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Posterior Reversible Encephalopathy Syndrome: Case Report

Posterior Reversibl Ensefalopati Sendromu: Vaka Raporu

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Abstract

Posterior reversible encephalopathy syndrome (PRES) is a neurological disease characterized by a range of neurological signs, symptoms and different neuroimaging findings reflecting vasogenic edema. Etiology of PRES may include hypertension, eclampsia/preeclampsia, sepsis, immunosuppressive agents, chemotherapy, collagen-vascular diseases and renal failure. Although SLE is a rare cause of PRES, endothelial damage and increased blood pressure due to vasculitic involvement are thought to play a role in the etiology. In this case report, a 38-year-old patient with a diagnosis of SLE who has a hypertensive attack and visual loss was presented.

Keywords: Posterior Reversible Encephalopathy Syndrome, Systemic Lupus Erythematosus, Hypertension.

Özet

Posterior geri dönüşümlü ensefalopati sendromu (PRES), bir dizi nörolojik belirti, semptom ve vazojenik ödemi yansıtan farklı nörogörüntüleme bulguları ile karakterize nörolojik bir hastalıktır. PRES etiyolojisinde hipertansiyon, eklampsi/preeklampsi, sepsis, immünosüpresif ajanlar, kemoterapi, kollajen- vasküler hastalıklar ve böbrek yetmezliği yer alabilir. SLE, PRES'in nadir bir nedeni olmasına rağmen, vaskülitik tutuluma bağlı endotel hasarı ve artmış kan basıncının etiyolojide rol aldığı düşünülmektedir. Bu olgu sunumunda 38 yaşında SLE tanısı olan, hipertansif atak ve görme kaybı ile başvuran bir hasta sunuldu.

Anahtar Kelimeler: Posterior Geri Dönüşümlü Ensefalopati Sendromu, Sistemik Lupus Eritamatozus, Hipertansiyon.

INTRODUCTION

Posterior reversible encephalopathy syndrome (PRES) is a neurological disease characterized by a range of neurological signs, symptoms and different neuroimaging findings reflecting vasogenic edema. The reason why it is called reversible is that clinical neurological findings and imaging findings improve hours or days after treatment is started (1).

Clinical conditions that cause changes in blood pressure, drugs, dysautonomia, chemotherapy, and discontinuation of antihertensive treatment can be considered as risk factors for the development of PRES syndrome (2). The clinical symptoms of toxicity are extensive, but most commonly headache, mental status changes, seizures, nausea/vomiting, and focal

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neurological disorders are observed (3). Visual findings such as hemianopia, cortical blindness, visual hallucinations, and visual acuity deterioration may occur in approximately two-thirds of PRES patients due to occipital lobe involvement (4).

This case report aims to, present a patient with a diagnosis of Systemic Lupus Erythematosus (SLE) who presented to the emergency with sudden vision loss, nausea, vomiting and confusion.

CASE

A 38-year-old patient, who had been diagnosed with SLE for 7 years, presented to the emergency department with sudden vision loss, nausea, vomiting and confusion. It was learned that the patient had applied to the emergency department with 2 hypertensive attacks before, used ramipril 5 mg irregularly, and did not have regular follow-up and treatment. In the neurological examination of the patient, her consciousness was latergic and her orientation-cooperation was limited. In the pupillary midline, the light reflex was taken directly or indirectly. She could not cooperate with her eye movements, could not count fingers in one meter, and did not have facial asymmetry. All four extremities were spontaneously mobile, plantar reflex bilateral flexor and there was no nuchal rigidity.

The patient's blood pressure was 210/110 mmHg and fever: 36.9°C. Electrocardiogram was normal. In laboratory tests, white blood cell: 8.6 K/uL (4-10), hemoglobin: 13.1 g/dL (11-17), platelet: 310 K/uL (100-380), C-reactive protein: 6.2 mg/L (0-5), urea: 41.2 mg/dL (8-20), creatinine:1.47 mg/dL (0.51-0.95). Liver function tests, electrolytes, and thyroid function tests were normal. Complete urinalysis was normal. In brain magnetic resonance imaging (MRI), fluid-attenuated inversion application recovery (FLAIR) sections, symmetrical hyperintense vasogenic edema findings in bilateral occipitotemporal regions were observed (A). Apparent diffusion coefficient (ADC) map showed high signal (B). MRI angiography and venography were normal. (Figure 1)

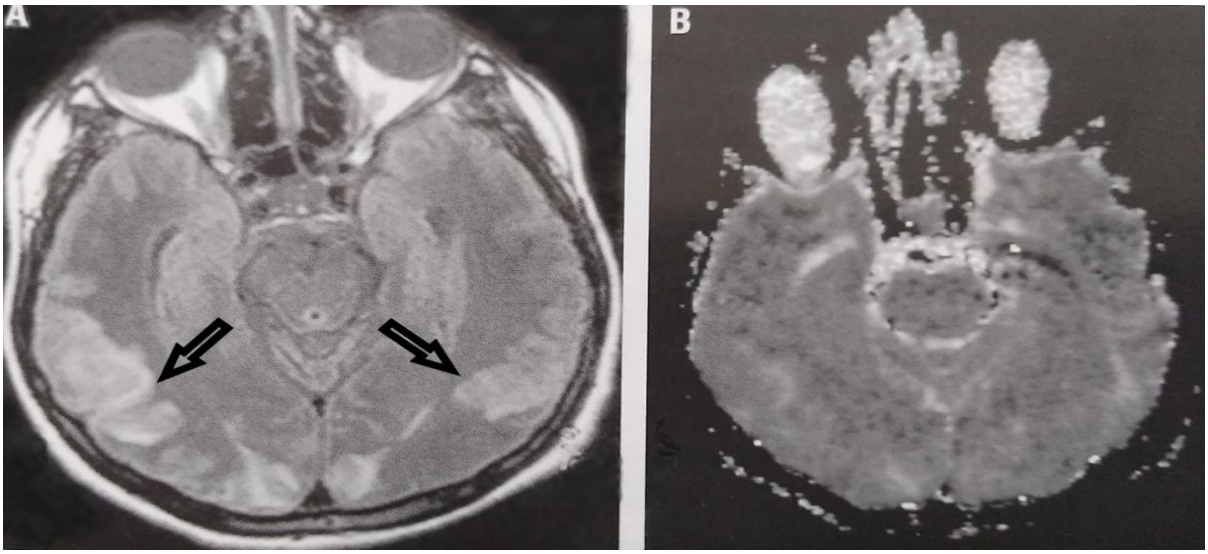

Figure 1. A. Fluid-attenuated inversion application recovery (FLAIR) sections. Symmetrical hyperintense vasogenic edema findings in bilateral occipitotemporal regions. B. Apparent diffusion coefficient (ADC) map showed high signal

The diagnosis of PRES was made by evaluating the patient's anamnesis, clinical findings and imaging. The patient was admitted to the neurology service and intravenous nitroglycerin

treatment was started to control blood pressure. Blood pressure values were gradually reduced and blood pressure regulation was achieved at the end of 12 hours. The patient was given 10 mg of mannitol 3 times on the first day as antiedema treatment, then the daily dose was tapered and discontinued. Since the patient could take it orally, 150 mg acetylsalicylic acid treatment was added. The patient's confusion resolved within 12 hours, and visual impairment almost completely resolved within 6 days. Cranial MR findings obtained at the end of a week showed almost complete improvement.

DISCUSSION

In this case report, we aimed to present a patient with a diagnosis of SLE who presented with a hypertensive episode with vision lossand confusion in the clinic and diagnosed with PRES.Although the pathophysiology of PRES is not known exactly, its etiology includes hypertension, eclampsia/preeclampsia, sepsis, immunosuppressive agents, chemotherapy, collagen-vascular diseases, and kidney failure (5). Our patient had a diagnosis of SLE and her blood pressure values were very high. Similarly, Sudan et al. reported a 32-year-old patient with a diagnosis of PRES who presented with a hypertensive episode and acute vision loss in both eyes (6).

PRES is a reversible picture characterized by segmental vasoconstriction and vasodilation in small cerebral vessels, often resulting from cerebral vascular tone irregularity (1). In its pathophysiology, disruption in cerebrovascular autoregulation and the resulting vasogenic edema are the main causes (7). Another reason is endothelial dysfunction caused by circulating endogenous or exogenous toxins (8). Although SLE is a rare cause of PRES, endothelial damage and increased blood pressure due to vasculitic involvement are thought to play a role in the etiology (9).

Brain MRI findings in patients with PRES characteristically show vasogenic edema. The most common neuroimaging finding is bilateral focal edematous areas in the frontal and inferior temporal-occipital junctions, especially in the parietal and occipital lobes (1). In our case, we observed hyperintense areas showing vasogenic edema in bilateral occipitotemporal regions in FLAIR sequences, consistent with the literature.

The primary goal in PRES treatment is to address the underlying cause, such as lowering blood pressure, taking antiepileptics or correcting electrolyte disturbances with sedation, hydration. In cases with acute hypertension, blood pressure should be reduced gradually, since rapid correction of blood pressure may lead to coronary, renal and cerebral ischemia (3).

In conclusion, late diagnosis or inadequate treatment in PRES may contribute to long-term sequelae such as permanent neurological disability, progressive brain edema, intracranial hemorrhage, and death. PRES should be considered in the differential diagnosis of patients presenting with hypertensive episodes and atypical clinical findings in SLE.Early diagnosis and correct interventions are crucial to achieve a favorable clinical outcome, as they can rapidly improve clinical symptoms and radiological findings.

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Conflict of interest: None declared.

Informed consent: The patient was informed for publication of the case report andaccompanying imagesconsent was obtained.

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