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Posterior reversible encephalopathy syndrome [PRES] - A rare and reversible cause of vision loss

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Abstract

Posterior reversible encephalopathy syndrome [PRES] is a reversible clinico-radiological syndrome characterized by headache, seizures, altered consciousness and visual disturbances. It has association with many causes. A significant relationship with acute hypertension is found. Multiple theories are proposed for explaining pathophysiology. MRI is the gold standard for diagnosis. Early detection and immediate treatment helps in full recovery. Our aim is to discuss PRES in detail with the help of a case of a young lady with pre-eclampsia diagnosed with PRES. Reversibility of clinical features and characteristic MRI findings lead to the diagnosis. Patient fully recovered with conservative management. PRES is a rare but possible cause of vision loss.

Keywords: PRES; reversible; vision loss; pre-eclampsia

1. Introduction

Posterior reversible encephalopathy syndrome (PRES) is a clinical syndrome with characteristic radiological features. Common symptoms of PRES are headache, seizures, altered consciousness and visual disturbances ^[1]. Posterior reversible encephalopathy syndrome (PRES) was first described by Hinchey *et al* in 1996 ^[2]. Previously various names have been used viz., reversible posterior leukoencephalopathy syndrome, reversible posterior cerebral edema syndrome and reversible occipital parietal encephalopathy. But now PRES is the most widely accepted and commonly used term ^[3]. A significant relation is associated with acute hypertension, but it's not invariably present ^[1]. PRES is associated with pre-eclampsia/ eclampsia, acute and chronic renal diseases, haemolytic uremic syndrome, autoimmune diseases, use of cytotoxic and immunosuppressant drugs, blood transfusion, electrolyte disturbances, cancer chemotherapy, infection /sepsis ^[2, 4].

2. Materials and Methods

Proper and clear history was taken from the patient. Thorough clinical bedside examination was performed initially, with full-fledged slit lamp examination performed later. Ophthalmological findings are documented in the form of fundus photo and Spectral domain optical coherence tomography [SD-OCT]. To confirm the clinical diagnosis, different investigations including MRI Brain has been performed. Proper follow ups of the patient were done to evaluate the progress.

3. Case History

A 28-year-old primi with twin pregnancy presented to labor room in latent phase of labor. Her first and second trimesters were uneventful. On examination, her BP was 180/120mm Hg. She was started on Tab Nifedipine 20mg twice daily, Tab Labetalol 100 mg thrice daily, Inj Mannitol 100 cc intravenous. She was taken up for emergency lower segment Caesarean section under spinal anesthesia and she delivered two baby girls who were healthy. She complained of blurring of vision and headache on her first post-operative day. She has been referred to the department of Ophthalmology. There was sudden onset of bilateral loss of vision associated with headache. There was no history of altered mental status, seizures, loss of consciousness. Not a known case of diabetes, hypertension, epilepsy, cardiac disease, chronic renal disease or autoimmune disease. No history of allergies.

4. Results

On bedside examination on post-operative day 1; patient was conscious and coherent with blood pressure of 150/110 mm Hg. Systemic examination was normal. On bedside ocular examination, there was no facial asymmetry, visual acuity was perception of light with doubtful projection of rays in both the eyes extra ocular movements were full, free and painless in all direction. Anterior segment was within normal limits with both direct and consensual light reflexes intact. On dilated fundoscopy, the media was clear, disc was pink in colour with normal shape, size, defined margins and cup:disc ratio being 0.3:1. Macula was normal with no haemorrhages or cotton wool spots. Retinal vessels were within normal limits. [Fig 1]

Complete blood count, Renal function tests, liver function tests were found to be normal. CT Brain was done and it was found to be normal. MRI Brain showed multiple cortical and sub cortical white matter hyper intensities in bilateral cerebral hemispheres predominantly in parietal, temporal and occipital lobes and bilateral cerebellar hemispheres, most likely Posterior reversible encephalopathy syndrome. Fig 2]

Patient was reviewed in Ophthalmology outpatient department on post-operative day 4. On examination, blood pressure was 140/90 mm Hg (under medication); visual acuity was counting finger close in both the eyes; pupils were normal, round, regular and reactive in both eyes. Anterior segment and dilated fundoscopy of both eyes were within normal limits. SD-OCT was within normal limits in both eyes. [Fig 3] She was reviewed again in Ophthalmology outpatient department on post-operative day 10 just before her discharge. On examination, her blood pressure was 120/80mmHg under medication. Distant vision was 6/9 in both eyes; near vision was N6 in both eyes. Color vision was normal in both eyes. On slit lamp examination, anterior segment and dilated fundus were normal. Patient denied HVF Perimetry and Check MRI.



Fig 1: Fundus photo of right and left eye showing normal fundus

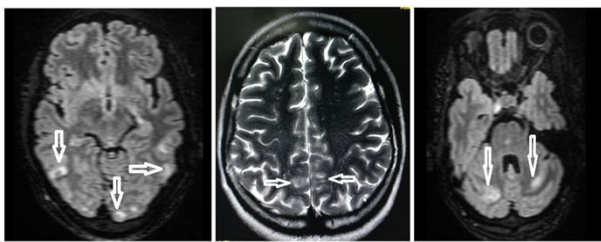


Fig 2: MRI Brain showing multiple cortical and subcortical white matter hyperintensities in bilateral cerebral hemispheres predominantly in parietal, temporal and occipital lobes and bilateral cerebellar hemispheres

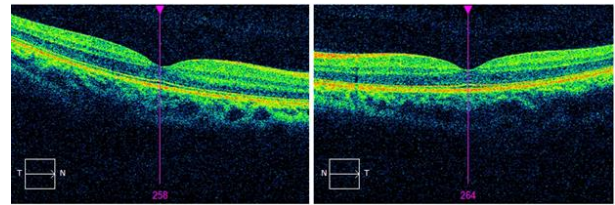


Fig 3: SD-OCT image of both eyes showing no abnormality

5. Discussion

As the name indicates it is a reversible neurological entity. Its characteristic symptoms being headache, visual disturbances, altered mental status, seizures and unconsciousness [5]. It is a rare condition with unknown exact incidence [6]. There is no age specification in its incidence. Females are commonly affected owing to the fact that one of the common causes of PRES is pre-eclampsia/eclampsia developing during pregnancy [7]. Different theories have been proposed to explain the pathophysiology of PRES: i) the most widely accepted theory is cerebral vasoconstriction in response to acute hypertension [8]. ii) Vasogenic edema as a result of hyper perfusion and toxins that cause endothelial injury and disrupt the blood brain barrier [9]. iii) Activation of cellular immune system that accompanies vasoconstriction contributes to delayed hypoperfusion and edema [10]. Hemianopia, Visual neglect, auras, visual hallucinations, and cortical blindness are the commonly encountered visual abnormalities seen in PRES [11]. The conditions to be considered while dealing with PRES are stroke, meningoencephalitis, demyelinating lesions of the brain and cerebral venous thrombosis. MRI is the gold standard for diagnosis with predominant lesions in the posterior sub cortical white matter, especially in the occipital and parietal lobes of the brain [12]. In our patient, it is pre-eclampsia induced PRES, hence the key treatment is to control hypertension, prevention or treatment of seizure and termination of pregnancy. Corticosteroids are choice only used in selected cases [5]. With correct diagnosis and timely control of hypertension, the patient prognosis is usually good. Time period to regain visual loss is usually of 4 hours to 8 days after treatment [11]. Delayed recognition of PRES can lead to secondary complications, of which status epilepticus, intracranial haemorrhage and ischemic infarction are considered common [13]. Recurrence rate is 5-10% [14].

6. Conclusion

Posterior reversible encephalopathy syndrome [PRES] is a reversible and relatively rare condition with good prognosis. The key to full recovery of PRES is early diagnosis and treatment. Misdiagnosis and delayed treatment may cause permanent neurological damage and even death.

7. References

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