SPSS 21.0 software.

RESULTS

Distribution of PRES according to gender.

A total of 19 patients were studied during study period. Out of these 19 cases there were 14 (73.68) females and 5 (26.3) males. There was a female preponderance with a M:F ratio of 1:3.75.

The analysis of age group of the studied cases showed that the most common affected age group was between 11-30 years (42.1%) followed by 31-50 years (36.8%). It was found to be uncommon in patients below 10 years of age (10.5%) and above 50 years (10.5%).

Out of total 19 patients, typical location was involved in 12 patients (63%) of the patient as compared to 7 patients with distribution of vasogenic oedema in atypical location (36.8%).

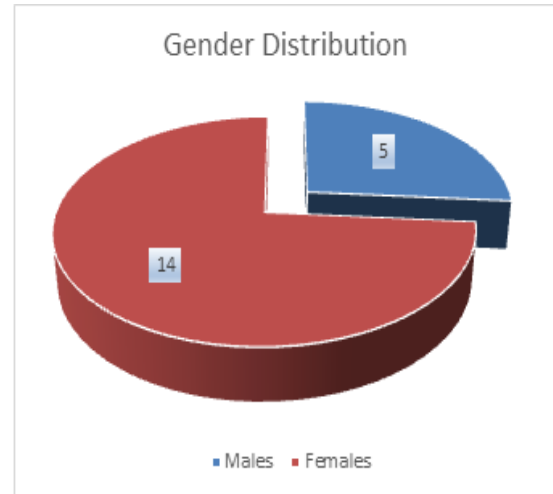


Figure 1: Gender Distribution of studied cases.

Table 1: Age distribution of the studied cases.

| Age group | No of patients | percentage |
|-------------|----------------|------------|
| 0-10 years | 2 | 10.5% |
| 10-30 years | 8 | 42.1% |
| ears | 7 | 36.8% |
| >50 years | 2 | 10.5% |

Table 2: Typical and Atypical Pres Imaging Features

| | No of patient | percentage |
|----------|---------------|------------|
| Typical | 12 | 63.1% |
| Atypical | 7 | 36.8% |

The analysis of imaging features showed that in majority of the patients (94.7%) parieto-occipital lobe was involved. The other common locations were found to be frontal lobe (78.9%) temporal lobe (47.3%) and cerebellum (42.1%). Thalamus (15.7%), brainstem (15.7%) and basal ganglia (21.05%) were less commonly involved.

Table 3: Anatomical site of involvement in studied cases.

| Location | No of patients | Percentage |
|---------------------------|----------------|------------|
| Typical parieto-occipital | 18 | 94.7% |
| Frontal | 15 | 78.9% |
| Temporal | 9 | 47.3% |
| Cerebellum | 8 | 42.1% |
| Thalamus | 3 | 15.7% |
| Brainstem | 3 | 15.7% |
| Basal ganglia | 4 | 21.05% |

On Diffusion weighted imaging restricted diffusion was seen in 4 (26.6%) patients whereas restricted diffusion was absent in 15 (78.9%) patients.

Table 4: Diffusion restricted imaging in studied cases.

| Restriction on DW images | No of patient | percentage |
|--------------------------|---------------|------------|
| Present | 4 | 26.6% |
| Absent | 15 | 78.9% |

Out of total 19 patients only 2 patient showed presence of intraparenchymal hematoma / subarachnoid haemorrhage.

Table 5: Focal Hematoma/ SAH in studied cases.

| Focal Hematoma / SAH | No of Patient | Percentage |
|----------------------|---------------|------------|
| Present | 2 | 10.5% |
| Absent | 17 | 89.5% |

The analysis of presence of etiological factors showed that acute hypertension was the most frequent cause of PRES in our study which was seen in 6 (31.5%) patients. The other common etiological factors were found to be postpartum epilepsy and eclampsia (26.3%) and convulsions (21%). Chemotherapy (10.5%) and trauma (10.5%) were the associated factors in comparatively a smaller number of cases.

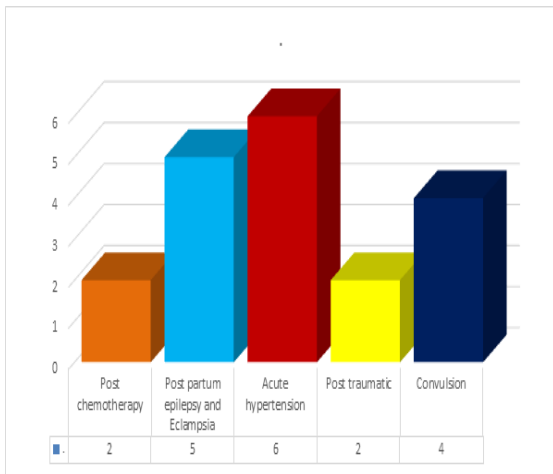


Figure 2: Etiological Factors in studied cases.

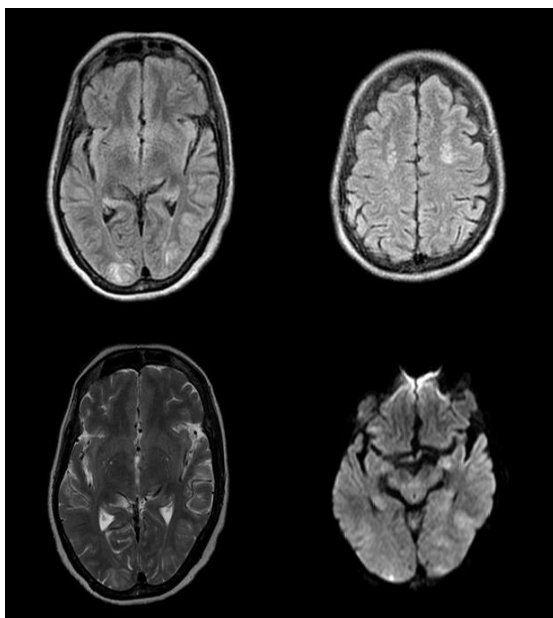


Figure 3: A 65-year-old female, came with the complain of drowsiness following sudden onset increase in blood pressure. MRI finding suggest hyperintense signal intensity in cortical and subcortical region of bilateral front parietal lobe suggestive of vasogenic edema. It also shows foci of restricted diffusion on DW images in bilateral occipital lobe

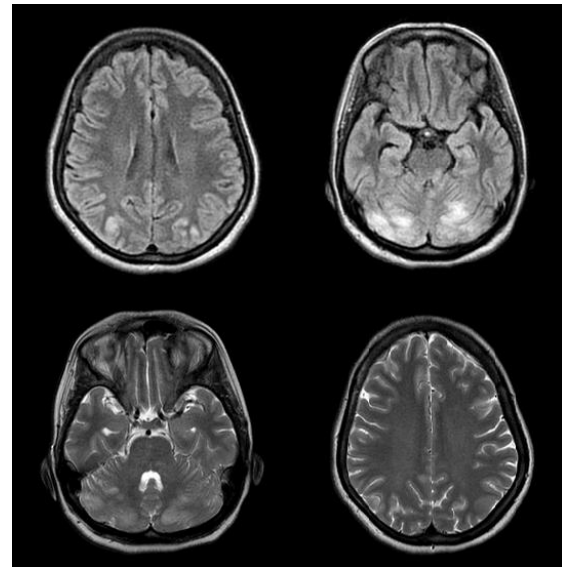


Figure 4: A 32-year-old female presented with post-partum headache and diminution of vision. MRI scan was performed and T2 and FLAIR hyperintense signal intensity was noted in bilateral parietal and occipital lobes.

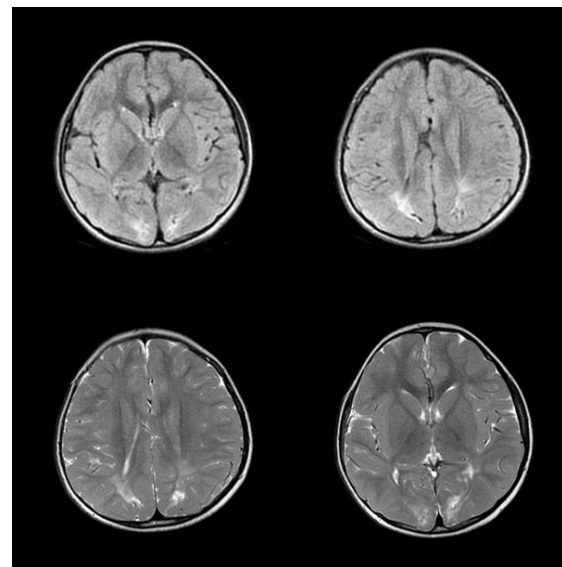


Figure 5: A 5-year-old boy, a known case on B –ALL presented with drowsiness, irritability following chemotherapy. MRI scan was performed, T2 and FLAIR hyperintense signal intensity was noted in bilateral fronto-parietal lobes.

DISCUSSION

From the above study we were able to know the different imaging findings seen in posterior reversible encephalopathy syndrome. As the name suggestive the imaging findings does not only involve the posterior cerebral region and findings are seen in other regions as well. Imaging findings can involve the typical locations such as parieto-occipital region, frontal, temporal region and cerebellar hemisphere or can involve uncommon location such

as brain stem, basal ganglia and corpus callosum.^[9-11]

In this study we saw that out of total 19 patients, 12 patient (63%) had imaging findings in typical locations and 7 had findings in atypical locations. This is comparable to study done by W.S. Bartynski and J.F. Boardman in which atypical location was seen in 26% of the cases. In the study the most common region involved in parieto-occipital region (94%) followed by frontal (78%). Less commonly involved areas are the brain stem, basal ganglia and corpus callosum. This was comparable to a study done by McKinney et al were also the most common region involved was parieto-occipital followed by frontal.^[12-14]

Most of the patient shows symmetrical involvement of bilateral cerebral hemisphere but asymmetrical involvement is also common. In our study about 15 patients had symmetrical involvement (78.9%) and 21.1 % of the patient had asymmetrical involvement (5 patients) which is comparable with a study done by W.S.Bartynski and J.F.Boardman in which asymmetrical involvement was seen in 27% of the patients.^[14]

Restricted diffusion is considered among the atypical findings of PRES. In patients with PRES some may also give restricted diffusion on DW images. In this study about 26% of the patient had restricted diffusion on DW images. This finding was comparable with a study done by McKinney et al. in which 28% of the patient had restricted diffusion.¹²In the study we found that the PRES has a slightly female predilection. In the study we found that 15 out of the 19 patients were female. It is mostly due to common occurrence of PRES in patients of eclampsia.

Most common presenting complain of the patient with PRES was found to be acute hypertension and eclampsia involving approximately 31% and 26% of the patients. Less common cause were found to be post chemotherapy (10.5%) and post traumatic (10.5%). The most common location to be involved was found to parieto-occipital lobe seen in approximately 94% of the patients followed by frontal and temporal lobes. These findings are comparable to study done by Jennifer E. Fugate et al. The least common site to be involved were brain stem, basal ganglia and thalamus.^[15]

Intraparenchymal bleed or subarachnoid haemorrhage can also be seen in patients with PRES. These findings are included in atypical findings of PRES and are not seen commonly. In this study out of 19 patients 2 patients had intraparenchymal bleed.

CONCLUSION

- Posterior reversible encephalopathy syndrome presents with classical imaging manifestations of

symmetrical parieto-occipital subcortical white matter hyper intensities.

- Other locations such as the frontal lobes, basal ganglia, brainstem and cerebellum may also be involved.
- The radiologists should be aware of the atypical manifestations such as unusual locations, restricted diffusion, hemorrhage and unilateral involvement so that diagnosis could be made and early treatment could be initiated.

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