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Posterior Reversible Encephalopathy Syndrome (PRES) as an Initial Presentation of Systemic Lupus Erythematosus (SLE)

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Abstract

Background: Posterior Reversible Encephalopathy Syndrome (PRES) refers to a clinico-radiological syndrome characterized by headache, altered mental function, visual disturbance, seizures and transient posterior changes on neuro-imaging.

Case report: We present a case of a 38 year old female who presented with sudden onset behavioural change, visual disturbance and subsequent witnessed generalized tonic-clonic seizure. A cerebral MRI showed extensive signal abnormality consistent with vasogenic oedema in the cerebellar hemispheres and subcortical white matter of the occipito-parietal lobes consistent with a neuro-radiological diagnosis of Posterior Reversible Encephalopathy Syndrome (PRES).

Conclusion: We describe a case of a dramatic first presentation of Systemic Lupus Erythematosus (SLE) with neurological manifestations of PRES, occurring in the absence of accompanying evidence of lupus nephritis, cerebritis or other usual causative factors, such as immunosuppression.

Keywords: Posterior reversible encephalopathy syndrome; Systemic lupus; Erythematosus

Introduction

Posterior Reversible Encephalopathy Syndrome (PRES), first described in 1996, is a neuro-radiological entity characterized by headache, altered mental function, visual disturbance, seizures and posterior transient changes on neuro-imaging [1]. Classical MRI findings were initially described as bilateral sub-cortical hyper-intense areas involving occipital and parietal lobes although involvement of other lobes including the cerebellum have since been reported [2,3].

Systemic Lupus Erythematosus (SLE) is a chronic multisystem inflammatory disease that follows a relapsing and remitting course. It is characterized by an auto-antibody response to nuclear and cytoplasmic antigens. SLE can affect any organ system, but mainly involves the skin, joints, kidneys, blood cells, and nervous system [4].

PRES can occur at anytime in an SLE patient, but the usual identified risk factors are hypertension, renal involvement and the use of immunosuppressive agents, particularly cyclosporine [4].

Case Presentation

A 38 year old female, Fijian Indian descent, presented with sudden onset behavioural change, confusion, visual disturbance and subsequent witnessed generalized tonic-clonic seizure on a background of a 1 week history of a probable viral prodrome of vomiting and diarrhoea. There were no other neurological symptoms at the time. On arrival to the emergency department, she was noted to be hypertensive with a BP of 176/104 mmHg. This was treated acutely with intravenous hydralazine, which resulted in a short-term reduction in blood pressure. She then had a second witnessed generalized tonic-clonic seizure. Examination revealed no focal neurological deficits and the absence of neck stiffness or papilloedema.

A cerebral CT revealed no mass lesion however a non-specific low density area in the left putamen was seen. Lumbar puncture showed clear CSF, which was acellular, with a protein of 1.39g/L (0.15-0.45), and normal glucose. She was given a bolus of intravenous phenytoin and commenced on acyclovir, cefotaxime and benzylpenicillin for a provisional diagnosis of meningo-enchephalitis. An Electro Encephalo Gram (EEG) showed generalised slowing with delta slow wave activity throughout the recording, with no epileptiform activity, consistent with a diffuse encephalopathy.

She subsequently developed blurring of her vision, following the lumbar puncture, and a cerebral MRI was organized. This showed extensive signal abnormality consistent with vasogenic oedema in the cerebellar hemispheres and subcortical white matter of the occipito-parietal lobes (Figure 1). Based on the clinical presentation and imaging changes, a diagnosis of Posterior Reversible Encephalopathy Syndrome (PRES) was made, to be due to undiagnosed hypertension.

Oral levitiracetam was commenced for seizure control and oral ramipril for management of blood pressure, which was up to 190/120 [6]. She continued to receive intermittent intravenous hydralazine. Despite the addition further oral anti-hypertensive therapies, amlodipine and subsequently metoprolol, confusion progressed to unresponsiveness necessitating intubation with intermittent positive pressure ventilation, 48 hours after admission.

A repeat EEG showed changes of non-convulsive status, with generalized slowing and right temporal epileptiform activity. A repeat CT brain revealed gross cerebral oedema with early herniation. Mannitol was administered and an extra-ventricular drain placed for presumed raised intracranial pressure.

Further investigations revealed a homogenous ANA at a titre

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of 1:640, markedly elevated dsDNA antibody level of > 80 KlU/L, elevated C-reactive protein of 181 and low C3 and C4. A diagnosis of Systemic Lupus Eyrthematosus was made and intravenous high-dose methyl prednisone, 1 gram daily, was instituted with clinical and blood pressure improvement.

Subsequent directed history from family revealed a 2 year history of recurrent skin rash over the lower limbs, mild alopecia and anaemia. There was no history of arthritis, malar rash, miscarriage or thromboembolism. She also had no previous history of hypertension.

The patient's condition improved gradually, with an improvement in mental state, cognition and there were no further seizures. Eight days later, she was extubated and the extra-ventricular drain removed. Anti-cardiolipin and beta-2-glycoprotein 1 antibodies were not detected although lupus anti-coagulant was positive. Even though she had a normal urea and creatinine, a renal biopsy was performed due to the presence of proteinuria (0.5g/day) and persisting microscopic haematuria. This revealed changes consistent with hypertension and a resolving post-infective or focal proliferative glomerulonephritis, rather than lupus nephritis.

Prior to discharge, she was commenced on Mycophenolate, 500 mg twice a day, and a weaning regimen of oral prednisone. Long-term, the patient has shown a response with no further neurological symptoms, and improvement in dsDNA antibody, C3 and C4 to near-normal levels. A repeat MRI at 3 months showed a marked improvement with near-complete resolution of the abnormal high T2 signal in cerebral and cerebellar hemispheres (Figure 2).

Discussion

Posterior Reversible Encephalopathy Syndrome (PRES) is a neuro-radiological entity characterized by headache, altered mental function, visual disturbance, seizures and posterior transient changes on neuro-imaging [1]. Classical MRI findings were initially described as bilateral sub-cortical hyper-intense areas involving occipital and parietal lobes although involvement of other lobes including the cerebellum have since been reported [2,3].

PRES has been described in association with systemic vasculitis, thrombotic thrombocytopenic purpura, haemolytic uraemic syndrome, infection and other miscellaneous conditions [7]. PRES has also been described in association with a number of immunosuppressive and cytotoxic agents including corticosteroids, cyclophosphamide, mycophenolate and rituximab [7]. Although typically a reversible entity, some patients have developed permanent neurological deficits after an episode of PRES [7].

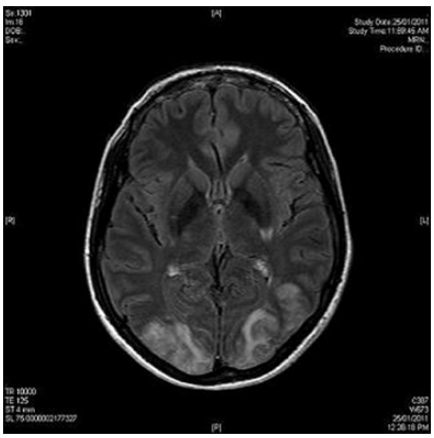

Figure 1: Hyperintense T2 cortical and sub cortical white matter intensity involving both occipital and parietal lobes].

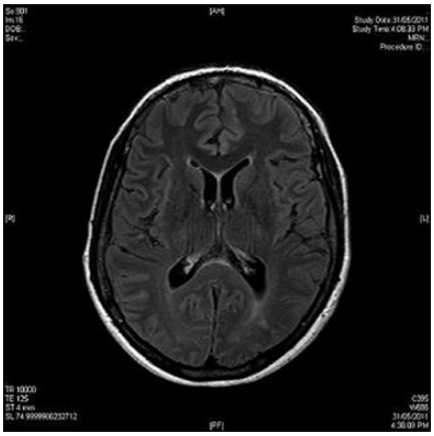

Figure 2: Repeat MRI Brain showing near complete resolution of abnormal high T2 signal in the cerebral hemispheres.

Although the exact pathogenesis of PRES remains unclear, vasogenic brain oedema secondary to hypertensive encephalopathy and disordered cerebral autoregulation play a major role. The posterior circulation, supplied by the vertebro-basilar system has poor sympathetic innervations and is therefore frequently affected. Vascular endothelial toxicity and endothelial dysfunction due to immunosuppression or inflammatory disease activity has also been proposed [1] and is supported by the description of PRES in normotensive patients [5].

Systemic lupus erythematosus (SLE) is a chronic multisystem inflammatory disease that follows a relapsing and remitting course. It is characterized by an auto-antibody response to nuclear and cytoplasmic antigens. SLE can affect any organ system, but mainly involves the skin, joints, kidneys, blood cells, and nervous system [4]. PRES can occur at anytime in an SLE patient, but the usual identified risk factors are hypertension, renal involvement and the use of immunosuppressive agents, particularly cyclosporine [5]. The mainstay of treatment is supportive control of BP and seizures, withdrawal of an offending drug and directed treatment of systemic SLE activity including corticosteroids and cyclophosphamide. The challenge of treatment thus is balancing the requirement for immunosuppression for management of disease activity against the risk of immunosuppression-associated disease or relapse.

This case demonstrates a very dramatic first presentation of SLE with neurological manifestations of PRES, rather than hypertensive encephalopathy with lupus cerebritis [8,9] occurring in the absence of accompanying lupus nephritis or other causative factors. In a case series by Leroux et al. [10] 91% of patients with SLE and PRES had associated lupus nephritis. Other published case series of PRES in SLE are consistent with the majority of patients having accompanying nephritis [5,8]. In a recent case series by Varaprasad et al. [11] PRES occurred as early as 1.5 months of initial diagnosis of SLE being made. Even in Baizabal-Carvallo et al. [12] review paper on PRES due to acute lupus activity, all 21 cases had known SLE at the time of presentation with PRES.

Conclusion

To our knowledge, this is the first reported case of PRES leading to a diagnosis of SLE. Given that our patient was treatment-naïve, this highlights the ability of active SLE to cause PRES in its own right, without the confounding causative role of immunosuppression and renal impairment.

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