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Case Report

Posterior reversible encephalopathy syndrome following septicemia in patient with myasthenia gravis ☆,☆☆

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ABSTRACT

Posterior reversible encephalopathy syndrome is a clinicoradiologic entity characterized by seizure, headaches, visual symptoms, impaired consciousness, and vasogenic cerebral edema of occipital and parietal lobes of the brain. Magnetic resonance imaging (MRI) is the diagnostic gold standard. The pathophysiology of posterior reversible encephalopathy syndrome is still unknown, but it is thought to be closely related to several medical conditions including hypertension, preeclampsia, eclampsia, immunosuppressive agents, transplantation, and sepsis. We report a rare case of posterior reversible encephalopathy syndrome in patient with myasthenia gravis and sepsis. A 22-year-old male was diagnosed with myasthenia gravis combined with sepsis due to pneumonia. During his recovery, the patient suffered multiple generalized convulsions and subsequent loss of consciousness. On cranial MRI, the abnormalities were observed with hyperintense within the subcortical white matter of the temporal, parietal, and bilateral occipital lobes on T2-weighted and T2 FLAIR. Reversibility of the symptoms and characteristic imaging findings led us to a diagnosis of posterior reversible encephalopathy syndrome. Early recognition and management of posterior reversible encephalopathy syndrome as a cause of encephalopathy in patients with septicemia and myasthenia gravis is necessary to prevent secondary complications in this condition.

Abbreviations: PRES, Posterior reversible encephalopathy syndrome; MRI, Magnetic resonance imaging; RT-PCR, Reverse transcriptase-polymerase chain reaction.

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Introduction

Posterior reversible encephalopathy syndrome (PRES) is characterized by the acute onset of several symptoms, including headache, convulsions, altered consciousness, and visual disturbances. Some patients present with acute hypertension [1,2]. The radiographic presentation is a prominent white matter vasogenic edema lesion in the brain's posterior circulation and parietal lobes [3]. PRES is also known by many other names, such as reversible posterior cerebral edema syndrome, posterior leukoencephalopathy syndrome, or brain capillary syndrome. PRES is the most commonly accepted nomenclature in medical literature nowadays [4]. Early diagnosis and treatment are essential to prevent secondary complications in this condition. Our understanding of PRES is still unclear, but it is thought to be closely related to several medical conditions including hypertension, preeclampsia, eclampsia, immunosuppressive agents, post-transplantation, renal disease, or even sepsis [5–7]. We introduce a rare case of PRES in a patient with septicemia and myasthenia gravis.

Case presentation

A 22-year-old male with no relevant personal history presented to the emergency department of local hospital with 2-week history of drooping eyelids, paralysis of upper limbs and lower limbs, double vision, rapidly progressive dyspnea, and respiratory distress. He was performed tracheal intubation and transferred to 108 Military Central Hospital.

At the time of presentation, his temperature was 36.4°C, heart rate of 97 bpm, blood pressure of 110/60 mmHg, respiratory rate of 21 breaths/min, and oxygen saturation of 98% on 45% oxygen (FiO_2 45%). He was alertness, well oriented. Repetitive nerve stimulation test revealed disorders of postsynaptic neuromuscular transmission typical for myasthenia gravis. Computed tomography (CT) of the thorax showed no thymoma. Reverse transcriptase-polymerase chain reaction (RT-PCR) testing showed negative for SARS-COV-2. He was managed with mechanical ventilation, pyridostigmine 60 mg every 6 hours, solumedrol 80 mg per day, and a typical course of five plasma exchanges with 3000 mL of fresh frozen plasma exchanged for each session. On the 10th day, the patient had fever and secretion of sputum. Chest X-ray showed pneumonia with the consolidation of right lower lung lobe. The cultures of blood and tracheal secretion were positive for *Staphylococcus aureus*. He was treated with vancomycin 1 g every 12 hours for 14 days. On the 21st admission day, his infectious condition and pneumonia improved. He was weaned from the ventilator. However, on the 23rd day, he suddenly had multiple generalized seizures, each lasting 3-5 minutes, followed by loss of consciousness. These seizures are repeated 2-3 times per day. There were no signs of meningeal irritation. The abnormalities observed on cranial MRI showed abnormalities in the white matter and the subcortical region of the temporal, parietal, bilateral occipital lobes, and posterior fossa with findings of increased signal on T2- FLAIR images and T2-weighted images (Fig. 1). Cerebrospinal fluid analysis revealed a slight increase in protein level (0.59 g/L) and normal white blood cells; Depakine 600 mg per day was administered. One week later, the patient's symptoms were controlled, and his consciousness recovered. The patient was discharged after that.

Discussion

PRES is seen in various clinical settings first recognized in the medical nomenclature in a 1996 case series. It is characterized by clinical manifestations and MRI lesions consistent with posterior cerebral white matter edema [3]. PRES is increasingly reported in case reports and case series. However, the

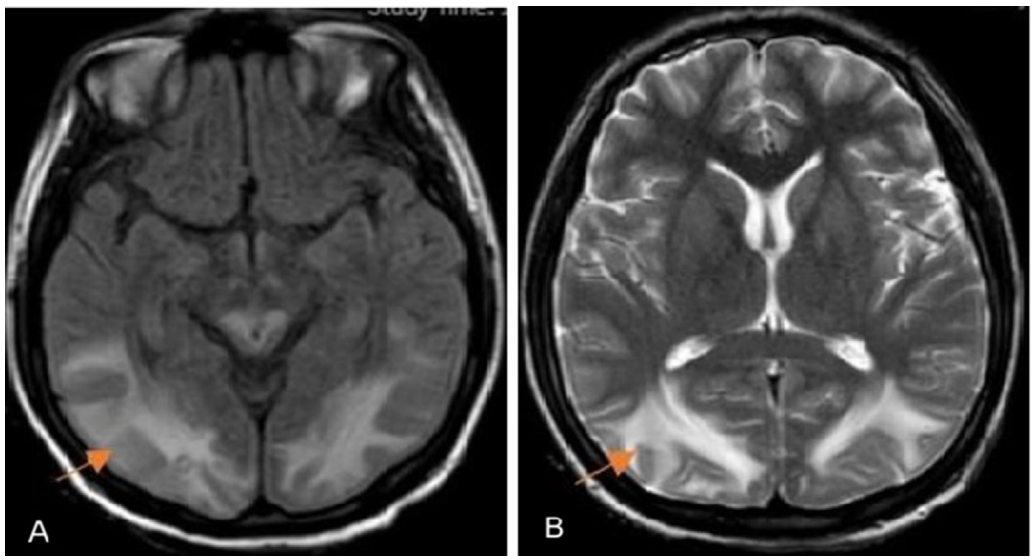

Fig. 1 – Hyperintensity in the occipital regions bilaterally on axial T2-FLAIR MRI image (A) and axial T2-weighted MRI image (B).

incidence of PRES is still unknown [8]. In addition, the pathogenesis of PRES is currently not clearly understood. Some suggested several mechanisms for PRES, including autoregulatory failure, hypertension leading to vasogenic edema, cerebral vasoconstriction with infarcts in the brain, and endothelial dysfunction that can lead to blood-brain barrier damage and capillary leakage, and the result is vasogenic edema [3,9]. Patients with PRES usually make a full recovery with proper treatment and diagnosis. However, there have recently been a few reports of persistent neurological damage and a mortality rate of approximately 15% [10,11]. Controlling high blood pressure is one of the essential treatments of PRES. When blood pressure is controlled slowly, the patient will usually improve markedly. Blood pressure goal should be achieved in 2-6 hours at a reduced rate not exceeding 25% of the presenting systolic blood pressure [12]. Target blood pressure should be assessed in conjunction with the patient's clinical recovery [13]. Patients with epileptic manifestations should receive antiseizure medication immediately [14]. Dexamethasone has been used to treat patients with PRES because it seems to improve vasogenic edema, but there is not enough strong evidence for this usage [13].

Approximately one-third to one-half of patients with PRES are associated with autoimmune diseases including systemic lupus erythematosus, thrombotic thrombocytopenic purpura, inflammatory bowel disease, rheumatoid arthritis and, neuromyelitis disorders [15]. To the best of our knowledge, this is the rare case of PRES in patient with **s**epticemia and myasthenia gravis.

Conclusions

Early recognition and management of PRES can reduce morbidity, mortality and may lead to early recovery, especially in patients with **s**epticemia and myasthenia gravis.

Patient consent

A written consent was obtained from the patient for publication of this case and any accompanying images.

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