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Case Report

Posterior reversible encephalopathic syndrome in systemic lupus erythematous: A case report

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Abstract

Posterior reversible encephalopathy syndrome (PRES) implies to a reversible subcortical vasogenic brain edema, which leads to some acute neurological symptoms. PRES occurs usually in the setting of renal failure, blood pressure fluctuations, cytotoxic drugs, autoimmune disorders, and pre-eclampsia or eclampsia.This is a 40 year old female patient with history of systemic lupus erythematous (SLE) from twenty years ago, admitted to our neurology ward due to severe and refractory headache with multiple convulsive events.Brain MRI showed vasogenic brain edema in occipito-parieto-frontal lobes white matter compatible with PRES. Treatment is omitting of the etiology, but in these patients, discontinuation of immunosuppressive drugs is not possible and probably intravenous methylprednisolone pulse is a good treatment option especially in patient without response to aggressive control of blood pressure

Introduction

Posterior reversible encephalopathy syndrome (PRES) first illustrated by Hinchey et al in 1996[1] imply to a reversible subcortical vasogenic brain edema lead to some acute neurological symptoms such as seizures, encephalopathy, headache, and visual obscuration. PRES occurs usually in the setting of renal failure, blood pressure fluctuations, cytotoxic drugs, autoimmune disorders, and pre-eclampsia or eclampsia. Brain imaging usually reveals vasogenic edema predominantly queering the bilateral parieto occipital regions. Possible pathophysiology of PRES is endothelial injury related to abrupt blood pressure changes or direct impact of cytokines on the endothelial tissues, which leads to breakdown of the blood–brain barrier and subsequent brain edema. PRES is generally reversible, both radiological and clinically, and has a desirable prognosis.[2]

Typical magnetic resonance imaging (MRI) findings include bilateral white matter hypointensity or isointensity on T1, and hyperintensity on T2 and fluid attenuated inversion recovery (FLAIR) in the parieto-occipital lobes, which is more or less symmetrical in most patients.[3]

Case Report

A 40-year-old female patient with history of systemic lupus erythematous (SLE) from 20 ago under treatment with low dose oral prednisone and hydroxychloroquine, without any recent history of steroid pulse admitted to our neurology ward due to severe and refractory headache with multiple convulsive events.

The patient had history of common migraine headache and generalized epilepsy from the beginning of the disease. But these new headaches were more severe, generalized, compressive in quality and refractory to routine medications. Also the recent convulsive seizures were secondary generalized with focal onset.

She was under treatment by low dose of steroid, hydroxychloroquine and azathioprine. On examination, the patient was alert and oriented. Temperature was 37.2°C, blood pressure was 160/90 mm Hg. Neurological exam was normal. There were not meningeal signs or pupil edema. The other physical exam was normal.

There was not detected any acute pathological event in the first day brain CT scan.

The patient's serum creatinine level and creatinine clearance were normal but urinalysis revealed proteinuria (+3). Anemia was seen as well as mild thrombocytopenia. Other biochemical tests and electrolytes were normal. Lumbar puncture was performed to rule out the infective or inflammatory meningeal process. Although IgG index in CSF was 0.7, but CSF analysis was normal and oligoclonal bands were negative.

Electroencephalogram) EEG) did not show any underlying convulsive disorder. Brain magnetic resonance venography (MRV) was normal. Brain magnetic resonance imaging (MRI), in T2 and FLIAR sequences showed hyperintensity in occipito-parieto-frontal white matter with predominance in occipital lobes, without

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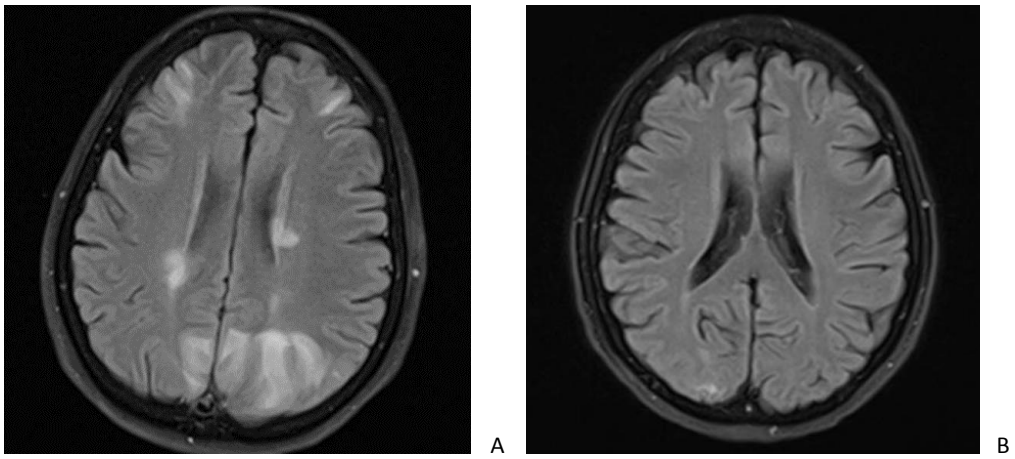

Figure 1. Brain magnetic resonance imaging (MRI), in FLIAR sequences showed hyperintensity in occipito-parieto-frontal with predominance in occipital lobes (A), with resolution of most hyperintensity in control brain MRI after 2 weeks (B).

any restriction in diffusion weighted sequences (DWI), compatible with brain edema (Figure 1). Cervical cord MRI was normal too.

Seizures eliminated with adjuvant levetiracetam, but headaches did not response to aggressive control of blood pressure, and we tried intravenous methylprednisolone pulse (IVMP), 1000 mg/day.

After three days of IVMP, her headache completely subsided and control brain MRI showed resolution of most hyperintense regions.

Discussion

Most clinicians and researchers believe that the meaning of the disease name (abbreviated as PRES) can be spurious since brain edema in this disease is frequently not restricted to posterior regions and sometimes the syndrome is not reversible. There is increasing reports that some of patients with PRES will endure sustain neurological disability or mortality.[4]

SLE is one of the predisposing conditions related to PRES, particularly when accompanied by lupus nephritis.[5-7] Most patients with SLE who experienced PRES were younger, had active lupus nephritis at the time of event, and suffered from hypertension, headache, visual obscuration and seizures.[8]

The treatment of PRES in the sitting of SLE principally related to etiology. In the setting of hypertension, effective treatment consists aggressive control of blood pressure and use of antiepileptic drugs if necessitate. In the condition PRES is due to high doses of corticosteroids and immunosuppressive drugs, key point of treatment is discontinuation of this medication.[2] But because most patients with SLE need continuous or even increase immunosuppressive drugs to control the active SLE attack, immunosuppressive drugs should not be discontinue or reduce but rather increase.[9]

Near to 90% of patients completely improve from neurological symptoms within one week.[9] Actually, postponement in diagnosis and treatment may lead to death or sever neurological disability.[10]

Conclusion

PRES can complicate SLE in the setting of hypertension, lupus nephritis and immunosuppressive drugs. Treatment is omitting of the etiology, but in these patients, discontinuation of immunosuppressive drugs is not possible and probably intravenous methylprednisolone pulse is a good treatment option especially in patient without response to aggressive control of blood pressure.

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Author Contributions

SH: diagnosis, treatment and report the case, critic and writing.

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Ethical Approval

To report this case, we got inform consent from the patient and her husband.

Conflict of Interest

None.

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