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Case Report

Brain Stem Posterior Reversible Encephalopathy Syndrome in Nephrotic Syndrome

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Abstract

Posterior reversible encephalopathy syndrome is typically characterized by a clinico-neuroradiological entity. It is well known that severe hypertension, vasoconstriction or systemic toxicity predispose patient with nephrotic syndrome to PRES. A 45-year old male with nephrotic syndrome was referred for sudden developed vertigo and disequilibrium. His blood pressure was within normal limit. He had not been on immunosuppressive or cytotoxic drugs. Horizontal jerk nystagmus was observed by eccentric right gaze. He showed hemiparesis and limb ataxia in right side. Brain MRI showed vasogenic edema in whole midbrain, pons and cerebellar peduncle symmetrically. He was given intravenous dexamethasone and diuretics to reduce vasogenic edema and raise the intravascular osmotic pressure. Nystagmus, motor weakness and limb ataxia improved gradually. He recovered without any neurologic symptom and sign. Follow-up MRI showed decreased vasogenic edema fairly. We report a case of brain stem PRES in normotensive nephrotic syndrome patient.

Keywords: *Posterior Reversible Encephalopathy Syndrome; Nephrotic Syndrome*

Abbreviations

PRES: Posterior Reversible Encephalopathy Syndrome

Introduction

Posterior reversible encephalopathy syndrome (PRES) is typically characterized by acute onset headache, altered mentality, seizures and visual loss associated with imaging findings of bilateral subcortical and cortical edema with a predominantly posterior distribution [1]. PRES is commonly identified in patients with eclampsia, organ transplantation, abrupt hypertension, systemic lupus erythematosus, Wegener granulomatosis, postchemotherapy and non-specific renal inflammatory conditions (glomerulonephritis, hepatorenal syndrome) [2]. Nephrotic syndrome is a syndrome comprising signs of proteinuria, hypoalbuminemia, and edema. It is well known that severe hypertension, vasoconstriction, use of immunosuppressive or cytotoxic drugs predispose patient with nephrotic syndrome to PRES [3]. Here, we report a case of PRES with atypical lesion in nephrotic syndrome without hypertension and use of immunosuppressive drug. We suggest that low intravascular osmotic pressure may play a role in developing PRES in nephrotic syndrome with review of literature.

Case

A 45-year old male was referred for sudden developed vertigo, disequilibrium. 5 months ago, he was diagnosed nephrotic syndrome and had taken diuretics. His medical history included diabetes controlled with medication. Renal biopsy revealed diabetic nephropathy as the cause of nephrotic syndrome. He was hospitalized because of generalized edema a few days ago. His vital signs were blood pres-

sure of 120/70 mmHg, pulse rate of 72/min and no fever. On neurologic examination, his mental state was alert. Horizontal jerk nystagmus was observed by eccentric right gaze. He showed good grade motor weakness and ataxia in right upper and lower limbs without other sensory abnormality. Because of a history of diabetes, stroke was highly suspected. The initial Brain MRI was performed 3 hours after the onset of neurologic symptoms. Brain MRI showed increased signal intensity in FLAIR and apparent diffusion coefficient (ADC) mapping, decreased signal intensity in T1 image and focal enhanced lesion in T1 contrast image at whole midbrain, pons and cerebellar peduncle symmetrically, which was compatible with vasogenic edema (Figure 1). According from MRI feature, PRES and CNS vasculitis were suspected. Laboratory findings showed severe proteinuria and hypoalbuminemia. Urine protein / creatinine ratio was 8.494, serum creatinine 1.27 mg/dl, albumin 2.4 g/dl, normal magnesium and electrolyte. We diagnosed as brainstem PRES. We began treatment of intravenous dexamethasone 15 mg/day and oral furosemide 240 mg/day to reduce vasogenic edema and raise the intravascular osmotic pressure. We terminated intravenous dexamethasone after 3 days, but maintained oral furosemide. Nystagmus, motor weakness and limb ataxia improved gradually. 2weeks later, He recovered without any neurologic symptoms and signs. Follow-up MRI showed decreased vasogenic edema fairly (Figure 2). He has not had a recurrence to date in over 1year follow-up.

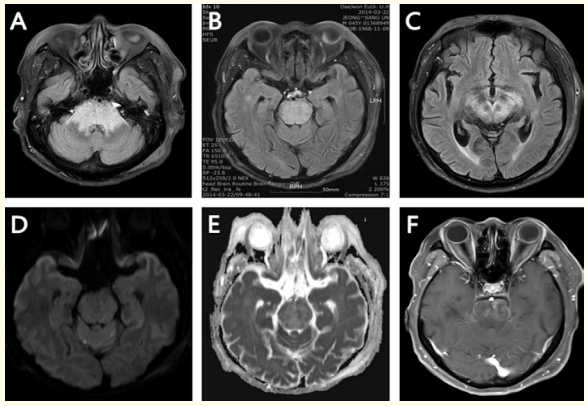

Figure 1: *Initial MRI of patient. Fluid-attenuated inversion recovery (FLAIR) image both revealed symmetrical hyperintensities at whole midbrain, pons and cerebellar peduncle symmetrically (A, B, C). Diffusion-weighted imaging (DWI) showed low intensity at the same areas, while apparent diffusion coefficient (ADC) mapping showed high intensity (D, E). Focal enhanced lesion in T1 contrast image at same area. These findings indicated vasogenic edema (F).*

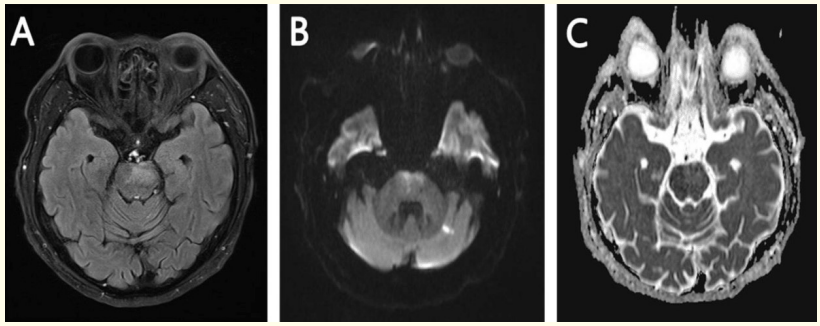

Figure 2: *Follow-up MRI performed 14 days showed that previous high signal in the pons and cerebellar peduncle decreased in FLAIR MRI (A). Diffusion MRI and apparent diffusion coefficient (ADC) mapping showed iso-signal intensity at same regions (B, C).*

Discussion

PRES usually affects the posterior regions. The posterior brain regions can be particularly susceptible to hyperperfusion because little sympathetic innervation exists in the posterior fossa. It could also developed in brain stem of 13% of cases [2]. In cases of brain stem lesion, higher blood pressure is usually noticed likewise in that of posterior regions. But, PRES could be developed in patients with normotensive state [4,5]. The pathophysiology of PRES, although not fully elucidated, is assumed to be potentiated by elevated arterial pressure exceeding the limits of cerebral autoregulation leading to hydrostatic brain edema or autoregulatory vasoconstriction leading to ischemia and subsequent edema. Other potential pathogenic mechanisms for PRES may include T-cell activation leading to inflammatory cytokine production, which up-regulates cell adhesion molecules and increased leukocyte trafficking leading to cerebral microcirculatory dysfunction [5]. Although the hypertension/hyperperfusion theory is favored due to the common presence of elevated blood pressure and perceived response to hypertension management, key issues remain problematic, including PRES in normotensives, toxicity pressures rarely reaching autoregulatory limits, and brain edema lower in severe hypertensives. A recent study has shown that vasogenic edema on the neuroimaging was significantly associated with hypoalbuminemia and that its severity was correlated with serum albumin level in PRES patients with other underlying disease [6]. Serum albumin normally accounts for 75% of protein in blood plasma and, therefore, also for 75% of colloid osmotic pressure. Colloid osmotic pressure affects osmotic pressure because the negative charges attract sodium, thus holding water and exerting an antagonistic effect on perfusion pressure. In conditions with endothelial damage due to inflammatory processes, reduction of colloid osmotic pressure may facilitate fluid extravasation and development of vasogenic edema. A possible correlation between low albumin levels and PRES is supported by case report described 7 pediatric patients who developed PRES during nephrotic syndrome and low levels of serum albumin. In these patients, PRES rapidly abated after substitution of albumin, despite further treatment with cytotoxic drugs [7]. A reversible course had been revealed after proper management including adequate control of blood pressure, treatment of seizures, and removal or reduction of causative factors. In our case, we selected diuretics as a treatment of PRES instead of substitution of albumin to raise intravascular osmotic pressure, which is supported by the course that diuretics abolished symptoms of patient and MR abnormalities successfully.

Conclusion

We report a case of brain stem PRES in normotensive nephrotic syndrome patient.

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