Case Report

Posterior reversible encephalopathy syndrome following paroxysmal nocturnal hemoglobinuria: a case report and literature review

Dongxue Ding, Kai Li, Guoliang Li, Xiaoyan Long

Department of Neurology, Xiangya Hospital, Central South University, Xiangya Road, Changsha, China

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Abstract: Paroxysmal nocturnal hemoglobinuria (PNH) is a rare acquired disorder characterized by hemolytic anemia, marrow failure, and a high incidence of life-threatening venous thrombosis. It is subject to a considerable variety of complications like intestinal obstruction and visceral embolism. The current study firstly presents a 40-year-old male with a previous diagnosis of PNH who developed posterior reversible encephalopathy syndrome (PRES) during treatment with methylprednisolone. He was referred to our department with headache and two episodes of generalized tonic-clonic seizures. Laboratory examination revealed peripheral blood cytopenias and elevated count of reticulocyte. Brain magnetic resonance imaging (MRI) exhibited abnormal signal in the bilateral parieto-occipital lobes with symmetric distribution which confirmed the diagnosis of PRES. After receive treatment of dexamethasone, anti-hypertensive and neurotropic drugs, the patient made a complete clinical recovery; and the abnormal signals of MRI were almost completely absorbed. This case shows that PRES might be a rare complication of PNH. Furthermore, it points out the necessity of rapid diagnosis and treatment of PRES.

Keywords: Paroxysmal nocturnal hemoglobinuria, posterior reversible encephalopathy syndrome, seizures, methylprednisolone, magnetic resonance imaging

Introduction

PNH is a rare acquired hematopoietic stem cell disorder which manifests with hemolytic anemia, thrombosis, bone marrow failure and peripheral blood cytopenias [1]. It is subject to a considerable variety of complications like intestinal obstruction and visceral embolism [2, 3]. Here we firstly describe a 40-year-old male with a previous diagnosis of PNH who developed posterior reversible encephalopathy syndrome (PRES) during treatment with methylprednisolone.

PRES was first described by Hinchen et al in 1996 [4]. Clinically it is marked by typical symptoms including headache, seizures, visual disturbances, altered mental status, nausea, vomiting, and focal neurological signs and accompanied by a typical computed tomography (CT) or MRI pattern [4]. The causes of PRES are varied and have been largely attributed to hypertension, eclampsia, malignancy, renal insufficiency, organ transplantation and the use of immune-suppressants [5, 6]. Although clinical manifestations of PRES are usually reversible and patients with PRES are mostly have a good prognosis, early diagnosis and treatment is essential as it may prevent progression to irreversible brain damage.

Case report

A 40-year-old man presented to the emergency department after headache and two episodes of generalized tonic-clonic seizures at home. In his medical history, he had been diagnosed with paroxysmal nocturnal hemoglobinuria (PNH) for 8 years and slightly hypertension for 2 years. During the past 8 years, he has consecutively taken methylprednisolone orally (5 mg/d) to control the disease progression of PNH. A month ago, he was diagnosed as left lower limb deep venous thrombosis, as symptom progressing, he had to accept an amputation surgery consequently.
Physical examination at the onset revealed blood pressure of 159/93 mmHg, heart rate of 69 beats/min; consciousness; equally big and round pupils with sensitive light reflex; no evidence of focal neurological deficit and meningeal signs were got by neurological examination. His laboratory examination showed hemoglobin of 77 g/L, a total white blood cells (WBC) count of 3.5×10^9/L, and platelet count of 83×10^9/L. The patient was negative for direct Coomb’s test and corrected reticulocyte was 3%. His serum biochemistry results and serum immunity indicator levels were within normal limits except that complements were low (C3: 664 mg/L; C4: 216 mg/L). Serology for human immunodeficiency virus (HIV), syphilis, Epstein-Barr virus, cytomegalovirus, hepatitis A, B, C viruses, and tubercle bacillus were negative. Cerebral fluid (CSF) studies revealed clear and colorless fluid with normal opening pressure, cell counts, glucose and protein contents. Evaluations for infection, including virus, Tuberculosis, Cryptococcus, Gram’s staining, India ink stain and cultures were unrevealing.

Brain MRI examination showed high signals on T2, fluid-attenuated inversion recovery (FLAIR) MRI, diffusion weight imaging (DWI) and appar-
PRES induced by PNH

The patient was diagnosed with PNH 8 years ago. His symptoms were well-regulated by a low dose glucocorticoid treatment. Further research is needed to study the association between PNH and PRES.

Although reversible by definition, if the patient did not undergo early recognition and timely treatment, irreversible brain damages may occur. Our patient's MRI revealed increased signal in the bilateral parieto-occipital lobe on T2-weighted, FLAIR, DWI and ADC sequences. These findings were consistent with PRES [15], attributable to a manifestation of vasogenic and cytotoxic edema. Treatment of PRES includes better blood pressure control, withdrawal/decreased doses of the offending medications, and seizure management [16]. In our patient, the antiepileptic drugs were not used and we continue the glucocorticoid treatment. Following the treatment mentioned above, his neurological complaints improved. We thus conjecture that PRES should be a complication of PNH, as well as thrombosis. To our knowledge, our patient was the first patient to develop PRES associated with PNH.

In conclusion, PRES is a rare complication of different diseases or some special treatments; it also can be followed by PNH. The diagnosis of PRES is made by recognition of the clinical manifestation, Imaging feature, especially MRI. MRI is very helpful to rule out alternative causes and to confirm the diagnosis. In order to prevent progression to permanent neurological damage and death, the underlying cause should be addressed, and treatment should be initiated as soon as possible [17].

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Disclosure of conflict of interest

None.

Address correspondence to: Xiaoyan Long, Department of Neurology, Xiangya Hospital, Central
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South University, Xiangya Road, Changsha, China.
E-mail: longxyan@sina.com

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