e - ISSN - 2349 - 8005

INTERNATIONAL JOURNAL OF ADVANCES IN CASE REPORTS

Journal homepage: www.mcmed.us/journal/ijacr

AN UNUSUAL CASE OF POSTERIOR REVERSIBLE ENCEPHALOPATHY SYNDROME IN PATIENT WITH AUTOIMMUNE HEMOLYTIC ANEMIA

Sagar Sunka, Pradeep M. Venkategowda, S Manimala Rao*, Jaydeep Ray Chaudhary

Department of Critical Care Medicine, Yashoda Multi-Speciality Hospital, Somajiguda, Hyderabad, Telangana, India 500082.

Corresponding Author:- **S Manimala. Rao**
E-mail: manimalarao@hotmail.com

Article Info
Received 15/04/2015
Revised 25/04/2015
Accepted 06/05/2015

Key words:
Autoimmune, Cyclophosphamide, Hypertension, Hemolysis.

ABSTRACT
Posterior reversible encephalopathy syndrome (PRES) commonly seen in hypertensive patients. It is due to failure of auto regulatory mechanism of posterior circulation of the brain due to acute rise in blood pressure (BP) leading to cerebral edema. This is a case report of 18 years old female a known case of autoimmune hemolytic anemia (AIHA)-cold agglutinin type who came to our hospital with history of headache and generalized tonic clonic seizures for 8-10 minutes. MRI brain showed bilateral asymmetric hyperintense lesions in parieto-occipital area and left fronto-temporal area. In view of above clinico-radiological entity, PRES due to AIHA was diagnosed and managed successfully. This case report mainly highlights that PRES can occur even with AIHA which is rather unusual entity.

INTRODUCTION

Posterior reversible encephalopathy syndrome also known as reversible posterior leukoencephalopathy syndrome. Initially described by Hinchey *et al* [1] as clinico-neuroradiological entity characterized by headache, nausea, vomiting, altered mental status, blurring of vision and seizures. Radiological features are edema in the white matter of the brain perfused by posterior circulation. The causes are hypertension (most common), preeclampsia, HELLP syndrome, immunosuppressive / cytotoxic drugs, sepsis, systemic lupus erythematosis and nephrotic state.

CASE REPORT

This is a case report of an 18 year-old female, a known case of autoimmune hemolytic anemia, cold agglutinin type, diagnosed 4 months previously. She came to our tertiary hospital emergency department (ED) with history of headache, nausea followed by generalized tonic clonic seizures for 8-10 minutes. In the ED she was drowsy, her pupil were bilaterally equal and reactive, no neck stiffness and moving all four limbs. Funduscopic examination was normal. She was afebrile with a pulse rate of 98 beats/min, blood pressure was 108/50 mmHg, on auscultation, bilateral normal vesicular breath sounds were heard.

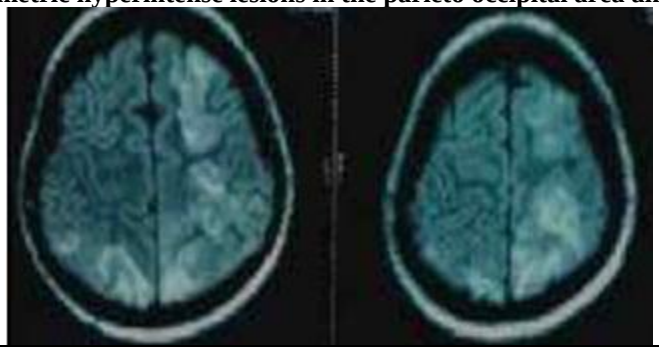
She had one more episode of seizures. She received 4mg lorazepam IV followed by 1000mg phenytoin IV over 20 minutes. She was endotracheally intubated and mechanically ventilated. The patient was transferred to ICU for further management.

Laboratory studies showed hemoglobin 9.4 gm/dl, WBC 4400 cells/mm^3 (neutrophils- 58%), platelets 2 lakhs/cumm. Arterial blood gas analysis showed pH-7.32, PO_2-286, PCO_2-38, and HCO_3-22. Blood sugar was 134 mg/dl. Serum electrolytes, liver function test, kidney function test and routine urine analysis were normal. CSF showed 2 cells with 100% lymphocytes with normal protein and sugar. Procalcitonin was <0.5 ng/ml. Blood, urine and endotracheal cultures were negative for any growth. EEG was normal. Chest x-ray, ECG and 2D-echocardiogram were also normal. MRI of the brain

showed bilateral asymmetric hyperintense lesions in the parieto occipital area and left fronto-temporal area (Figure-1).

In view of the clinical and radiological finding, posterior reversible encephalopathy syndrome (PRES) due to autoimmune hemolytic anemia was diagnosed. The patient was managed with supportive care (mechanical ventilation, phenytoin, pantoprazole, enoxaparin) along with once daily dose of tablet cyclophosphamide 100mg and prednisolone 20mg. Her level of consciousness improved and she was extubated on the 3rd day, and discharged home on day 6.

Fig 1. Showing bilateral asymmetric hyperintense lesions in the parieto occipital area and left fronto-temporal area



DISCUSSION AND CONCLUSION

Posterior reversible encephalopathy syndrome (PRES) is due to failure of auto regulatory mechanism of posterior circulation of the brain due to acute rise in BP leading to cerebral edema. The global incidence of PRES is unknown and reported in patients aged 4 to 90 years with marked female predominance.

Two theories have been postulated in the development of PRES. Vasogenic edema theory is due to failure of compensatory mechanism of the CNS to limit the blood flow in acute hypertension [2] leading to damage of blood-brain barrier (BBB) causing vasogenic edema in the brain. This can be seen in any part of the brain but more common at posterior circulation. Secondly the cytotoxic theory which states that acute rise in BP produces hypoperfusion leading to hypoxia resulting in endothelial damage with subsequent edema.

The various causes of PRES are hypertension, preeclampsia, HELLP syndrome, immunosuppressive / cytotoxic drugs, sepsis, systemic lupus erythematosis and nephrotic state. Sepsis and septic shock associated with PRES due to endothelial derangement and microcirculation disturbances. Autoimmune disease has been encountered in 8 % to 10 % of cases. Systemic lupus erythematosis [3], Systemic sclerosis [4], Polyarteritis nodosa [5], Wegener's granulomatosis [6], Thrombotic microangiopathy [7], Polyangiitis [8], Takayasu arteritis [9] and Crohn's disease [10] are some of the causes of PRES. Our patient had AIHA and the occurrence of PRES in this disease is unusual.

Clinical features are headache, nausea, vomiting, altered mental status, blurring of vision, seizures, lethargy, stupor, somnolence and coma. Signs of motor dysfunction such as hemiparesis, dystonia and dysmetria may be present. If brain stem involved then patient can manifest with dyspnea, anarthria and dysphagia. Complications of PRES are cerebral infarction, cerebral hemorrhage and cerebral herniation, coma and status epilepticus,

There are no consensus guidelines for diagnosis of PRES. The combination of clinical manifestations and radiological criteria establishes the diagnosis. Magnetic resonance imaging (MRI) is the imaging of choice [11]. The four radiological patterns of PRES have been described, such as (a). Holohemispheric watershed pattern (23 %), (b). Superior frontal sulcus pattern (27 %), (c). Dominant parietal-occipital pattern (22 %), and (d). Partial or asymmetric expression of the primary patterns (28 %). Our patient had dominant parieto – occipital pattern of PRES on MRI as shown in figure-1.

Treatment is mainly supportive, early diagnosis and treatment to avoid irreversible damage to brain. Close monitoring of cardiac, respiratory and neurological status along with removal of triggering agent. In case of high BP induced PRES, mean arterial pressure (MAP) should be reduced not more than 20-25% within the first 2 hours [12]. In case of Preeclampsia related PRES, prompt delivery along with supportive measures are required. Our patient had AIHA hence treatment of this disease (Tablet cyclophosphamide 100mg OD and Tablet prednisolone 20mg OD) was the key to the successful management of this condition. This case report mainly highlights the occurrence of PRES in autoimmine hemolytic anemia which is unusual.

ACKNOWLEDGEMENTS

We gratefully acknowledge Dr.Ganesh Jaishetwar (Hemato - oncologist), Respiratory therapists, Physiotherapists, nurses and management of the hospital for their valuable support.

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