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Recurrent Posterior Reversible Encephalopathy Syndrome in Chronic Kidney Disease

KEYWORDS : Posterior Reversible Encephalopathy Syndrome, PRES, Chronic Kidney Disease, Peritoneal Dialysis

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ABSTRACT *Posterior reversible encephalopathy syndrome (PRES) is a clinical syndrome of encephalopathy, headache, visual disturbance, and seizures. One essential feature of PRES is the presence of reversible cerebral vasogenic edema that has a predominantly posterior distribution on brain imaging. In most cases, both clinical and radiological findings are reversible, although permanent imaging abnormalities and residual neurological sequelae can be seen in a minority of patients. Treatment of hypertension and seizures, and withdrawal of causative agents are the mainstays of therapy in PRES. Chronic kidney disease patients may be especially vulnerable to this syndrome because they are frequently exposed to several of its possible causes, including uremia and hypertension. In its most severe form, PRES can manifest clinically as seizures, coma or death. However, if properly diagnosed and treated, this syndrome can be completely reversible. We report the case of a very young hypertensive chronic kidney disease patient, brought to the emergency room with headache and left tonic-clonic seizures; the cerebral magnetic resonance imaging features were impressive. Anti-hypertensive therapy and Peritoneal Dialysis allowed complete recovery including complete resolution of follow-up MRI. Recurrence though uncommon was observed in the child who was now on Continous Ambulatory Peritoneal Dialysis after 5 months of initial episode. Timely diagnosis and therapy led to complete reversal of PRES and therefore it should be kept in mind in the differential diagnosis of consciousness impairment, seizure activity, headache or visual abnormality in chronic kidney disease patients. Prompt correction of the cause is crucial to decrease the risk of ischemia or bleeding and therefore to avoid permanent disability or death.*

INTRODUCTION

Posterior reversible encephalopathy syndrome (PRES), first described by Hinchey *et al*. in 1996,1 is a clinical condition presenting with neurological symptoms including headache, seizures, altered sensorium and loss of vision accompanied by characteristic magnetic resonance imaging (MRI) findings which are potentially reversible.

Over the years, this condition has been described by various names including reversible posterior leukoencephalopathy, reversible posterior cerebral oedema, reversible occipitoparietal encephalopathy, and hypertensive encephalopathy. [2]

Postulated underlying causes include sudden rise in blood pressure, immunosuppression, chemotherapeutic agents for lymphoma and leukemia, severe hypercalcemia, thrombocytopenic syndromes, Henoch-Schönlein purpura, vasculitis, and renal failure, [1,2,3] of which sudden rise in blood pressure and renal failure appear to be the most common. [2]

Magnetic resonance imaging (MRI) typically demonstrates vasogenic edema in the posterior (parieto-occipital lobes) white matter. Atypical manifestations of PRES, in which the main lesions are discovered in regions other than parieto-occipital lobes, have been reported [4-8].

CASE REPORT

An 8 year old girl was admitted to the emergency department with complaints of severe headache, vomiting, generalized tonic-clonic seizure. She did not have findings of meningeal irritation. Her blood pressure was 135/95 mmHg (>95 p), and physical examination was normal. Fundoscopy was normal and revealed no sign of papilledema or hypertensive retinopathy.The Laboratory Findings revealed Hb 7.6 g/dl, White Blood Counts : 11300/cmm, Renal Funtion Tests : Blood urea nitrogen : 185 mg/dl, creatinine 8.1mg/dl, Calcium, : 6.6 mg/dl and Phosphorus : 7.8 mg/dl, eGFR 9ml/min/1.73, ABG : pH : 7.25 HCO3 : 16 .Ultrasound -Abdomen revealed bilateral small shrunken kidneys, suggestive of dysplastic kidney suggestive of chronic kidney disease. Her electroencephalography showed slow rhythm in the background. MRI showed low signal intensity on T1-weighted images and high signal intensities on axial FLAIR and T2-weighted images involving the cortical and partially subcortical regions of the bilateral temporo-occipital lobes.(Figure 1)

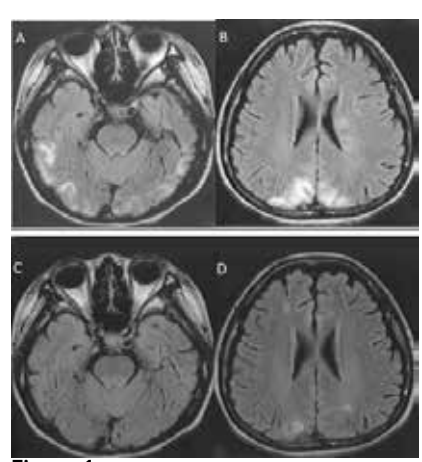

Figure 1

Figure 1 : (A-B) Fluid attenuated inversion recovery (FLAIR) sequence showing symmetrical hyperintensity over cortical

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and subcortical white matter of the bilateral occipital, parietal and posterior temporal lobes.(C-D) Nearly complete interval resolution of prior abnormal signal at the same locations is noted in follow-up imaging study 1 month later, consistent with reversible vasogenic edema.

The patient was drowsy and developed blurring of vision which progressed to loss of vision. Anticonvulsants, antihypertensives were started and acute peritoneal dialysis was performed. Later she was shifted to CAPD(Continuous Ambulatory Peritoneal Dialysis) .She recovered five days after the appropriate therapy for chronic renal failure was instituted with consultation of pediatric nephrologist. Repeat MRI was completely normal after one month on follow up.(Figure 1)

Five months after the first attack, the patient was readmitted to the emergency department with loss of vision, headache and left tonic clonic seizure. Her blood pressure was high (166/108 mmHg) and her physical examination revealed severe pallor. The laboratory findings revealed blood urea nitrogen 190 mg/dl and creatinine 9.0 mg/dl. Other laboratory parameters were normal.

MRI revealed edema in the posterior cerebral region as shown in Figure 2. The findings were suggested of recurrent PRES. Anticonvulsant and antihypertensives therapies were instituted. Hypertonic peritoneal dialysis (2.5%Dextrose instead of 1.5%) were used to augment ultrafiltration. The patient recovered within 3 days of institution of therapy. The MRI repeated after one month did not demonstrate any abnormality. The patient is managed conservatively (Figure 2)

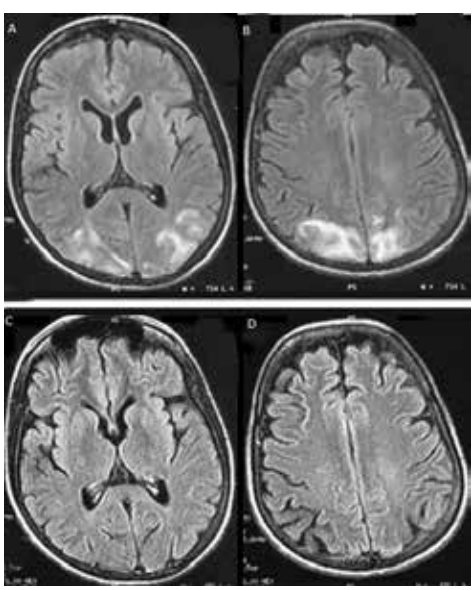

Figure 2

Figure 2 : (A-B) Fluid-attenuated inversion recovery (FLAIR) sequence showing symmetrical bilateral hyperintense lesions in the parieto-occipital regions affecting the cortex and subcortical white matter.(C-D) Fluid-attenuated inversion recovery (FLAIR) sequence reveal that the lesions have disappeared following treatment.

DISCUSSION

Posterior reversible encephalopathy syndrome clinically presents with seizures, severe headaches, cortical blindness and other visual abnormalities, altered mental status, and focal neurologic signs[1,4,6,7].

The cause of the PRES is multifactorial. The main cause of PRES is acute elevation of blood pressure above the upper limit of cerebral blood flow

autoregulation.[9] The uremia in End Stage Renal Disease (ESRD) has also been proposed as an independent triggering factor.[6] Patients with ESRD have a dysfunction in both vasopressor homeostasis and endothelial function related to elevations in lipoproteins,blood pressure, uremia, fluid overload, and drug therapies.[10] Chronic renal failure and hypertension were the common triggers in this patient.

Two possible mechanisms have been proposed in the pathophysiology of PRES. The first is vasospasm due to acutely increased blood pressure, and the second is loss of autoregulation. In the first hypothesis, it has been suggested that vasospasm contributes to ischemia and cytotoxic edema at regions of the arterial border zone.[11] The second hypothesis is supported by diffusion images suggesting that dilation develops in cerebral arterioles due to autoregulatory failure. The objective of cerebral autoregulation is to keep blood flow constant, and to protect the brain during changes in blood pressure; however, sudden and severe increases in blood pressure can impair autoregulation, and such impairment can, in turn, lead to arteriolar vasodilation and endothelial dysfunction. In this condition, plasma and red blood cells migrate to the extravascular space from the intravascular space, and vasogenic edema occurs.[12] The preferential involvement of the posterior circulation has been postulated as being due the sympathetic innervation protecting the brain from sudden increase in blood pressure being relatively less in the arterioles supplied by the vertebrobasilar system than in the anterior circulation.[3]

When promptly recognized and treated, radiological and clinical abnormalities can completely resolve. However, the lesions may not be reversible in all cases, and unrecognized patients can progress to ischemia and massive infarction with death.[6] This condition can cause neurological sequelae such as persistent brain damage and epilepsy, especially in those with hemorrhage on MRI arising from delays in diagnosis and therapy. [10,12-15] Early suspicion, identification of triggers and timely treatment of this syndrome can prevent long-term sequelae.

As indicated by the name of this syndrome, appropriate treatment is expected to ensure a full recovery. However, permanent complications and fatal cases have been reported, leading some authors to suggest that a better name may be "potentially reversible encephalopathy syndrome". [16]

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