Transient cortical blindness at delivery-an unusual case report on posterior reversible encephalopathy syndrome

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ABSTRACT

Background: Posterior reversible encephalopathy syndrome (PRES) is an unusual clinical entity seen associated with blood pressure (BP) fluctuations presenting with acute neurological symptoms. This is an unusual case of PRES due to severe pre-eclampsia, with clinical presentation of cortical blindness and hypertension, with absence of headache or seizures.

Case Presentation: A primigravida, with no significant medical history, presented with significant proteinuria and worsening generalized itching and normotensive at 37 weeks 6 days of gestation. Due to elevated urate and raised urine protein induction of labor was planned. Ten minutes after vaginal delivery, patient reported sudden loss of vision with only being able to perceive light. She developed hypertension, hyperreflexia, and clonus. Aggressive management for severe pre-eclampsia was initiated. Her vision started improving as soon as her BP started normalizing. Imaging revealed the possibility of PRES with a superimposed focus of acute ischemia. She was discharged on antihypertensives and followed up by renal physicians. Within 2 months she recovered very well.

Conclusion: This is an unusual case of PRES due to severe pre-eclampsia, with clinical presentation of only cortical blindness with absence of headache or seizures. Prompt recognition and aggressive management of severe pre-eclampsia resulted in resolution of symptoms.

Keywords: Posterior reversible encephalopathy syndrome, pre-eclampsia, cortical blindness, hypertension, headache, pregnancy.

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Background

Posterior reversible encephalopathy syndrome (PRES) is an unusual and not a clearly understood clinical syndrome. Patients present with acute neurological symptoms with fluctuations of blood pressure (BP), as in preeclampsia or eclampsia, renal failure, use of cytotoxic drugs, and autoimmune conditions. Clinical presentation may vary and can include rapid onset of headache, seizures, confusion, blurred vision, and nausea. The exact pathophysiology is unclear; however, endothelial dysfunction is considered to be the underlying mechanism. Typical magnetic resonance imaging (MRI) appearance includes subcortical vasogenic oedema from leakage across blood brain barrier due to endothelial dysfunction or failure of cerebrovascular autoregulation due to severe arterial hypertension, usually seen in the parieto-occipital lobes.

Although the incidence of PRES is unknown, but it is reported to be seen in young to middle aged patients, especially in females. This is an unusual case of PRES due to severe pre-eclampsia, with clinical presentation of cortical blindness and hypertension, with absence of headache or seizures.

Case Presentation

A 30-year-old P0, Caucasian ethnicity, body mass index of 27.3, booked antenatally with a normal BP of 120/66 and no proteinuria. Her antenatal period was mostly uneventful. At 37 weeks 6 days, she was noted to have BP 128/82 with +++ proteinuria and worsening itching. Urine protein creatinine ratio (UPCR) was found to be elevated at 0.124 g/l. Urate was elevated at 426 mmol/l. Urine protein was significantly high at 1.24 g/mmol. She was otherwise asymptomatic with normal reflexes and no clonus. Hence, induction of labor was planned for her. During the induction process, the patient needed one dose of 200 mg labetalol for control of her raised BP. She progressed well in labor and delivered vaginally. She had remifentanil for analgesia. Platelets were noted slightly lower at 133 × 10^9/l during labor. Liver enzymes was normal at alanine transaminase (ALT)-12 units/l & aspartate transaminase (AST)- 22 units/l. Post-delivery her BP started rising (Table 1). Initially 160/106 to 170/113. Within 10 minutes of delivery, the patient reported inability to see her baby. She could appreciate between light and dark but

nothing else. At that point her BP was 189/143. On neurological examination, power and tone were normal with brisk knee reflexes bilaterally and two beats of clonus. Multidisciplinary team management including anesthetist, physician, ophthalmologist, and radiologist were involved. Pupils were reacting to light bilaterally. Fundus was grossly normal bilaterally. Aggressive management of severe pre-eclampsia was initiated with prophylactic magnesium sulphate (MgSO4) bolus and IV labetalol, as per protocol. An urgent computed tomography (CT) scan was requested and the working diagnosis at that time was retinal vessel spasm due to sudden rise in BP or a venous thrombosis. While she was being transferred for the CT scan, her vision started to return.

The differential diagnosis considered were cortical venous sinus thrombosis, transient ischemic attack, and cerebral vasospasm due to severe arterial hypertension. The CT scan showed patchy low attenuation in the occipital regions bilaterally and a small area in the left high parietal parafalcine region. No hemorrhage or any particular vascular distribution was noted. These appearances raised the possibility of PRES. An MRI scan was requested, and this demonstrated bilateral areas of white matter T2 hyperintensity in the occipital lobes. There were foci of diffusion restriction seen in the right occipital and bilateral parietal lobes and also in the head of the left caudate nucleus. The overall appearance was consistent with PRES with superimposed foci of acute ischemia.

Table 1. *Investigation results.*

INVESTIGATIONS	PRE-DELIVERY	IMMEDIATE POSTPARTUM	2 MONTHS POSTPARTUM	NORMAL RANGE
U&E				
Urea		4.0	3.2	5.8 2.5-7.8 (mmol/l)
Creatinine		69	62	61 50-100 (µmol/l)
Sodium		139	138	137 133-146 (mmol/l)
Potassium		3.9	3.9	4.9 3.5-5.3 (mmol/l)
Chloride		101	101	103 95-108 (mmol/l)
estimated Glomerular Filtration Rate (eGFR) result/1.73 m^2		>60	>60	>60 60-61 (ml/minute)
Full blood count				
RBC		4.05	5.22	3.88-4.99 (10^{12}/l)
Monocytes		0.9	0.5	0.2-0.9 (10^9/l)
Haemoglobin		104	142	122-165 (g/l)
White cell count		13.7	9.5	3.9-11.1 (10^9/l)
Platelets		151	291	150-400 (10^9/l)
Hematocrit		0.33	0.43	0.36-0.48 (%)
MCV		81.0	82	82-98 (fl)
MCH		25.7	27.2	27.3-32.6 (pg)
MCHC		318	330	316-349 (g/l)
Neutrophils		10.7	5.8	1.8-7.4 (10^9/l)
Lymphocytes		1.8	3	1.1-5 (10^9/l)
Eosinophils		0.2	0.2	0.1-0.7 (10^9/l)
Basophils		0	0	0-0.1 (10^9/l)
Liver function test				
Total bilirubin		3	5	3-21 (µmol/l)
ALP		142	129	30-130 (U/l)
AST		16	29	
ALT		14	49	5-55 (U/l)
Total protein		55	77	60-80 (g/l)
Albumin		28	51	35-50 (g/l)
Globulin		27	26	21-35 (g/l)
Urate	564	562		140-360 (umol/l)

INVESTIGATIONS	PRE-DELIVERY	IMMEDIATE POSTPARTUM	2 MONTHS POSTPARTUM	NORMAL RANGE
Urine				
Urine creatinine	10.04		4.81	
Urine Protein/creatinine ratio	0.124		0.010	0-0.014 (g/mmol)
Urine protein	1.24		0.05	0-0.5 (g/l)

MCV-Mean corpuscular volume; MCH-Mean corpuscular hemoglobin; MCHC-Mean corpuscular hemoglobin concentration; ALP-Alkaline phosphatase; AST-Aspartate transaminase; ALT-Alanine transaminase.

As soon as BP started returning to normal, the patient's vision improved within minutes. She continued to remain well and was discharged on eight postnatal day on antihypertensive labetalol 200 mg three times a day (TID) and nifedipine slow release (SR) 10 mg twice a day (BD). Bloods continued to be normal with platelets at 155 × 10^9/l, ALT-15 units/l and AST-32 units/l, just prior to discharge (Table 1).

During the follow-up by renal physicians, at 12 months postpartum, urine protein returned to normal at 0.07 g/mmol with UPCR 0.016 g/l. Further investigation including protein electrophoresis for paraproteins and immunoglobulins were reported normal. Antinuclear antibodies and double stranded DNA was normal, thus ruling out other renal causes of proteinuria.

Discussion

PRES is a rare condition which is characterized by a reversible clinico-radiological syndrome requiring aggressive management. It is more commonly seen in females in relation to preeclampsia or eclampsia with severe BP fluctuations. Fisher et al. [1] reported PRES associated with 62.5% of eclamptics and 10.6% of pre-eclamptics. Our case involved pregnancy with pre-eclampsia at delivery. Initial presentation was in the form of proteinuria which eventually led to fluctuation of BP during induction of labour. She presented only with transient cortical blindness without any other symptoms, which was very unusual for PRES.

Varied range of presentation in case reports is attributed to the region of brain affected. PRES is predominantly reported to involve parietal and occipital lobes. However, as per Bartynski et al. [2] involvement of the frontal lobe, temporal lobe, and cerebellar hemispheres is common in PRES, along with the occasional presence of lesions in the brain stem, basal ganglia, deep white matter, and splenium. Sanders et al. [3] have also mentioned possibility of involvement of basal ganglia and deep white matter.

PRES is manifested by neurologic symptoms: headache, nausea or vomiting, generalized seizures, visual disturbance, and altered sensorium. Recurrent seizures are common and visual disturbances are present ranging from hemianopsia and visual neglect to cortical blindness [4]. In the present case, absence of headache and seizures, with only visual disturbance was rather atypical.

Rare case of sensory and motor deficit and amnesia due to unequal posterior cerebral involvement has been documented [5]. Cases of postpartum reversible posterior leukoencephalopathy syndrome with involvement of anterior brain regions after eclampsia complicated by hemolysis, elevated liver enzymes, low platelet (HELLP) syndrome are very rare [6]. Visual disturbances are attributed to vasogenic oedema of the parietal lobe.

Rapid diagnosis and aggressive management are the key to prevent irreversible neurological sequelae and death [7,8]. Cozzolino et al. [9] reported that the goal of the therapy is to control elevated BP and to prevent seizures or promptly manage it [10].

Conclusion

PRES is rare entity in pregnancy with pre-eclampsia. It has varied presentation, ranging from headache, nausea, vomiting, seizures, visual disturbance, and altered sensorium in different combination. As in this case, it can present with a solitary symptom of cortical blindness. It requires early diagnosis and aggressive management with multidisciplinary team involvement for good outcome and ensuring patient safety.

What is new?

PRES is an unusual clinical entity seen associated with BP fluctuations presenting with acute neurological symptoms. It usually has varied presentation, ranging from headache, nausea, vomiting, seizures, visual disturbance, and altered sensorium either in different combination or with solitary symptom of cortical blindness, as in our case. It requires early diagnosis and aggressive management with multidisciplinary team involvement for good outcome and ensuring patient safety.

List of Abbreviations

ALT	Alanine transaminase
AST	Aspartate transaminase
BP	Blood pressure
CT	Computed tomography
MgSO4	Magnesium sulphate
MRI	Magnetic resonance imaging
PRES	Posterior reversible encephalopathy syndrome
UPCR	Urine protein creatinine ratio

Conflict of interests

The authors declare that there is no conflict of interest regarding the publication of this article.

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Consent for publication
A written informed consent to publish/present this case was obtained from the patient.

Ethical approval
Ethical approval is not required at our institution to publish an anonymous case report.

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Summary of the case

1	**Patient details**	Female, 77 years old
2	**Symptoms**	Syncope
3	**Final diagnosis**	Syncope secondary to AHCM
4	**Clinical procedures**	Echocardiogram
5	**Clinical specialty**	Cardiology
6	**Interesting features**	Lateral ST segment elevation on ECG secondary to AHCM