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Case Report

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Posterior Reversible Encephalopathy Syndrome (PRES) Associated with Papilledema Mimicking Multiple Sclerosis and Boomerang Sign on Imaging: Report of Two Cases

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Abstract

Posterior reversible encephalopathy is a clinical syndrome of acute neurological symptoms like seizures, disturbed vision, headache, and altered mental state in the setting of renal failure, heart failure, blood pressure fluctuations, cytotoxic & anti-epileptic drug overdose or withdrawal, autoimmune disorder and eclampsia leading to sub cortical vasogenic edema as a result of endothelial injury. Here we are reporting two cases that presented as altered mental state and diagnosed posterior reversible encephalopathy syndrome (PRES) in the presence of risk factors, including renal, heart failure and eclampsia and fluctuations of blood pressure with subcortical white matter hyperintensities on brain imaging. Our first case had bilateral papilledema and MRI brain revealed subcortical white matter hyperintesities mimicking multiple sclerosis. Second case showed diffusion restrictions in the posterior splenium region with reduced corresponding ADC level (Boomerang sign) on neuroimaging. Both of our patients were managed symptomatically and discharged in a stable condition. The clinicians should be aware of such uncommon presentations of PRES in view varied clinical and radiological features.

Keywords

Posterior reversible encephalopathy syndrome (PRES), Papilledema, Boomerang sign, Multiple sclerosis

Introduction

Posterior reversible encephalopathy is a clinical syndrome of acute neurological symptoms in the setting of renal failure, blood pressure fluctuations, cytotoxic drugs, anti-epileptic drug overdose or withdrawal, autoimmune disorder and preeclampsia or eclampsia leading to sub cortical vasogenic edema as a result of endothelial injury predominantly involving the bilateral parietoccipital region on brain imaging [1].

There are various differentials of posterior reversible encephalopathy syndrome (PRES) including infectious encephalitis, autoimmune or paraneoplastic encephalitis, lymphoma, gliomatosis cerebri, sub cortical leukoaraiosis, CNS vasculitis, progressive multi focal leukoencephalopathy, osmotic demyelinating syndrome, acute demyelinating encephalomyelitis and anti-epileptic drugs or withdrawal [2].

The proposed mechanism in PRES was fluctuating rather than the absolute rise in blood pressure leading to endothelial dysfunction and disruption of the blood-brain barrier allowing the interstitial extravasation of plasma and macromolecules resulting in vasogenic or intramyelinic edema due to inflammatory cells, predominantly involving white matter and responsible for

signal changes in posterior splenium region [2, 3].

Case Report-1

A 37 years old male alcoholic presented with seven-day history of acute onset, mild to moderate holocranial headache associated with nausea, vomiting, blurring of vision and vertigo. He also had one episode of loss of consciousness lasting for 1-2 minutes. There was no history of fever, rash, trauma, tuberculosis, hearing loss or antiepileptic drug intake or withdrawal. General physical examination revealed PR 78/min, blood pressure 200/110 mm of Hg, which was decreased to 150/100 mm of Hg with antihypertensive medication over a period of 3-5 days. Fundus revealed bilateral papilledema (Figure 1) and rest of neurological examination was unremarkable. Routine hemogram, including HIV, VDRL, ESR and thyroid profile were normal while biochemistry revealed blood sugar 92 mg/dl, blood urea 79 mg/dl and serum creatinine 3.65 mg/dl. MRI brain showed cortical and subcortical, predominantly white matter hyperintensities in corona radiata, frontoparietooccipital and ponine region on T2 and FLAIR images (Figure 2).

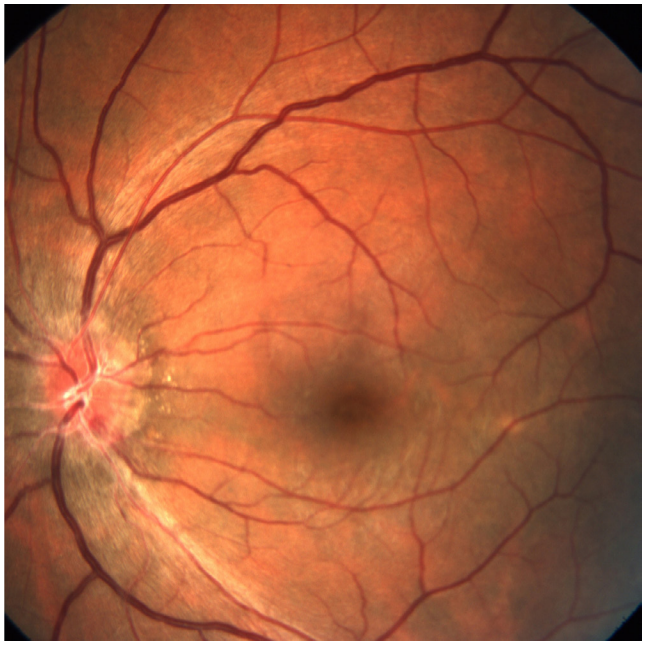

Figure 1: Fundus photograph showed fully developed papilloedema-disc margin blurred, obscuration of cup with exudates and venous engorgement.

Papilledema alone was not consistent with hypertension. Therefore, four differential diagnosis, including PRES, cortical venous thrombosis, Susac syndrome and idiopathic intracranial hypertension (IIH) were considered. Ultrasound of the whole abdomen showed normal kidney size. Serum electrolytes and bilateral renal Doppler were also normal. Further investigations like audiometry, autoimmune and vasculitis profile (ANA, Anti-ds-DNA, Anti-nucleosome, Anti-histones, Anti-Sm, Anti-SS-A, Anti-RO, Anti-Scl-70, Anti Anti-Rib-Protein, Anti-JO, Anti-SS-B) were also carried out to rule out Susac syndrome. MR venogram and angiogram of the brain were performed in view of cerebral venous thrombosis (CVT), and CSF pressure was 220 mm of Hg, which was not consistent with IIH diagnosis. 2D-ECHO showed global hypokinesia with ejection fraction 30%. In view of the presence of risk factors, including renal and heart failure, the diagnosis of posterior reversible encephalopathy syndrome was made [2]. He was treated with IV fluid, tablet amlodipine 5 mg/day, atenolol 50 mg/day and tab torsemide 20 mg/day. Patient was discharged in a stable condition.

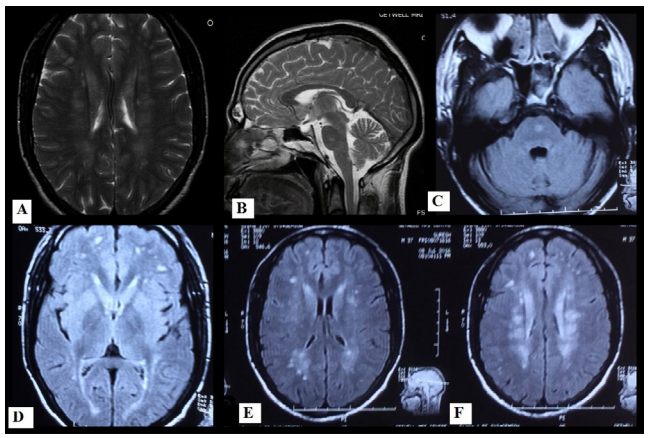

Figure 2: MRI brain showed cortical and subcortical predominantly white matter hyperintensities in corona radiata **(A, E, F)**, fontoparietooccipital **(D, E)** and pons **(B, C)** on T2 **(A, B)** and FLAIR **(C-F)** images.

Case Report-2

A 27 years old female presented with altered sensorium after normal vaginal delivery. Next day, she developed irritable behaviour and occasional irrelevant talking. There was no history of fever, rash, trauma, tuberculosis, anti-epileptic drug intake or withdrawal. General physical examination revealed PR 92/min, blood pressure 180/100 mm of Hg, which decreased to 160/96 mm of Hg without any antihypertensive medication over a period of 2-3 days. Neurological examination was normal except behavioral abnormalities. Routine hemogram and biochemistry, including ESR, HIV, VDRL, thyroid profile, renal and liver function tests were normal. Urinalysis

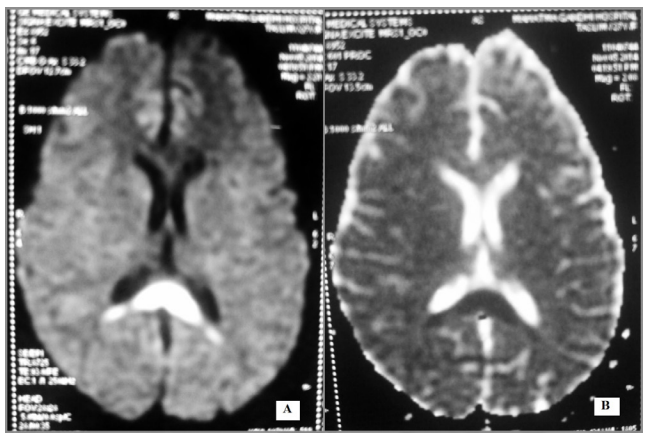

Figure 3: MRI Brain DWI **(A)** revealed diffusion restrictions in posterior splenium region with **(B)** reduced corresponding ADC level (Boombrang sign).

showed proteinuria. MRI Brain revealed diffusion restrictions in the posterior splenium region with reduced corresponding ADC level (Boomerang sign) (Figure 3). Therefore, diagnosis of PRES in setting of eclampsia was made [2]. She was treated with IV fluid, tablet olanzapine 5 mg and tablet lorazepam 2 mg hs for five days. At the time of discharge, her blood pressure was within normal limits. She could follow verbal commands, and behavioral aspect was normal.

Discussion

PRES evolves over a matter of hours, with varied clinical presentation like seizures, disturbed vision, headache, and altered mental state [4]. Posterior brain region may be more susceptible to hyper perfusion because of fewer sympathetic innervation occurs in the posterior fossa. Signal changes in the corpus callosum occur in several pathologic conditions, which may be transient, including seizures, antiepileptic drug overdose or abrupt drug withdrawal, infections, pre-eclampsia, axonal injury, drug toxicity cyclosporine, metronidazole and fluorouracil or permanent as seen in multiple sclerosis, Marchia fava- Bignami disease, tumor, leukodystrophy and HIV-related encephalopathy. These signal changes can be divided into two types according to shape and extent: circumscribed, well defined, oval lesion in middle or broad with fewer regular borders involving entire splenium mimicking boomerang sign as observed in our second case [5]. Diffusion restriction in the splenium is present in 15-30% cases of PRES [6]. Association of renal failure with PRES is present in 55% cases [7]. Prognosis of PRES is favourable, and most patients recover fully in about 75-90% cases [8] as seen clinically in both of our cases. There are three MRI patterns reported in 70% cases in radiology literature: a dominant parietooccipital pattern, holohemispheric watershed pattern and superior frontal sulcus pattern. Holohemispheric watershed pattern as seen in our first case, lesions were oval shaped, perpendicular to the ventricle, predominantly involving subcortical white matter mimicking multiple sclerosis [9].

Conclusion

PRES is a clinical syndrome diagnosed in the setting of renal failure, heart failure, blood pressure fluctuations, eclampsia and other conditions leading to sub cortical vasogenic edema as a result of endothelial injury. Here reported two cases presented as altered mental state and diagnosed PRES. One of them developed bilateral papilledema and subcortical white matter hyperintesities mimicking multiple sclerosis on brain imaging, which is an interesting observation. In second case, we reported "Boomerang" sign on neuroimaging, unusual presentation of PRES. So neuro physician should be aware of varied and atypical presentation of PRES with dealing such cases.

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