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Case Report

Posterior Reversible Encephalopathy Syndrome in Systemic Lupus Erythematosus: An Underdiagnosed Condition

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Abstract

Posterior reversible encephalopathy syndrome (PRES) is a rare underestimated neurological condition characterized by non-specific symptoms like headache, seizure, altered mental status, visual disturbance, and a typical finding of edema in the posterior white matter of the brain, without any signs of infarction. PRES can occur in conjunction with eclampsia, kidney disease, cytotoxic drugs, and immunosuppressants. Its manifestation in patients with Systemic Lupus Erythematosus (SLE) is particularly uncommon. An early and accurate diagnosis of PRES is essential to improve the outcome and ensure a better prognosis for the patient. Delayed diagnosis can lead to permanent neurological damage. Treatment primarily involves supportive care and addressing the underlying disease. We report a case of PRES in a 21-year-old female with SLE with lupus nephritis class five.

Keywords: Posterior Reversible Encephalopathy (PRES); PRES Treatment; Pathogenesis of Pres; Systemic Lupus Encephalopathy

Introduction

Posterior Reversible Encephalopathy Syndrome (PRES) is a complex neurological disorder that presents with a myriad of symptoms such as headache, stroke, seizures, meningitis, intracranial hypertension, and disorientation. It has a common radiological finding of parieto-occipital watershed-region edema without overt infarction. The risk factors include high blood pressure and systemic toxicity associated with immune activation, such as the coexistence of systemic lupus erythematosus [1]. PRES in systemic lupus erythematosus (SLE) is an infrequent presentation [2]. Significant therapeutic and prognostic implications can be achieved by reversing the clinical and radiologic abnormalities through rigorous blood pressure control, cessation of the offending medicine, and treatment of the underlying illness. If undiagnosed, it might progress to cytotoxic edema, which is permanent. The literature is relatively sparse regarding the occurrence of PRES in patients with SLE. Consequently, this report details the case of a 21-year-old female diagnosed with SLE with lupus nephritis stage five who was subsequently identified through radiographic imaging as having PRES. This case contributes to the limited knowledge of the co-existence of these two complex conditions.

Case Presentation

A 21-year-old female patient, a known case of SLE with lupus nephritis class five was brought to the emergency department by her mother after experiencing a generalized tonic-clonic seizure at home. She had a severe headache that started the day before the seizure episode. She took two tablets of acetaminophen, which did not alleviate her symptoms, leading to four instances of vomiting.

Later that day, while lying down, she experienced an episode of seizure that was resolved spontaneously. It was not accompanied by tongue bite or urinary incontinence. The patient was examined in a postictal condition with a blood pressure of 218/143 mm of Hg, a pulse rate of 120 per minute, a respiratory rate of 26 breaths per minute, and a SpO2 level of 98%. There were no signs of neck stiffness and neurological impairments. The cranial nerves and motor tests were normal. She experienced another seizure episode in the emergency room and was treated with intravenous lorazepam 4mg, levetiracetam 1g, and was started on intravenous nicardipine. The consumption of alcohol or illicit drugs was absent. There was no personal or familial history of seizures.

Laboratory data showed elevated erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), creatinine, leukocyte count, and anemia. Lab values are shown in table 1. The non-contrast computed tomography (NCCT) brain scan revealed symmetrical areas of reduced density in the white matter of the occipital hemispheres as shown in figure 1 [3].

Labs	Units	Values	Reference range
WBC COUNT	10*3/uL	19.60	4-10
HEMATOCRIT	%	49.6	37-47%
HEMOGLOBIN	g/dL	15.9	12-16
PLATELETS	10*3/uL	178	150-450
MCV	fL	88.7	80-98
RDW	%	15.3	9-14.5%
SODIUM	mmol/L	139	136-145
POTASSIUM	mmol/L	4.6	3.5-5.0
CHLORIDE	mmol/L	105	95-105
BUN	mg/dL	21	8-20
CREATININE	mg/dL	1.84	0.8-1.3
GLUCOSE	mg/dL	104	70-99
CALCIUM	mg/dL	7.7	8.5-10.2
MAGNESIUM	mg/dL	2.6	1.6-2.6
PHOSPHORUS	mg/dL	7.8	2.5-4.5
ESR	mm/hr	80	0-20
CRP	mg/dL	16	0.3-1

Table 1: Laboratory values.

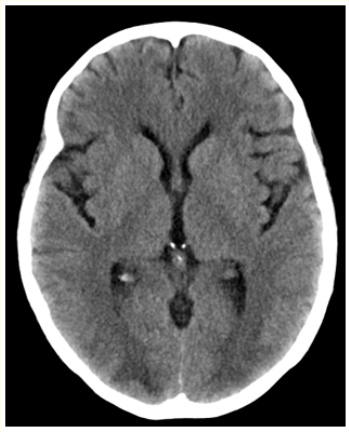

Figure 1: The non-contrast computed tomography (NCCT) brain scan shows symmetrical areas of reduced density in the white matter of the occipital hemispheres. (Image courtesy: Dr. Bruno Di Muzio, Radiopaedia.org, rID 55441).

The results of the video electroencephalogram monitoring were within normal limits. She was monitored for two days, during which she did not experience any further seizures, and her blood pressure was effectively managed. The patient has been prescribed 500 mg of levetiracetam twice a day for two weeks along with Fioricet every six hours to manage headaches. She was started on ACE inhibitors and was instructed to monitor her blood pressure periodically. CT findings were shown to have returned to normal imaging after 2 weeks.

Discussion

PRES is associated with a diverse array of medical conditions, as mentioned in table 2. These conditions include, but are not limited to, bone marrow transplantation, eclampsia/preeclampsia, organ transplantation, autoimmune diseases, and the administration of high-dose chemotherapy [4]. This extensive range of associations underscores PRES's complexity and multifaceted nature, necessitating a broad understanding of its potential triggers in various clinical scenarios.

General conditions	Autoimmune diseases	Toxins	Cytotoxic and immunosuppressive medications	Other medications
Hypertension Sepsis Transplantations Renal failure Malignancy Preeclampsia and eclampsia Hypomagnesemia Hypercalcemia Hypercholesterolemia Radiation induced brain injury SMART Syndrome	Rheumatoid arthritis Crohn's disease Systemic lupus encephalopathy Scleroderma Vasculitis Neuromyelitis spectrum disorder	Scorpion poison LSD intoxication Ephedra overdose Alcohol intoxication Cocaine	Cyclosporine Tacrolimus Hydroxydaunorubicin/adriamycin Vincristine/vinblastine Gemcitabine Platinum containing drugs Bortezomib Interferons Corticosteroids Etoposide Capecitabine/5fluorouracil Rituximab Methotrexate Azathioprine Mycophenolate mofetil	Lithium Linezolid Intravenous contrast Intravenous immunoglobulins Tyrosine kinase inhibitors

Table 2: Causes of PRES.

In SLE, a systemic disorder is usually the first manifestation but it can also start as a neurological presentation. Seizures are seen very commonly in SLE patients along with intracranial hypertension, TIA, and psychiatric disturbances [5]. It can also present initially as PRES [6]. When SLE presents with neurological symptoms multiple differentials like CNS infections, thrombotic events, PRES, and cerebrovascular events should be ruled out.

The majority of cases are characterized by endothelial activation and hypertension. Hypertension is considered the primary reversible cause of PRES [7]. There are two opposing theories regarding the pathophysiology of the PRES. First, according to the latest vasogenic theory, severe hypertension exceeds the autoregulatory capacity causing blood vessel dilation, increased permeability, and the development of brain edema. The older cytotoxic theory says that hypertension causes cerebral vasoconstriction, ischemia, and brain edema [8,9]. However, hypertension alone is unlikely to explain the mechanism of PRES as it can be seen in SLE patients with normal blood pressure too [10].

Inflammatory mechanisms may influence PRES in SLE patients. The key mediator is interleukin-6 [11]. Recent research on patients with SLE and PRES revealed an increase in serum interleukin-6 levels compared to active SLE patients without PRES [12]. The Signal transducer and activator of the transcription-3 (STAT-3) pathway is activated by interleukin 6 and upregulates nitric oxide synthase expression and other vasodilators [13]. These molecules stimulate the blood vessels' endothelium, making them more permeable and breaking down the blood-brain barrier. This makes patients more likely to get vasogenic edema and neurological injury from high blood flow.

The treatment for PRES is supportive and depends on the underlying condition. Whether PRES was due to high blood pressure or SLE-related factors or immunosuppressive medications remains speculative. In hypertension or drug-induced PRES, it is crucial to withdraw the offending drug, control the blood pressure, and give antiedema and anti-convulsive therapy along with temporary renal support. SLE-related PRES is managed with steroids and alkylating agents. Acute hypertension patients should progressively lower their blood pressure by 20%-25% in the first few hours to avoid cerebral, coronary, and renal ischemia, aiming for a mean arterial pressure between 105-125 mm of Hg [14].

Conclusion

PRES is a largely underdiagnosed condition. Since PRES is reversible, early diagnosis with clinical examination and neuroimaging is very crucial to avoid complications and mortality. Hence, being a time-sensitive disease providing the

essential treatment at the right time is the need of the hour for such patients. In reviewing the existing case literature, two significant gaps were identified. Firstly, there is a lack of research into the potential risk factors that may increase the likelihood of developing PRES in patients with SLE. Understanding these factors along with the pathophysiology could facilitate the development of tailored prevention and treatment strategies. The impact of conventional SLE therapies, including corticosteroids and other immunosuppressive drugs, on the progression and resolution of PRES remains poorly understood. Further, there is a need for a thorough understanding of diverse clinical presentations including the range of neurological symptoms. Further studies addressing these issues are essential to enhance our management approaches and improve patient outcomes.

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