A Case of Chronic Renal Failure and Recurrent Posterior Reversible Encephalopathy Syndrome

Kronik Böbrek Yetersizliği ve Rekürran Posterior Reverzibl Ensefalopati Sendromu (PRES) Olgusu

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ABSTRACT

Posterior reversible encephalopathy syndrome (PRES) is a rare clinical and radiological entity of neurological origin which has different etiologies. A 27-year-old male patient with a history of type 1 diabetes, uncontrolled hypertension and chronic renal failure presented to the emergency department with complaints of confusion, high blood pressure and seizures. Magnetic resonance imaging (MRI) revealed bilateral hyperintense lesions in the occipital lobes with vasogenic edema. The lesions had disappeared at follow-up MRI. Two months after the onset, the patient presented with atypical headache and high blood pressure and, the MRI showed new, multiple hyperintense lesions in the parieto-occipital region. The diagnosis of recurrent PRES was considered based on the onset of symptoms and MRI findings. There were only 5 cases of recurrent PRES reported in the literature and only one with chronic renal disease. The hallmark of PRES is vasogenic edema in the territories of the posterior circulation. provides powerful means to establish the diagnosis and gives prognostic information about the follow-up of the patient. These patients should be carefully monitored, and the physician should be alert for possible recurrence. (Archives of Neuropsychiatry 2011;48: 270-3)

Key words: Renal failure, recurrent PRES, vasogenic edema

Introduction

Posterior reversible encephalopathy syndrome (PRES) is a rare clinical and radiological entity of neurological origin which has different etiologies. Headache, altered mental status, cortical blindness, seizures, focal neurological signs and blood pressure instability are the major clinical findings of this syndrome (1,2). There is no consensus in the literature regarding the causative factors, but hypertensive encephalopathy, eclampsia, metabolic disorders, and iatrogenic or toxic agents have all been implicated in the etiology (2-5).

The typical radiological findings of PRES are symmetric, bilateral subcortical and/or cortical hyperintensity on T2-weighted magnetic resonance imaging (MRI) scans, especially in the parieto-occipital lobes; less commonly, the brainstem, basal ganglia, and cerebellum may also be involved (1,6-8). Computed tomography (CT) and MRI characteristically reveal symmetrically distributed areas of vasogenic edema predominantly located within the territories of the posterior circulation of the brain (3). The pathological process affects primarily the white matter, but the cortex may also be involved (7,9). Diffusion-weighted MRI (DWI) has been shown to be a

Ozet


Anahtar kelimeler: Böbrek yetersizliği, rekürran PRES, vasojenik ödem

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reliable method in distinguishing vasogenic edema in PRES from cytotoxic edema seen in cerebral ischemia (10,11).

In spite of the dramatic manifestations, the clinical picture may be fully reversible with early diagnosis and adequate treatment. Otherwise, the disease may lead to permanent cerebral damage and even death (1,12-16).

There are few recurrent cases of PRES published in the literature and only one case was reported with coexisting end-stage renal disease (17). In this paper, we present a case of recurrent PRES in a patient with chronic renal failure who responded well to treatment and whose radiological findings improved.

Case Report

A twenty-seven-year-old male patient with a history of type 1 diabetes mellitus, uncontrolled hypertension and chronic renal failure presented to the emergency department with confusion and high blood pressure. He had a history of several hypoglycemic attacks during the past year. Generalized tonic-clonic seizures started in the emergency room and EEG revealed diffuse slowing with 6Hz theta waves. Emergent MRI was performed and, T2-weighted and FLAIR images revealed bilateral hyperintense lesions in the occipital lobes with cortical-subcortical and partially deep white matter involvement; DWI showed vasogenic edema (Figure 1a,1b,1c).

The patient was hospitalized and the seizures resolved with 20 mg/kg intravenous phenytoin infusion, followed by 1000 mg levetiracetam (per os) treatment. The blood pressure normalized after glyceryl trinitrate infusion. Immediately after the blood pressure and seizures were controlled, the patient became conscious and EEG findings returned to normal.

Laboratory tests, including routine biochemical tests, were within normal limits; hepatitis A, B, C, HIV, syphilis (VDRL, TPHA), vasculitic (ANA, RF, antiphospholipid antibodies, ENA profile) markers were negative; B12 and folic acid levels were normal. Thyroid hormone levels were within normal range (TSH: 1.85 mU/L (0.27-4.2), free T3: 3.92 (pmol/l) (1.57-9.71), free T4: 1.57 (pmol/l) (0.93-1.7). Cerebrospinal fluid (CSF) analysis revealed normal cell count and normal glucose, protein and LDH levels. Erythrocyte sedimentation rate (ESR) was elevated (60mm/hour).

The clinical presentation, which included confusion, seizures, high blood pressure and MRI findings showing vasogenic edema in the posterior regions, suggested PRES (1,2). ESR was elevated, but the patient had neither fever nor signs suggestive of acute infection. Underlying chronic disease was thought to be responsible for the elevated ESR.

The patient was discharged and the cranial MRI examination was repeated 1 month later (Figure 2a, 2b). The lesions had disappeared at the follow-up MRI and the patient was symptom-free. One month after the second MRI, the patient attended the neurology outpatient unit with complaints of severe headache, nausea and high blood pressure (190/100 mmHg). Neurological examination was normal, but MRI showed new, multiple hyperintense lesions in the parieto-occipital region (Figure 3). The patient was hospitalized again.

Figure 1 a, 1b, 1c. Sagittal T2-weighted images (1a) show hyperintense lesion in the left parietal white matter (arrows). Bright signal on the axial diffusion-weighted image (arrow, 1b) and hypointensity on the apparent diffusion coefficient map (arrow, 1c) indicate the presence of vasogenic edema.

Figure 2 a, 2b. Sagittal T2- (2a) and T1-weighted (2b) pre- and post-contrast images show the disappearance of the lesions.

Figure 3. Sagittal T2-weighted images show recurrence of the parietal white matter lesions, very similar to the initial presentation (arrows).
Lowering the blood pressure to normal limits lead to amelioration of the headache and nausea symptoms and, he was discharged. MRI performed 3 months later showed that the former lesions had resolved considerably; the DWI scans were normal (Figure 4a, 4b).

Discussion

PRES is defined as a cliniconeuroradiological entity (1,2). The major clinical findings of PRES are headache, seizures, visual disturbances, altered mental status and other focal neurological signs. Many etiologic factors are implicated, such as uremia, renal failure with hypertension, collagen vascular disease, pre-eclampsia/eclampsia, immunosuppressive and cytotoxic medications, organ transplantation and porphyria (2,18,19).

The mechanism of PRES is not clear, but there are two main hypotheses. The first one suggests that severe hypertension exceeds the limits of autoregulation and results in brain edema; the second hypothesis suggests that vasoconstriction and hypoperfusion are the factors that produce brain ischemia and vasogenic edema (20). Sudden increases in blood pressure may impair autoregulation and lead to arteriolar vasodilatation with endothelial dysfunction. Following this pathological process, the plasmocytes and the red blood cells migrate to the extravascular space and vasogenic edema occurs (21).

CT and MRI typically show symmetrically distributed areas of vasogenic edema, predominantly in the posterior circulation territories. The abnormalities affect primarily the white matter, but the cortex is also involved (1,2,5,10-12,14). Previous studies (5,10,11) have shown that vasogenic edema is responsible for the changes seen in PRES, therefore, the hallmark of PRES is vasogenic edema in the territories of the posterior circulation, which can be differentiated from cytotoxic edema in other etiologies by using DWI as well as apparent diffusion coefficient (ADC) map that shows elevated ADC values (22).

To our knowledge, there are only 5 cases of recurrent PRES published in the literature. Hagemann et al. (23) have reported a case of recurrent PRES of unknown etiology following intensive care unit treatment with only moderately elevated blood pressure. Sweany et al. (24) have presented 3 patients with sickle cell disease, antibody-positive autoimmune disease, and allogenic bone marrow transplantation who developed recurrent PRES. Finسترer et al. (25) have reported PRES associated with multi-system mitochondrial disorder. Ergün et al. (17) have presented the first recurrent PRES case in a patient on hemodialysis for end-stage renal disease with multiple congenital anomalies including ectopic single kidney.

In our case, the patient had chronic renal failure due to type I diabetes mellitus and was treated with peritoneal dialysis. Uncontrolled blood pressure and hypoglycemic attacks began in the last year. First PRES episode occurred with high blood pressure, confusion and seizures and the second episode with headache and high blood pressure. In both of the episodes, there were bilateral parieto-occipital lesions. Vasogenic edema was detected during the first episode by DWI. During the second episode, we could not perform DWI because of technical problems. DWI had been performed in some of the previously reported cases of recurrent PRES. Hagemann et al. (23) and Sweany et al. (24) showed vasogenic edema on FLAIR sequences, while DWIs were normal. Ergün et al. did not perform DWI, but they also detected posterior localization of the lesions on FLAIR sequences (17).

Sweeney et al. (24) proposed that infections might trigger this process and they have documented infection in both episodes of all their 3 patients with recurrent PRES. In our patient, the ESR was elevated, but there were no other signs and symptoms of an underlying infectious process. In this case, the final MRI revealed only partial resolution of the lesions in the posterior regions, while the DWIs were normal.

Patients who experienced PRES should be very carefully monitored for recurrent PRES episodes, even in the absence of new symptoms. Covarrubias et al. (22) suggested that the DWI findings may represent an early sign of nonreversibility in PRES, heralding the conversion to infarction. Therefore, DWI provides not only powerful means of diagnosing PRES but also gives prognostic information about the follow-up of the patient.

The hallmark of this diagnosis is vasogenic edema in the posterior circulation territories, which can be reliably differentiated from cytotoxic edema seen in other pathologies using DWI method and ADC map.

The extent of T2 and DWI signal intensity correlates well with patient outcome and can help guide more aggressive treatment in more severely affected patients (22).

References