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Case Report

Posterior Reversible Encephalopathy Syndrome (PRES) in a Patient with Chronic Kidney Disease (CKD)

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ABSTRACT

A 24 year male known case of chronic kidney disease (CKD) presented with headache and seizure. MRI Brain showed hypointense lesions on T1 and hyperintense lesion on T2 weighted images in bilateral occipital and cerebellum region suggestive of Posterior reversible encephalopathy syndrome (PRES) which is a clinico-radiological entity with characteristic features on neuro-imaging and clinical symptoms. This condition is labelled by a variety of names like reversible posterior leukoencephalopathy syndrome, reversible posterior cerebral oedema syndrome and reversible occipito-parietal encephalopathy.

Keywords: Posterior reversible encephalopathy syndrome, seizure, chronic kidney disease, posterior leukoencephalopathy.

INTRODUCTION

Posterior reversible encephalopathy syndrome (PRES) is characterized by seizure, impairment of consciousness, headache, visual abnormalities, nausea, vomiting and focal neurological deficits, which are potentially reversible. The cerebral imaging abnormalities are often symmetric and predominate in posterior white matter. [1] The term PRES describes a potentially reversible imaging appearances and may occur in diverse situations including hypertension, eclampsia, preeclampsia, immunosuppressive medications such as cyclosporine, various anti-neoplastic agents, severe hypercalcemia, renal failure, post transplantation, infection, sepsis shock, Henoch -Scholin purpura (HSP), haemolytic uremic syndrome (HUS), amyloid angiopathy, Systemic lupus erythematous (SLE). [2] The findings on neuroimaging in PRES include non-enhancing white matter abnormalities that appear as areas of low attenuation on CT scan and hypointense on T1-weighted imaging and hyperintense on T2- weighted imaging on MRI. The lesions are mainly seen in posterior region of cerebral hemispheres. These abnormalities partially or completely resolve on follow up scanning, suggesting subcortical oedema without infarction. [3] Here we report a case of chronic kidney disease (CKD) with accelerated hypertension, presented with

generalised tonic clonic seizure and found to have posterior reversible encephalopathy syndrome (PRES) confirmed by neuro-imaging.

CASE REPORT

A 24 year male presented to emergency department with nausea, headache and new onset generalised tonic clonic seizure. He was known case of chronic kidney disease (CKD) on twice weekly maintenance haemodialysis (renal replacement therapy). For last 2 weeks he didn't undergo haemodialysis. On general examination blood pressure was 230/120 mm of Hg at the time of admission. Fundoscopy revealed grade-II hypertensive retinopathy. Neurologically patient was in postictal confusion with no focal neuro-deficites. CT brain imaging showed ill defined hypodensities in the bilateral cerebellar hemisphere and occipital lobes. **[Figure no.1]** MRI brain showed areas of altered signal intensity noted in bilateral occipital gyri and cerebellum which were hypointense on T1 and hyperintense on T2/FLAIR with no area of blooming on gradient recalled echo (GRE) features suggestive of PRES. Hyperintense lesions were noted in basal ganglia, cerebral peduncle bilaterally on T1 and hypointense on T2/FLAIR with no blooming on GRE suggestive of encephalopathy. **[Figure no 2]** His previous ultrasound abdomen was suggestive of right kidney atrophic and grade-2 medical renal disease (MRD) in left kidney. On admission serum urea: 170 mg/dl, serum creatinine: 18 mg/dl, sodium (Na^+) 143 mEq/l and potassium (K^+): 5.4 mEq/l. Hemoglobin: 9.5 gm%, Total leucocyte count (TLC): 12130 /cu mm and platelet count: 2.85 lac.

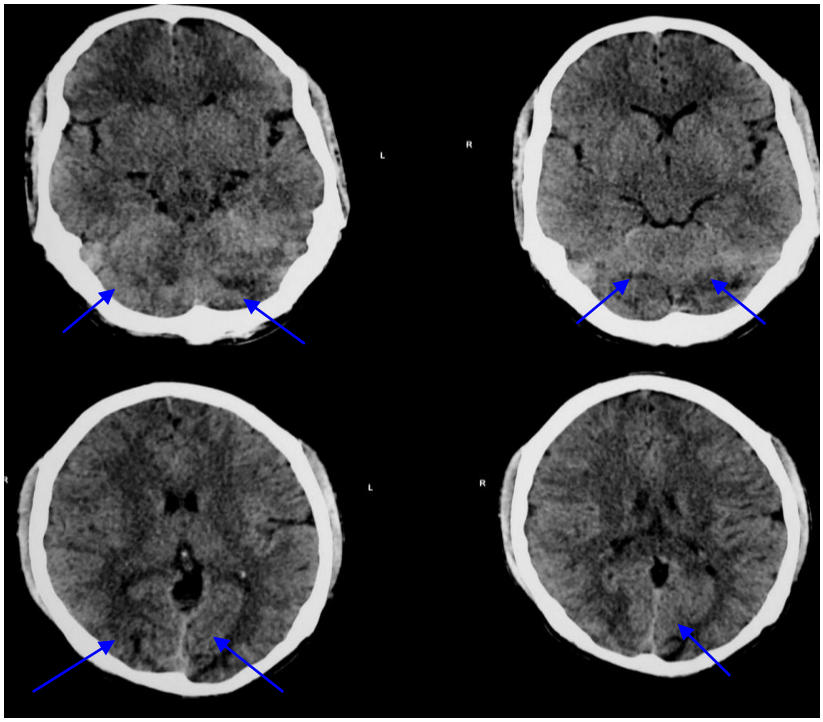

Figure no.1: CT Brain Showing cerebral edema with effaced sulcal spaces (arrow) predominantly in cerebellar hemispheres and occipital region with multiple hypodense lesion (arrow) in same region.

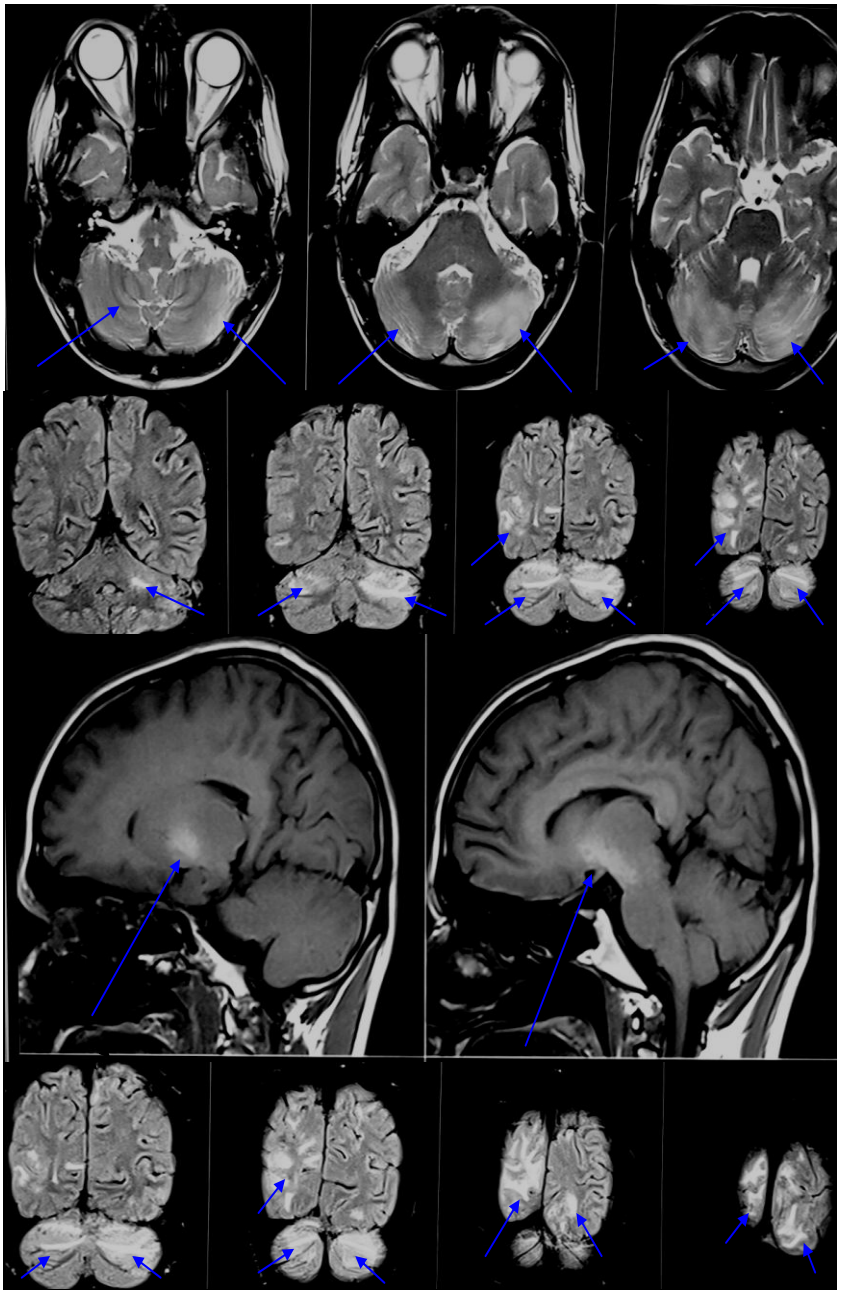

Figure no. 2: MRI Brain showed areas of altered signal intensity noted in bilateral occipital gyri and cerebellum which were hypointense on T1 and hyperintense on T2/FLAIR with no area of blooming on gradient recalled echo (GRE) features suggestive of PRES. Hyperintense lesions were noted in basal ganglia, cerebral peduncle bilaterally on T1 and hypointense on T2/FLAIR with no blooming on GRE suggestive of encephalopathy.

DISCUSSION

The typical features of PRES consist of impairment of consciousness, seizures, headache, visual abnormalities, nausea, vomiting and focal neurological signs. Impairment of consciousness may range in severity from confusion, somnolence and lethargy to encephalopathy or coma. [1] Our patient presented with headache, seizure and accelerated hypertension**.** A frequently associated sign, acute hypertension is not usually described among the main signs of PRES; however hypertension has been reported in most studies in 67% to 80% of

patients. PRES was believed to consistently produce bilateral and symmetric regions of oedema typically located in the white matter and predominating in posterior parietal and occipital lobes.The topographic distribution of Radiological patterns reported in cohort studies is given as [4-6] a) Holo-hemispheric watershed pattern: a swath of confluent vasogenic oedema extends through frontal, parietal and occipital lobes . Involvement of temporal lobes is less marked this matches to watershed zone between anterior and posterior cerebral arteries on one hand and middle cerebral artery on other hand b) Superior frontal sulcus pattern: patchy oedema predominant in frontal lobes along superior frontal sulci c) Dominant parieto – occipital pattern d) Partial expression of three primary patterns. [7-9] The exact pathophysiology of PRES remains unknown. However, three hypotheses exist that may explain the radiological findings in PRES and are related to disorganized cerebrovascular autoregulation, endothelial dysfunction and vasospasm related to acute increases in blood pressure. Sudden elevation of systemic blood pressure causes cerebrovascular autoregulation failure and breakdown of the blood brain barrier, resulting in hyperperfusion. This loss of autoregulation more prominently affects the parieto-occipital regions because there is decreased sympathetic innervation in the posterior cerebral arterial circulation. [10] The loss of cerebral autoregulation leads to arteriolar vasodilation and endothelial dysfunction, resulting in transudation of fluids and proteins, causing cerebral vasogenic edema. Moreover, cerebral vasospasm can lead to hypoxia and cytotoxic ischemia leading to edema predominantly found in the parietal-occipital regions and watershed areas of the brain. [11] In the setting of renal failure, azotemia may cause interstitial brain edema by increasing the permeability of capillaries and cytotoxic edema by direct injury (endothelial dysfunction) of the brain parenchyma. [12] The overall occurrence of PRES is currently unknown. However, a retrospective study of 36 patients with PRES reported 45% (17 patients) had the comorbidity of renal failure, making renal failure a common condition found in patients diagnosed with PRES. PRES can be a complication of dialysis-dependent patients, but it is due to uremia that dialysis-dependent patients develop as opposed to dialysis disequilibrium syndrome that occurs within hours of treatment. [13] A number of medications have been implicated in inducing PRES, including erythropoietin which is commonly prescribed to CKD patients to treat the associated normocytic anemia. [14] Erythropoietin works by stimulating the bone marrow to release more reticulocytes [15] and known to induce or exacerbate hypertension in end stage renal disease patients. [16] This worsening of BP may play role in cerebral dysregulation and ultimately PRES. Medication withdrawal has also been implicated in the development of PRES. Nakabou and colleagues report a case of PRES following the discontinuation of antihypertensive medication in a man with ESRD, [17] and resolution of symptoms when antihypertensives were restarted. Clonidine, which can cause a rebound hypertension if discontinued quickly, [18] has also been implicated in PRES. [19] Patient in case report presented with renal failure (chronic kidney disease), hypertension and seizures. MRI Brain suggestive of PRES supported by follow up scan, responded well to supportive line of management in the form of antihypertensive, antiepileptic drugs with thrice a week haemodialysis and metabolic correction.

CONCLUSIONS

The diagnosis of PRES is challenging as other neurological conditions

shows similar clinico-radiological findings. Prompt recognition of this condition is critical, to ensure reversibility of the vasogenic edema as seen in present case report. The underlying mechanism of PRES, in our patient, could be endothelial dysfunction caused by renal failure added by accelerated phase of hypertension. PRES should be considered in patients who developed neurological manifestations in the form of altered sensorium, seizures, unconsciousness and or focal neuro-deficites in patient with hypertension with renal failure.

Conflict of interest: nil
Support: nil

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