Posterior reversible encephalopathy syndrome-associated bilateral cortical blindness as presenting feature of severe pre-eclampsia

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DESCRIPTION

A 24-year-old primigravida (gravida 1, para 0), at 34 weeks of gestation, presented to the emergency department of our hospital with a sudden painless bilateral visual loss for 1 day associated with mild headache. She did not have any history of loss of consciousness, trauma, seizure, fever, hypertension, diabetes, thyroid disorder or any cardiovascular diseases. Her antenatal history was insignificant. Ocular examination revealed visual acuity of only light perception and intact pupillary light responses in both eyes. Fundus (figure 1A,B) was unremarkable bilaterally, except a single dot haemorrhage temporal to the fovea in the right eye. No disc oedema, retinal detachment or any retinal vascular abnormality was noted in either eye. Optical coherence tomography (figure 1C,D) showed bilateral normal foveal contour and retinal nerve fibre layer thickness. Her blood pressure was found to be 170/100 mm Hg, recorded multiple times. Obstetrical examinations including fetal assessment and other systemic evaluations were normal. Laboratory investigations including complete haemogram, liver function test, renal function test, serum electrolytes and coagulation profile were unremarkable, except hypocalcaemia (6.5 mg/dL) and raised serum alkaline phosphatase levels (454.8 U/L). Urinalysis revealed 2+ proteinuria and elevated levels of 24-hour urinary protein. Considering severe

Figure 2 Brain MRI showing hyperintensities on T2-weighted images and fluid-attenuated inversion recovery images in bilateral parietal (A and D), occipital (B and E) and cerebellar hemispheres (C and F).

pre-eclampsia, treatment with magnesium sulfate (according to Pritchard regimen) and antihypertensive medication (labetalol) was initiated. An urgent MRI of the brain with venography revealed areas of T2/fluid-attenuated inversion recovery hyperintensities (figure 2A–F) in bilateral parieto-occipital lobes and cerebellar hemispheres, suggesting posterior reversible encephalopathy syndrome (PRES). A diagnosis of bilateral cortical blindness due to severe pre-eclampsia-associated PRES (PE-PRES) was made, and an emergency caesarean section was performed. The perinatal period was uneventful with a healthy mother and child. One week post-operatively, her visual acuity gradually restored to 20/20 in both eyes.

Pre-eclampsia and eclampsia are among the most common conditions associated with PRES, others being blood pressure fluctuations, renal failure, autoimmune diseases, cytotoxic drugs and organ transplants. PRES is characterised by reversible vasogenic cerebral oedema, often presenting with seizures, encephalopathy, headache or visual disturbances.^{1 2} Various mechanisms, including failure of cerebral autoregulation, endothelial injury by sudden blood pressure changes and direct cytokine effects on endothelium, have been postulated. All of these factors can lead to blood–brain barrier breakdown and consequent brain oedema.¹

As parietal and occipital lobes are preferentially involved, visual symptoms are common in PRES. Lifson *et al* have reported visual complaints in 27% of patients diagnosed with PRES.³ However,

Figure 1 Fundus photograph of both eyes (A,B) showing normal fundus except tiny retinal haemorrhage temporal to the fovea in the right eye. Optical coherence tomography demonstrating normal foveal contour (C,D) with insets showing normal retinal nerve fibre layer thickness bilaterally.

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Images in...

clinical presentations in PE-PRES may vary from PRES due to other causes. PE-PRES was reported to have a higher prevalence of headache, less prevalence of altered mental status, equal prevalence of seizures and visual disturbances, less severe presentation and relatively better outcome than PRES due to other aetiologies. On neuroimaging also, patients with PE-PRES demonstrated less severe oedema, haemorrhage and contrast enhancement at presentation. More frequent resolution of oedema and less frequent residual structural lesions were noted on follow-up imaging in this category.⁴

Cortical blindness and serous retinal detachment are two important causes of reversible severe visual loss in pre-eclampsia.^{5–7} The visual loss in PRES is usually regained with

the treatment of the predisposing cause.^{3 5} This report depicts an uncommon PRES-associated severe visual loss as the sole initial presenting feature of severe pre-eclampsia. Immediate bedside fundus evaluation, urgent neuroimaging and standard management of pre-eclampsia led to complete visual recovery within a week.

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Patient's perspective

I was extremely scared when I lost vision in both eyes suddenly and I thought I would never regain vision. However, I did not have any major problem or illnesses during my pregnancy. On examination by a team of doctors, I came to know that my blood pressure is raised and was advised some medications with immediate caesarean section. They also assured me that my vision is likely to improve following childbirth. Fortunately, I regained my vision in about a week after surgery.

Learning points

- Posterior reversible encephalopathy syndrome-associated bilateral cortical blindness may be the initial presenting feature of pre-eclampsia.
- Simple bedside fundus evaluation and urgent brain MRI are the mainstay of diagnosis.
- Rapid and complete visual recovery may take place with definitive management of pre-eclampsia.

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