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Case Report

Posterior reversible encephalopathy syndrome (PRES): A case report

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ABSTRACT

Patients with preeclampsia or eclampsia may be associated with hypertension, seizures and other neurologic symptoms and deficits. A young female patient presented with a neurological and visual disturbance which included variable symptoms like visual disturbances, headache, seizures and altered consciousness or mental status with a history of postpartum eclampsia after an uneventful LUCS. She also gave a history of a two-bag blood transfusion one month ago. A neurological and ophthalmic evaluation was performed. Detailed history taking and clinical evaluation followed by an imaging study, potentially magnetic resonance imaging (MRI), helped to confirm the diagnosis of Posterior reversible encephalopathy syndrome (PRES). PRES is a clinico-neuroradiological disorder of neurotoxicity that typically involves headache, mental confusion, seizures, and occasionally loss of vision. The exact pathophysiology of PRES is still unclear and has not been thoroughly explained. Hypertension and endothelial cell injury may be pathognomic. Prompt management may help to recover early.

1. Introduction

Posterior reversible encephalopathy syndrome (PRES) is a clinico-neuroradiological disorder characterized by symptoms including headache, seizures, altered consciousness or mental status and visual disturbances.[1] PRES was first described by Hinchey et al. in 1996.[2] Shortly after the description in 1996, two cases were published after a short period of the PRES reported in 1996.[3] Before this, the syndrome had been reported by various names (reversible posterior leukoencephalopathy syndrome, reversible occipital parietal encephalopathy, reversible posterior cerebral oedema syndrome). PRES is now the accepted term worldwide.[4] It is usually but not always associated with acute hypertension.[1] This clinical syndrome is commonly recognized because of the improvement and availability of brain imaging. The primary clinical conditions associated with PRES are preeclampsia, eclampsia, autoimmune disorders (like SLE, rheumatoid arthritis, Crohn's disease), acute or chronic renal diseases, massive blood transfusion, liver failure or transplantation, immunosuppressive or cytotoxic drugs, high dose steroids, cancer chemotherapy and hypertensive encephalopathy.[5,6] There is a wide variation in the severity of clinical symptoms; for example, visual disturbance can vary from blurred vision and homonymous hemianopia to cortical blindness. Altered consciousness may present as mild confusion or agitation, even coma. Nausea, vomiting, and the features of brainstem deficits may also present. Seizures and status epilepticus are usual symptoms, while non-convulsive status epilepticus is more common than

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generalized status epilepticus. Non-convulsive status should be cautiously evaluated in patients with prolonged altered consciousness, which may mistake as postictal confusion. Signs reported as stereotypic movements like staring, head turning or blinking eyes. Postictal confusion usually lasts for a few hours, but PRES and non-convulsive status may last for many days and can be mistaken for psychosis, drug intoxication, or psychogenic states. If PRES is diagnosed and treated early, the clinical syndrome can commonly resolve within a week. Here, we address a rare case on PRES reported in ophthalmic department.

2. Case Report

A 25-year-old female came to the Ophthalmology department of the Anwer Khan Modern Medical College & Hospital, Dhaka, Bangladesh on 7th July 2022 with complaints of visual disturbance for five days. According to the patient's statement, she was pregnant (Twin pregnancy with breech presentation) for 36 weeks, and she underwent lower uterine cesarian section (LUCS) on 20th June 2022 at the same hospital. Her per-operative period was uneventful with normotensive status. Still, she developed postpartum eclampsia on her 3rd POD with complaints of headache, high rise of BP (180/110 mm hg), restlessness, recurrent convulsion attacks and altered mental condition like acting violently to others (attacking and saying unpleasant things). Then she was transferred to the ICU on 23rd June 2022 for further management. After six days, depending on her physical and mental condition, she was shifted to her cabin; after improving her mental condition, she complained of visual disturbance for five days. Three days after shifting in the cabin, she describes her visual disturbances as she sees dirty things all over her bed, and her surroundings in the room are flooded when her baby urinates. She feels like sitting on the ceiling though she is on the bed. On 4th day her previous visual disturbances declined, and she complained of gradually fading colors to white towards the left side of the visual field when both eyes are open. The exact visual acuity could not be measured due to her altered mental consciousness.

She also gives a history of skin allergy and two bags of blood transfusion one month back. Her maternal period was uneventful. No history of preeclampsia, Gestational Diabetes mellitus, or maternal infection. No history of neonatal infection. Both the babies are in good health, and there is no history of anti-hypertensive, anti-diabetic, anticonvulsant, cytotoxic, or any pregnancy-contraindicated drugs.

On general examination, anaemia, jaundice, cyanosis, oedema, dehydration, clubbing, koilonychia, and leukonychia are all absent. Her pulse was recorded as 84 b/m, her blood pressure was 110/70 mm of Hg, and her temperature was reported as 98°F.

On the nervous system examination, she was well-oriented, and no abnormalities were found. Ocular examination revealed left homonymous hemianopia with dyschromatopsia (Table 1).

Table 1: Ophthalmic features of the patients with PRES

	Right eye	**Left eye**
Nystagmus	Absent	Absent
Ocular motility	Full in all cardinal gaze	Full in all cardinal gaze
Conjunctiva	Normal	Normal
Cornea	Normal	Normal
Pupil	Round, regular and reacting to light	Round, regular and reacting to light
Fundus	Normal	Normal
Visual Field	Loss of nasal field	Loss of temporal field
Color vision	Fades to white in nasal visual field on closure of left eye	Fades to white on temporal visual field on closure of right eye.

On cardiorespiratory, abdominal, and renal system examination, no abnormalities were found.

We have investigated CBC, RBS, SGPT, BT, CT, SGPT, Serum Creatinine, Serum urea, Urine R/M/E, ECG, and MRI of the Brain with MRV and Covid-19 RT PCR test. All the test report findings are in the normal range except for the Brain MRI, which shows Multiple variables sizes hyper signals in both basal nuclei, cortical and subcortical white matter of both cerebra, primarily posterior parieto-occipital regions (Figure 1). MRV findings were normal.

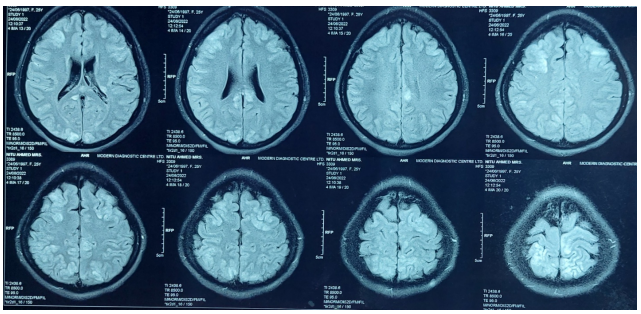

Fig. 1: Brain MRI showing multiple variables sizes hyper signals on Flair in both basal nuclei, cortical and subcortical white matter of both cerebrums primarily posterior parieto-occipital regions.

Depending upon the patient's history, clinical examination, and investigation findings, our diagnosis is posterior reversible encephalopathy syndrome (PRES).

3. Discussion

Posterior reversible encephalopathy syndrome (PRES) is a neuroradiological disorder of reversible vasogenic subcortical oedema without infarction, usually in a parieto-

occipital lobe pattern that manifests in headache symptoms, visual loss, seizures, variation of consciousness, and focal deficits. About 92-98% of patients with eclampsia revealed PRES on neuroimaging.[7–9] The exact pathophysiological mechanism of PRES is uncertain.

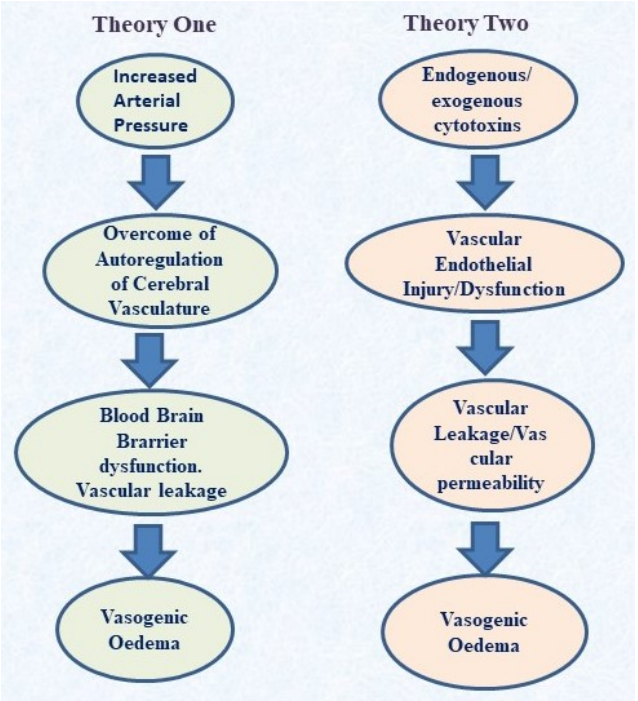

Fig. 2: Major theories of the pathophysiology of PRES

Two major and opposite controversial hypotheses are commonly described in the literature (Figure 2). The most popular theory suggests that severe arterial hypertension exceeds the limits of the autoregulation of the cerebral blood vessels, which leads to the compromise of the blood-brain barrier and vasogenic oedema.[8,10,11] The upper limit of autoregulation of the cerebral vasculature varies from 150-160 mm Hg. However, this range can extend to 30 mm Hg higher in acute sympathetic states. The brain area supplied by the posterior cerebral artery involves more than the area supplied by the middle and anterior cerebral arteries in the sudden rise of blood pressure. Because there are more rich sympathetic innervations of the internal carotid artery than the vertebral artery, raised blood pressure can be more easily regulated by the middle and anterior cerebral arteries than the posterior cerebral artery, leading to endothelial cell injury. The second theory suggests the fact that 15-30% of Patients with PRES may be normotensive or hypotensive during initial evaluation. They are not associated with accelerated hypertension, and not related to overcome the limits of autoregulation of the cerebral vasculature.[11] Vascular endothelial dysfunction caused by endogenous or exogenous cytotoxins is the main culprit according to the second theory. Post-transplant patients with immunosuppressive agents, patients with chemotherapeutic agents, sepsis may be associated for producing circulating toxins which cause vascular injury. Endothelial dysfunction allowing for increasing vascular permeability and ultimately develop vasogenic oedema. Another theory is related to theory one that suggests elevated hypertension leading to cerebral autoregulatory vasoconstriction, ischemia, and subsequent brain oedema.[7,10,12–14]

Clinically PRES may be presented with several types of clinical signs and symptoms. Approximately 50 to 80 per cent of PRES patients are encephalopathic, headache (50-87%) is the most common symptom; others include seizures (60-70%), altered mental status (51%), visual disturbances (34%), nausea/vomiting (10-19%), and 5-5% with status epilepticus.[8,13] Severe systolic hypertension may be associated with about 50% of patients with PRES.[9] Encephalopathy in patients may range from stupor to comatose stage. Seizures are often generalized, and status epilepticus may also manifest, but this is less common. Acute encephalopathy syndrome includes confusion, headache, vomiting, altered consciousness, and visual problems (blurred vision, hemianopsia, or complete loss of vision), which may lead to the clinician for place PRES in a differential diagnosis.

In a study on 47 patients with eclampsia, 98% of neuroimaging had changes consistent with PRES.[8] Another study showed that MRI had changed with PRES in 92% of patients with eclampsia, compared to only 19% of patients with preeclampsia and other neurologic disorders.[15]

MRI is beneficial in the diagnosis of PRES. Radiographic stigmata of the disease usually involve raised signal on T2 weighted image and T2 FLAIR (fluid-attenuated inversion recovery) sequences of subcortical white matter with vasogenic oedema predominantly include the parieto-occipital and posterior temporal lobes of both cerebrums of the Brain.[16] However, tissues of the other structures like as the anterior part of the cerebrum, deep white matter, brainstem, and cerebellum, may also be involved. The following three primary variations of radiological findings exist in about 70% of patients: a parietooccipital pattern, a holohemispheric watershed pattern, and a superior frontal sulcus pattern.[10] The involvement of the Frontal and temporal lobe can be seen in up to 75% of cases. Similarly, oedema can be affected the basal ganglia and the brainstem in up to 33% of cases and the cerebellum in up to 50%.[10] Finally, intracranial hemorrhage is common, complicating 10 to 25 per cent of cases. Intra-parenchymal bleeding is the most common type of intracranial hemorrhage, and subarachnoid hemorrhage is the second most common type. The prevalence of intracranial hypertension in PRES is 19.4%, and both subarachnoid hemorrhage (SAH) and Intraparenchymal hemorrhage (IPH) can occur in association with PRES.[17]

Radiologically any disturbance on parieto–occipital region leads to visual disturbance, which is seen in PRES as our visual pathway passes through this region. About 33% of patients are found suffering from visual disturbances in PRES. On the visual pathway after the optic chiasma, the optic tract goes to the thalamus's lateral geniculate body [LGB] situated above the midbrain. It forms a connection with the occipital cortex via optic radiation. In humans, the LGB is usually described as having six distinctive layers. The inner two layers (1 and 2) are magnocellular cell (M cell) layers, while the outer four layers (3,4,5 and 6) are parvocellular cell (P cell) layers. The neurons of these two layers carry various visual information; for example, P cells are concerned with a point-to-point vision or shape of objects, as well as color vision, and M cells are concerned with the location, position, and motion of objects. So, any disturbance around LGB may result in visual agnosia, dyschromatopsia, dyslexia, prosopagnosia, and schizophrenia.[18–20] Nerve fibers from the LGB form a connection with the primary visual cortex on the occipital lobe with the help of Optic radiation which acts as a connecting pathway. Disturbance in optic radiation results in homonymous quadrantanopia (superior or inferior) or homonymous hemianopia. Involvement of the occipital lobe may cause visual aphasia and temporary cortical blindness. As PRES is a reversible condition, visual disturbances caused by it are not permanent. It resolves gradually depending on the patient's recovery. The management of the PRES is mainly aimed at controlling the primary etiology causing PRES (44). For example, in cases of raised arterial pressures, treatment is aimed for correcting the raised blood pressures in a controlled environment. Always aware to drastically lowing the blood pressure to avoid for ischemic cerebral disease. Anticonvulsant medications may be used as adjunct therapy. Magnesium sulfate may use for seizure prophylaxis as hypomagnesemia is also one of the findings. Patients with PRES induced by chemotherapeutic or other immunosuppression agents, the better option is to taper or discontinue the medications.[21–24] PRES may recur in 5-10% of patients, usually in association with uncontrolled hypertension.[15,21]

4. Conclusion

PRES has been increasingly recognized in recent years and has been the cause of recurrent physician consultations for obstetric pre-eclamptic and eclamptic cases, along with ophthalmic and neurological consultations. In most patients, persistently increased blood pressures remain the chief culprit for the clinical symptoms and neurological deficits. Early diagnosis by Neuroradiological imaging (MRI) and differentiation from other causes of altered sensoria, such as seizures, meningitis, and psychosis, is essential to initiate treatment and prevent further complications. Reduction of blood pressure and seizure control remain the mainstays of therapy after prompt stabilization of the patient and removal of any known toxic insult. Although PRES is resolving successfully in most cases and carries a favorable outcome, patients with improper therapeutic support or delay in treatment may not project a positive outcome.

5. Declaration of Patient Consent

We have obtained informed written consent from the patient. All the images and other clinical information to be reported in the journal with the patients consent. The patient understand that her name and initials will not be published, and due efforts will be made to conceal her identity and gave her consent.

6. Source of Funding

None.

7. Conflict of Interest

None.

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