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CLINICAL, IMAGING SPECTRUM AND OUTCOME OF POSTERIOR REVERSIBLE ENCEPHALOPATHY SYNDROME

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Abstract

Background: The posterior reversible encephalopathy syndrome (PRES), also known as acute hypertensive encephalopathy and reversible leukoencephalopathy syndrome (RPLS), is a clinico-radiological syndrome characterised by white matter vasogenic edema primarily affecting the parietal and occipital lobes of the brain and characterised by rapid onset of headache, seizures, loss of consciousness, and visual loss. The imaging findings, on the other hand, can be found in a variety of places, including the frontal lobes, thalami, basal ganglia, and brainstem.

Objective: To study the clinical, imaging spectrum and final disease outcome of PRES.

Materials And Methods: The study included 104 participants who had clinical symptoms and a radiological diagnosis of PRES. Each patient's clinical, imaging, and outcome data were examined.

Results: The study included 104 individuals ranging in age from 10 to 65 years old. Seizures were the most common symptom, followed by headache (60 percent), vomiting (37 percent), visual disturbances (33 percent), altered sensorium (15 percent), thalamic aphasia (6 percent), hemiparesis (4 percent), paresthesia (4 percent), ataxia (4 percent), quadriparesis (4 percent), quadriparesis (4 percent), and facial numbness (2 percent). The frontal lobe (52 percent), temporal lobe (17 percent), cerebellum (27 percent), thalamus (15 percent), brainstem (13 percent), basal ganglia (10 percent), and corpus callosum (10 percent) were all involved on MR imaging in the majority of the cases (2 percent). Except in one case of central PRES, all lesions in atypical areas had lesions in usual locations. In 32% of instances, the dominant parieto-occipital pattern was detected, followed by the superior frontal sulcus pattern in 27%, holohemispheric pattern in 35%, and partial or asymmetric expression of primary patterns in 5%. Only about a quarter of the time, diffusion limitation and post-contrast enhancement were seen. In 91 percent of the cases, the symptoms were completely resolved, however four percent of the patients died. Symptoms persisted in 6% of cases after being followed up on.

Conclusion: PRES is a clinico-radiological illness that has a wide range of clinical and imaging manifestations. Atypical regions are frequently included. Lesions in atypical places, on the other hand, frequently have lesions in typical locations as well. Restricted diffusion and contrast enhancement are two examples of unusual imaging properties. Clinicians and radiologists must be aware of atypical lesion locations and imaging findings in order to correctly diagnose PRES in the appropriate clinical situation.

Keywords: Posterior reversible encephalopathy syndrome, imaging, reversible leukoencephalopathy syndrome

INTRODUCTION

Acute hypertensive encephalopathy (AHE) and reversible leukoencephalopathy syndrome (RPLS) are two clinico-radiological syndromes that manifest with headache, seizures, decreased consciousness, visual abnormalities, nausea, vomiting, and focal neurological symptoms[1]. Hypertension, pre-eclampsia, and eclampsia, renal failure, systemic lupus erythematosus, usage of certain immunosuppressive medications, thrombocytopenic purpura, and hemolytic uremic syndrome are all linked to it[2,3]. Other links have lately been proposed, including infection, sepsis, and shock. The normal imaging feature is bilateral and symmetrical vasogenic oedema covering the cortical and subcortical regions of the occipital and parietal lobes, while some patients have atypical distributions involving the anterior cerebral lobes, brain stem, cerebellum, and basal ganglia[4,5]. On MRI, atypical imaging features such as contrast enhancement, haemorrhage, and limited diffusion may be visible. As clinical and radiological expertise with PRES accumulates, certain unusual clinical and imaging signs are becoming more well recognised, and understanding these traits is critical for proper diagnosis and therapy[6]. The clinical, imaging, and outcome characteristics of PRES are described in this article.

MATERIAL AND METHODS

The current prospective longitudinal observational study lasted two years and took place in a tertiary care centre. The study comprised patients with PRES-like clinical and imaging characteristics. For each patient, a detailed clinical history was collected, as well as a comprehensive general physical, systemic, and neurological examination. A predesigned proforma was used to record all clinical and radiological data. On a 1.5 T MR machine, all patients had T1-weighted, T2 weighted, T2 FLAIR, susceptibility weighted, diffusion-weighted, contrast enhanced T1-weighted imaging sequences. On T2 weighted and T2 FLAIR images, patterns of lesion distribution in anatomical regions were found. Other features were detected, such as restricted diffusion on DWI, bleeding on SWI, and contrast enhancement. Cases were divided into four categories based on the patterns found in the literature: dominant parietooccipital pattern, superior frontal sulcus pattern, holohemispheric watershed pattern, and incomplete or asymmetric expression of the major pattern. In the cases that were available, follow-up imaging was included. Each patient's outcome was recorded.

RESULTS

We prospectively monitored 104 PRES patients with complete clinical and MRI data in order to describe the clinico-radiological profile and prognosis in a thorough manner in the current study. The study included 104 patients, 22 of whom were males and 82 of them were girls. The participants ranged in age from under 10 to 65 years old. The majority of the patients (68%) were between the ages of 21 and 30, followed by the groups of 31–40 years, 16 (15%), and 11–20 years 4 (4 percent). There were six patients under the age of ten (six percent) and ten patients beyond the age of fifty (ten percent) (10 percent).

Table 1: Predisposing factors of PRES in our study

Predisposing condition	Number of patients	Percentage
Eclampsia	74	71%
Chronic kidney disease	10	10%
Chemotherapy	6	6%
Post renal transplant	4	4%
Systemic lupus erythematosus	4	4%
Nephrotic syndrome	4	4%
IgA nephropathy	2	1%

Predisposing circumstances included eclampsia in 74 (71%) patients, CKD in 10 (10%) patients, chemotherapy in 6 (6%) patients, post renal transplant in 4 (4%) patients, SLE on Rituximab treatment in 4 (4%) patients, Nephrotic syndrome in 4 (4%) patients, and IgA nephropathy in 2 (2%) patients (Table 1). 64 of the 74 patients with eclampsia were in their first pregnancy, whereas 10 were in their second.

Table 2: Clinical features of PRES in our study

Clinical features	Number of patients	Percentage
Seizures	88	85%
Headache	62	60%
Vomitings	38	37%
Visual disturbances	34	33%
Altered sensorium	16	15%
Thalamic aphasia	6	6%
Hemiparesis	4	4%
Paresthesia	4	4%
Ataxia	4	4%
Quadriparesis	4	4%
Facial numbness	2	1%

Seizures were the most common symptom in our study (85 percent). The most common kind was generalised tonic clonic (57 percent). In 17% of cases, partial seizures were observed, and in 10% of cases, status epilepticus was observed. Other clinical characteristics such as headache (60 percent), vomiting (37 percent), visual abnormalities (33 percent), altered sensorium (15 percent), thalamic aphasia (6%), hemiparesis (4%), paresthesia (4%), ataxia (4%), quadriparesis (4%), and facial numbness (1%), were also reported in our study (Table 2). The parietal and occipital lobes were implicated in 96 percent and 94 percent of the patients, respectively, on MR imaging. In our analysis, lesions with unusual locations were not rare. Atypical lesion locations were seen in the frontal lobe (52%), temporal lobe

(17%), cerebellum (27%), thalamus (15%), brainstem (13%), basal ganglia (10%), and corpus callosum (10%). (1 percent). Except in one case of central PRES in which there was involvement of the midbrain, pons, and thalami with no cortical or subcortical edoema of the cerebrum, all of these cases displayed alterations in the normal places such as the parietooccipital white matter of bilateral cerebral hemispheres. In 32% of instances, the dominant parieto-occipital pattern was detected, followed by the superior frontal sulcus pattern in 27%, holohemispheric pattern in 35%, and partial or asymmetric expression of primary patterns in 5%. Restricted diffusion was found in 22 cases (21%) in our investigation. In the midst of significant vasogenic edoema, areas of limited diffusion were visible. In 14 (14%) of the cases, a punctate focus with restricted diffusion was observed, and in 10 (10%) of the cases, a focal gyral configuration was observed. In 17 percent of the cases, contrast enhancement was observed. In 54 individuals, a follow-up MRI was available. Reversibility of lesions was observed in 48 of these patients. In 18 of the 22 cases with restricted diffusion, follow-up was available. Lesions were resolved in 12 cases, but they were not resolved in 6 others. Complete symptom relief was achieved in 94 of the 104 individuals. On the fourth and fifth days of their hospitalisation, four individuals (4%) died. Neurological symptoms persisted in 3 (6%) of the individuals (one with seizures, one with hemiparesis, one with visual disturbances).

DISCUSSION

The clinical and imaging spectrum of PRES, as well as its outcome, are described in our work. According to the literature, PRES is a diverse condition with a wide range of clinical and radiological characteristics. The fact that eclampsia accounted for 71 percent of all cases in our analysis likely explains the female preponderance[7,8]. The most prevalent form of PRES was seen in primigravida. PRES was seen in a wide range of illnesses and predisposing factors in our study, including eclampsia in 71% of patients, CKD in 10%, chemotherapy in 6%, post renal transplant in 4%, SLE on Rituximab treatment in 4%, Nephrotic syndrome in 4%, and IgA nephropathy in 1% of patients[9]. Seizures were the most common symptom in our study (85 percent). The most prevalent tonic clonic kind was generalised tonic (57 percent of patients). In 17% of cases, partial seizures were observed, and in 10% of cases, status epilepticus was observed. In their investigation, Bartynski WS et al. found that seizures were the most common symptom in 71% of cases. Seizures were likewise the most prevalent symptom in 87 percent of cases, according to Lee VH et al. According to the literature, generalised tonic clonic seizures were detected in 54–64 percent of patients, partial seizures in 3–28%, and status epilepticus in 3–17% of cases[10]. Headache (60 percent), vomiting (37 percent), visual disturbances (33 percent), altered sensorium (15 percent), thalamic aphasia (6 percent), hemiparesis (4 percent), paresthesia (4 percent), ataxia (4 percent), quadriparesis (4 percent), and facial numbness (1 percent) were all noted in our research. The imaging manifestations of PRES can be varied, and they can occur in unusual places. The parietal and occipital lobes were implicated in 96.1 percent and 94.2 percent of the cases in our study, respectively. In 52 percent and 17 percent of instances, the frontal and temporal lobes were affected, respectively. The majority of the atypical region engagement happened in the centre zones (such as the basal ganglia, thalami, brainstem, basal ganglia and corpus callosum). In our study, there were some similar locations but differing incidences when compared to earlier studies; this could be owing to the various sample sizes and populations. Except in two cases (1 percent) of central PRES in which there was involvement of the midbrain, pons, and thalami with no cortical or subcortical edoema of the cerebrum, all of these cases had alterations in the normal regions such as the parietooccipital white matter of both brain hemispheres. McKinney et al. found that 4% of 124 individuals with PRES had MRI results of a "central variant" PRES, with involvement of the brainstem or deep grey nuclei but no involvement of the cerebral hemispheres[11]. They found that thalami were present in all five PRES patients with MR findings consistent with the central form, but that the posterior limb of the internal capsule, cerebellum, and periventricular white matter were all implicated in varying degrees. According to Bartynski WS et al., 22 percent of people have a dominant parieto-occipital pattern, 27 percent have a superior frontal sulcus pattern, 23 percent have a holohemispheric pattern, and 28 percent have partial or asymmetric expression of primary patterns. In our investigation, the dominant parieto-occipital pattern was seen in 32% of the cases, the superior frontal sulcus pattern was shown in 27%, the holohemispheric pattern was seen in 35%, and partial or asymmetric expression of primary patterns was seen in 5% of the cases. In 17 percent of instances, McKinney et al. found restricted diffusion, 17 percent bleeding, and 38 percent augmentation. Diffusion limitation, haemorrhage, and contrast enhancement were described in 30 percent, 22 percent, and 25 percent of patients, respectively, by Saurabh Bansal et al. Restricted diffusion was found in 22 cases (21.1%) in our investigation. In the midst of significant vasogenic edoema, areas of limited diffusion were visible[11,12]. Punctate restricted diffusion foci were observed in 14% of cases, and focal gyral structure was seen in 10% of instances. In our investigation, haemorrhage and contrast enhancement were detected in (14%) and (17%) patients, respectively. In 27 individuals, a follow-up MRI was available. Of this reversibility of lesions was seen in 24 patients.

CONCLUSION

PRES is a clinico-radiological illness that has a wide range of clinical and imaging manifestations. Atypical regions are frequently included. Lesions in atypical places, on the other hand, frequently have lesions in typical locations as well. Restricted diffusion and contrast enhancement are two examples of unusual imaging properties. Clinicians and

radiologists must be aware of atypical lesion locations and imaging findings in order to correctly diagnose PRES in the appropriate clinical situation.

REFERENCES

1. Hinchey J, Chaves C, Appignani B, Breen J, Pao L, Wang A, et al. A reversible posterior leukoencephalopathy syndrome. N Engl J Med (1996)334:494-500.
2. Onder AM, Lopez R, Teomete U, et al. Posterior reversible encephalopathy syndrome in the pediatric renal population. Pediatr Nephrol (2007)22:1921–9.
3. Datar S, Singh T, Rabinstein AA, Fugate JE, Hocker S. Long-term risk of seizures and epilepsy in patients with posterior reversible encephalopathy syndrome. Epilepsia (2015) 56:564–8.
4. Kastrup O, Gerwig M, Frings M, Diener HC. Posterior reversible encephalopathy syndrome (PRES): electroencephalographic findings and seizure patterns. J Neurol. (2012) 259:1383–9.
5. Sha Z, Moran BP, McKinney AM IV, Henry TR. Seizure outcomes of posterior reversible encephalopathy syndrome and correlations with electroencephalographic changes. Epilepsy Behav (2015) 48:70–4.
6. Karampekios SK, Contopoulou E, Basta M, et al. Hypertensive encephalopathy with predominant brain stem involvement: MRI findings. J Hum Hypertens (2004)18:133-4.
7. Choh NA, Jehangir M, Rasheed M, et al. Involvement of the cervical cord and medulla in posterior reversible encephalopathy syndrome. Ann Saudi Med (2011)31:90-2.
8. Chen TY, Lee HJ, Wu TC, et al. MR imaging findings of medulla oblongata involvement in posterior reversible encephalopathy syndrome secondary to hypertension. AJNR Am J Neuroradiol (2009) 30:755-7.
9. Ono Y, Manabe Y, Hamakawa Y, et al. Localized lesions on MRI in a case of hypertensive brainstem encephalopathy. Intern Med (2005) 44:1002-5.
10. Ekawa Y, Shiota M, Tobiume T, et al. Reversible posterior encephalopathy syndrome accompanying eclampsia: correct diagnosis using preoperative MRI. Tohoku J Exp Med (2012)226:55-8.
11. Schwartz RB, Jones KM, Kalina P, et al. Hypertensive encephalopathy: findings on CT, MR imaging and SPECT imaging in 14 cases. AJR (1992) 159:379–383
12. Schwartz RB, Bravo SM, Klufas RA, et al. Cyclosporine neurotoxicity and its relationship to hypertensive encephalopathy: CT and MR findings in 16 cases. AJR (1995) 165:627–631.