

Postoperative posterior reversible encephalopathy syndrome as initial presentation of systemic lupus erythematosus

Joshua Sunny George, Shahil Mehta, Patricia Calvo

ABSTRACT

Introduction: Posterior reversible leukoencephalopathy syndrome (PRES) is a syndrome consisting of neurological symptoms including headaches, visual changes, and seizures often occurring in the setting of uncontrolled hypertension. Diagnosis is often confirmed by characteristic findings on neuroimaging studies. **Case Report:** We present a case of a 25-year-old African-American woman with a history of chronic pelvic pain secondary to recurrent endometriosis presenting with chief complaints of fever and pelvic pain. She was treated with laparoscopic ablation three months prior. Workup revealed bilateral tubo-ovarian abscesses and the patient underwent total abdominal hysterectomy and bilateral salpingo-oophorectomy. Postoperatively, the patient had new onset hypertension which eventually lead to a seizure episode. The patient was transferred to the ICU, started on nicardipine and Keppra and her hypertension improved within several hours. Neuroimaging findings on MRI scan revealed lesions in the occipital and parietal lobes consistent with PRES. Outpatient workup conducted several months afterwards uncovered a diagnosis of systemic lupus erythematosus, leading us to conclude that the postoperative hypertensive emergency

and PRES were secondary to undiagnosed SLE. **Conclusion:** The rare complication of PRES has been described in a variety of settings including SLE in which endothelial dysfunction of the intracerebral vasculature leads to characteristic PRES symptoms. Patients, especially those in the postoperative setting covered by multiple specialty providers, with new onset hypertension and neurological symptoms should warrant further workup as they may indicate underlying etiologies such as SLE or other described risk factors for PRES.

Keywords: Hypertension, Posterior reversible encephalopathy syndrome, Systemic lupus erythematosus, Total hysterectomy

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Joshua Sunny George¹, Shahil Mehta¹, Patricia Calvo¹

Affiliation: ¹University of Miami Miller School of Medicine, Miami, FL, USA.

Corresponding Author: Joshua Sunny George, 82 Gordon Drive, Troy, Michigan, USA, 48098; Email: Jsg58@med.miami.edu

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INTRODUCTION

Posterior reversible leukoencephalopathy syndrome (PRES) is a syndrome consisting of neurological symptoms including headaches, visual changes, and seizures often occurring in the setting of uncontrolled hypertension [1]. Diagnosis is often confirmed by characteristic findings on neuroimaging studies. The PRES has been reported in association with systemic lupus erythematosus (SLE) and

is thought to be related to disease activity [2]. There have been several case series published regarding patients with SLE and PRES; the majority of patients were previously diagnosed and were on immunosuppressive therapy at the time of their PRES episode [3]. We describe a patient with no prior history of hypertension who experienced uncontrolled hypertension, vision changes, and seizure in the postoperative setting after a total abdominal hysterectomy and bilateral salpingo-oophorectomy (TAH-BSO). Imaging and clinical findings led to a diagnosis of PRES. She recovered appropriately after several days and was discharged without an etiology for her new onset hypertension. It was not until several months later that outpatient workup led to an underlying diagnosis of SLE.

CASE REPORT

A 25-year-old female with a past medical history of severe endometriosis status post laparoscopic ablation and recent intrauterine device placement presented to the hospital with debilitating abdominal pain, a fever of 39.3°C, and tachycardia. On admission, she also complained of chest pain and difficulty swallowing medication, feeling as though, the pills are getting stuck. This led to intravenous administration of medications, including an intravenous PPI for possible pill esophagitis secondary to doxycycline. Pelvic ultrasound demonstrated large bilateral complex adnexal masses and labs showed a normal white blood cell count and anemia with hemoglobin 7.6 and a hematocrit of 22%. All other laboratory studies were within normal limits. The patient requested definitive treatment for her symptoms and consented for TAH-BSO. Two units of packed red blood cells were given to the patient prior to surgery. The surgery was performed without any complications and a Jackson Pratt drain was placed in the RLQ. During surgery, the patient was confirmed to have bilateral tubo-ovarian abscesses.

On postoperative day-1, the patient was tachycardic with a heart rate of 100 beats per minute and blood pressure of 156/111 mmHg. Oral labetalol was ordered for the patient to control her tachycardia and blood pressure. Of note, she had no prior history of hypertension. She also continued to have difficulty swallowing pills which persisted throughout her hospital course. On postoperative day-2, the patient had a normal heart rate and blood pressure; however, she developed acute kidney injury with oliguria, which was corrected with intravenous fluids. For the following two days, the patient's blood pressure began to rise so the dose of labetalol was increased and oral amlodipine was started. On postoperative day-5, the patient developed worsening tachycardia and hypertension with a maximum blood pressure of 182/118 mmHg during the day. She also complained of a new onset persistent headache and blurry vision. Labetalol was discontinued, clonidine was

started and the dosage of amlodipine was increased. Later that day, the patient had one episode of vomitus and complained of worsening vision changes. She then experienced a seizure episode which lasted less than 3 minutes and was followed by a period of postictal confusion. Blood pressure was measured at 175/114 mmHg during the code rescue for the seizure. She denied any seizure history or family history of seizures. The patient was transferred to the intensive care unit (ICU) scan and Keppra (UCB Pharma Inc., Smyrna, Georgia, USA) was given for seizure prophylaxis and a nicardipine drip was started to control for her hypertension.

Magnetic resonance angiography (MRA) and magnetic resonance imaging (MRI) scan of brain without contrast were ordered for further evaluation. Magnetic resonance angiography of brain showed no evidence of thrombosis. Magnetic resonance imaging scan of brain without contrast showed edema signal changes in a cortical and subcortical distribution in both the subdural cortices, temporal cortices, in the right frontal and parietal cortex (Figure 1). There was mild reversal on diffusion. These imaging findings in context of the clinical picture suggested the diagnosis of posterior reversible encephalopathy syndrome. Following the seizure episode, further inquiry was made into the cause of uncontrolled hypertension during the patient's hospital stay. At the time, it was thought to be that the patient was not compliant with her

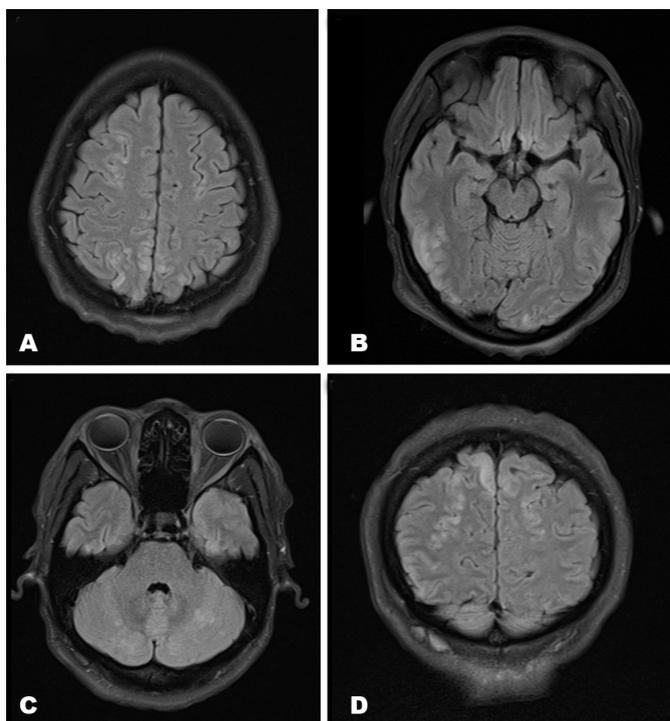


Figure 1: Axial views on T2 FLAIR revealed: (A) Patchy areas of increased signal with slight involvement of the right frontal lobe and posterior parietal lobe right greater than left, (B) Along the cortex in the right temporal and occipital lobes and the left occipital lobe, (C) In the cerebellar areas in the distribution of the posterior cerebral circulation, and (D) Coronal view of increased FLAIR signal changes typical in posterior reversible leukoencephalopathy syndrome.

oral hypertension medications during her hospital course as she had continued discomfort when swallowing pills.

During the remaining four days of the hospitalization, the patient did not experience any further neurological symptoms and had a negative EEG. She had a CT brain completed without contrast which showed no acute intracranial hemorrhage and loss of normal gray-white matter cortex along the frontal, parietal and occipital cortex and subcortical distribution, greater on the right side. She was weaned off of the nicardipine and transitioned to an oral medication regimen of amlodipine, chlorthalidone, and lisinopril. By this time, her ability to swallow oral medication improved. She was discharged on amlodipine, chlorthalidone, and lisinopril for blood pressure control and Keppra for seizure prophylaxis. She was encouraged to follow up with her primary care and to return within a month for further imaging studies.

The patient did not follow up for imaging; however, two months later, she was seen in the emergency department on two separate occasions with a chief complaint of arthralgias. At the first visit, she was discharged from the emergency department on corticosteroids with the diagnosis of arthralgias secondary to an adverse reaction from Lupron shot she had received months prior to treat her endometriosis. She was referred to a rheumatologist at this time. To note, the patient was anemic at this time with hemoglobin 9.3 g/dl and hematocrit 28.2. CRP and ESR of the patient were also elevated. She returned to the emergency department a week later with the same complaint, stating that her appointment to see the rheumatologist was not for several weeks. She was discharged again the same day with the same diagnosis. She was seen by her primary care physician later that month who eventually diagnosed her with SLE. She was found to have positive laboratory tests for ANA, anti-Smith, anti-dsDNA. She was started on a regimen of mycophenolate mofetil, hydroxychloroquine, and prednisone and her symptoms resolved. Further neurological workup at sixth month included MRI sequences of T2 FLAIR, diffusion and ADC, and T1 with and without contrast. Imaging appeared unremarkable with no lasting changes from prior PRES episode. In addition, she had a normal EEG and no further seizure episodes or any other clinical signs of neurological changes.

DISCUSSION

Posterior reversible leukoencephalopathy syndrome (PRES) was first described by Hinchey et al. as a recognizable pattern of headache, altered mental status, seizure, and vision loss [1]. Other sequelae of intracranial hypertension may be present as well, contributing to the high variability in clinical presentation [4]. The PRES is often found with characteristic findings on neuroimaging studies including edema in the posterior cortical white matter. Although named as such, findings again may be variable and diagnosis is not restricted to

strictly posterior or reversible lesions [5, 6]. It has been described in the literature in association with pregnancy, eclampsia, drug toxicity and autoimmune conditions such as systemic lupus erythematosus (SLE), as was the case for our patient. The underlying pathophysiology is focused around the endothelial hypothesis and high blood pressure [7, 8]. These theories purport an inciting factor for sustained or uncontrolled hypertension leading to endothelial dysfunction and loss of nitric oxide production. This results in subsequent vasogenic edema which is found on imaging studies.

Although our patient's course was diagnosed relatively quickly and resolved without complication, PRES identification in the acute setting is important for preventing associated morbidity. In a retrospective study of severe PRES, Legriél et al. reported increased prevalence of status epilepticus and unfavorable functional outcome based on Glasgow Outcome Score [9]. Time to control of causative factor was found to be an independent predictor of 90 day functional outcome, highlighting the importance of recognition of clinical presentation and intervention [10, 11]. In this case, delays in diagnosis of the patient were due to confounders in her clinical presentations. Our patient experienced hypertension preoperatively which was thought to be caused by pain from her chief complaint. This condition along with the medication noncompliance secondary to suspected pill esophagitis disguised the underlying cause of the patient's hypertension emergency, her undiagnosed SLE. No further workup for new onset hypertension was completed, because the clinical course was relatively benign after the seizure episode as blood pressure was switched to intravenous management and anti-epileptics were started. She was simply diagnosed with secondary hypertension. Closer follow-up on a telemetry/ICU floor was helpful in monitoring vital signs postoperatively and may be suitable for recommendation for all patients undergoing procedures with general anesthesia [12].

Comparing our case to a review of case series pertaining to SLE and PRES revealed similarities in most common presenting symptoms, age of onset, and brain lobes affected [2]. The unique component of this presentation is that PRES was the initial finding of SLE and that the onset occurred in the postoperative setting. The majority of PRES cases reported come after patients have already been diagnosed with SLE and started on immunosuppressive therapy. PRES onset is related to increase in disease activity and can signal the need for adjustments of immunosuppressive therapy to control lupus activity [2]. There are also other reports of PRES occurring after TAH; however, in those cases, PRES was attributed to other causes such as rapid correction of anemia and incomplete pain control [13, 14].

CONCLUSION

Posterior reversible leukoencephalopathy syndrome (PRES) is a rare complication has been described in a

variety of settings including systemic lupus erythematosus (SLE). The underlying endothelial dysfunction from lupus disease activity is suspected to contribute to the interruption of neurovascular autoregulation, placing patients at an increased risk for vasogenic edema and clinical symptoms. Multi-specialty care teams working together in the postoperative setting should take note of patients with new onset hypertension and neurological symptoms such as seizures as they may be indicators of underlying autoimmune processes such as SLE or other described risk factors for PRES. Early detection and management may lead to proper workup and treatment at an earlier clinical disease state, which may lead to better long term outcomes for patients.

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Author Contributions

Joshua Sunny George – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Shahil Mehta – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Patricia Calvo – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

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Authors declare no conflict of interest.

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