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Posterior Reversible Encephalopathy Syndrome in Systemic Lupus Erythematosus: Case Report of a Young Female Patient

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Abstract

Posterior Reversible Encephalopathy Syndrome is a rare manifestation of lupus erythematosus, in which there is altered mental function, headache, convulsions, visual impairment and posterior and transient changes on neurologic imagistics [1,2], occurring in a few hours and presenting with rapidly progressive neurologic symptoms [3]. It was first described in 1996. It is more frequent in patients with acute kidney injury or chronic kidney disease, and is associated with hypertension, autoimmune diseases, immunosuppressive therapies-especially antibody-based immunosuppressive therapy, and organ transplantation. It is clinically reversible within one week and imagistically reversible within 2-4 weeks. It is treatable and has a good prognosis. We present the case of a young woman of 27 years diagnosed with systemic erythematous lupus who developed convulsive crisis, headache, visual impairment, being under immunosuppressive therapy with azathiorpine. She had class IV lupus nephritis on the kidney biopsy and partial remission of the nephrotic syndrome. Other lupic manifestations were cutaneous, immunological, articular and hematological. She had a good short-term prognosis at 30 days.

Keywords: Erythematosus systemic lupus; Neurological manifestations of lupus; Posterior reversible encephalopathy; Seizures; Lupus nephritis; Reversible neurological lesions

Introduction

Posterior reversible encephalopathy is a rare condition usually associated with autoimmune diseases, immunosuppressive drugs – especially antibody-based therapies, organ transplantation and renal chronic or acute disease. It is a rare manifestation of lupus erythematosus, in which there is altered mental function, headache, convulsions, visual impairment and posterior and transient changes on neurologic imagistics [1,2], occurring in a few hours and presenting with rapidly progressive neurologic symptoms [3]. It was first described in 1996 by Hinchey et al. in 1996 based on 15 cases reports [1]. It can include focal neurological deficits, status epilepticus or coma, but these are less frequent [4]. On neuroimaging techniques, reversible lesions consistent with white matter cerebral vasogenic edema occur in the posterior reginos of the cerebral hemispheres – occipital, parietal lobes, rarely involving the brain stem, or the cerebellum (sub cortical edema without infarction [1]). It is presumed that the physiopathological mechanisms involve capillary leakage and disruption of the brain-blood barrier [5]. Several medical terms have been used to name this syndrome, mainly reversible posterior leukoencephalopathy syndrome, reversible posterior cerebral edema syndrome, and reversible occipital parietal encephalopathy [6,7]. Other imagistic elements that are compatible with this diagnosis include the presence of haemorrhage, restricted diffusion, contrast enhancement, and vasoconstriction are all compatible with a diagnosis [8].

The global incidence of PRES is not known. Epidemiological data come from retrospective studies of patients, mainly seen between 1988-2008, being reported in patients aged 4-90, most cases occurring in young to middle-aged adults, mean age between 39-47 years [9-11]. It is more frequent in females and patients usually have comorbidities such as chronic hypertension, chronic renal failure, eclampsia, preeclampsia, bone marrow or organ transplantation or autoimmune diseases. Mechanical ventilation is required in 35% to 40% of patients with PRES, for 3 to 7 days [10,12]. Status epilepticus may require ICU admission. It has a very low incidence in lupic patients, authors citing 0.69% of all lupic patients [7]. The real incidence in not firmly known, as there are cases in which the neuroimagistic results and follow-up are not available.

Definition

Posterior reversible encephalopathy syndrome (PRES) is a rare neurological disorder; its frequency is increasing in lupus

patients. It is a clinic-imagistic syndrome that includes sensory impairment, seizures, headache, visual disturbances and transient changes on MRI of the posterior brain and cerebellum showing sub cortical lesions of edema, it being caused by capillary leakage.

Physiopathology

PRES is a neurotoxic state accompanied by a trypical brain imaging of subcortical edema in occipital, parietal lobes, cerebellum. The mechanism behind the developing vasogenic edema and CT or MR imaging appearance of PRES is not known.

There are two theories:

a. Severe hypertension that damages the auto-regulation, subsequent hyperperfusion, with endothelial injury/ vasogenic edema.

b. Vasoconstriction and hypoperfusion leads to brain ischemia and subsequent vasogenic edema, immune challenge present and develop a similar state of T-cell/endothelial cell activation that can generate leukocyte trafficking and systemic/cerebral vasoconstriction. The imagistic findings support the older theory of vasoconstriction coupled with hypoperfusion as the mechanism, hypertension leading to cerebral autoregulatory vasoconstriction, ischemia and subsequent cerebral edema [13,14].

Treatment

Management of PRES in SLE includes the treatment of acute hypertension, seizure, lupus activity, and admission into an intensive care unit if necessary. Oral or intravenous antihypertensive drugs are used in most episodes of PRES, diuretics and renal replacement therapy are used for oliguria and edematous status not responsive to diuretic treatment. The elective treatment for lupus control are steroids, usually methylprednisolone pulse-therapy. Because immunosuppressive therapy is associated with PRES, a short discontinuation or dose reduction in classical immunosuppressives in recommended (azathioprine, mycophenolate, calcineurin-inhibitors, antibody-mediated immunosuppressives), until the patient is stabilized and then initiated under strict medical observation.

Case Report

We present the case of a young female patient of 27 years, diagnosed with systemic lupus erythematosus since decembrer 2009, with biopsy proven lupus nephritis class III-IV, with impure nephrotic syndrome since the diagnostic. She had cutaneous, articular, immunological, hematological and renal manifestations of lupus. The patient underwent temporary peritoneal dialysis in 2015 for an acute kidney injury episode due to lupus vasculitis, with partial recovery of renal function afterwards, maximum creatinine being of 4mg/dl, and after the remission of the acute renal failure the creatinine level being of 2.2mg/dl. She also had Cushing's syndrome after corticotherapy, secondary osteoporosis, mixed dyslipidemia, secondary arterial hypertension (reno-parenchymal), maximum arterial pressure of 190/110mmHg, chronic anemia secondary to the systemic lupus.

The patient received 6 monthly pulses of cyclophosphamide for rapidly progressive renal failure in 2015, and another acute kidney injury episode due to pre-renal ischemia in February 2016, maximum creatinine of 3.59mg/dl, medium hypocomplementemia, and received treatment with mycophenolate mofetil 1.5grams/day. The patient had a good immunological and renal response, but the anemia worsened, due to the mycophenolate treatment regimen. Considering all these aspects, in December 2016 oral mycophenolate mofetil was replaced with oral azathioprine in dosage of 1mg/kg/day associated with 7.5mg oral prednisone/day. In February 2017 the patient presented with generalized tonico-clonic seizures, altered conscience level – moderate comatose state, headache, altered vision. Clinically, she had no peripheral edema, normal dieresis, blood pressure of 190/11mmHg, generalized hyperreflexia, no cervical stiffness. Lumbar punction showed no pathological modifications of cerebrospinal fluid. EEG showed dominant alpha waves in posterior territory, without paroxysmal episodes and without interremispheric electrical bursts. Cerebral CT showed small hypodense areas in the cortico-subcrotical occipital lobes bilaterally and normal ventricles. IRM showed hypersignal areas T2/FLAIR predominantly in sub cortical areas at the border territories of the frontal, parietal and occipital lobes, simetrically, also in the inferior temporal lobes and in the superior cerebellum, and mild ventricular system distension. The patient had moderate renal failure, compatible with chronic kidney disease stage III B, normal ionogram and mild anemia, mild schysocytes. The patient received anti-convulsivant therapy with Levetiracetam 1g/day, diuretic, anti-hypertensive therapy with remission of convulsive crisis.

She presented in the nephrology department three days after with complete urinary retention, interpreted as secondary to levetiracetam, the dosage being reduced from 1g to 500mg/day in 7 days, and to 250mg/day afterwards. Few cases of complete urinary retention are known to be reported [15], but after the reduction of the dosage the patient regained urinary bladder control in 20 days. The patient had a good neurological prognosis, without any focal motor or sensory deficits. She remained on the anticonvulsant therapy in the dosage of 250 mg/day of Levetiracetam.

Discussion

PRES is a rare condition in lupic patients, the risk factors for developing PRES being female sex, young age, the presence of hypertension, lupus activity and lupus nephritis [16,17].

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