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Bilateral lateral geniculate body lesions causing reversible blindness in a patient with posterior reversible encephalopathy syndrome

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Abstract

Posterior reversible encephalopathy syndrome (PRES) is a completely reversible neuroradiological entity caused by accelerated hypertension, eclampsia, certain cytotoxic drugs and acute renal failure. PRES involves posterior circulation of the brain resulting in various manifestations, hence the name. Acute vision loss is one of the manifestations that occurs owing to the involvement of the visual pathway. However, loss of vision due to a lesion involving the lateral geniculate body alone is unusual. We report one such case of a young female who developed acute bilateral painless loss of vision without any other symptom during postpartum period. MRI brain showed features of PRES involving bilateral lateral geniculate body, hippocampus and brainstem. There was no involvement of retrogeniculate visual pathway, i.e. parieto-occipital cortex. The patient improved with optimal blood pressure control and was discharged after 5 days.

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Introduction

Posterior reversible encephalopathy syndrome (PRES) occurs owing to failed cerebral circulatory autoregulation and endothelial dysfunction. The primary involvement is of the areas of brain supplied by posterior circulation. The mechanism behind selective involvement of these areas is poorly understood. One explanation is that the intracerebral and pial vessels in posterior circulation are poorly supplied by sympathetic nerves when compared to the vessels in anterior cerebral circulation.[1] We came across only two reported cases of PRES involving the visual pathway at the lateral geniculate body (LGB) alone.[2,3] This paper, besides describing the case, reviews the literature and discusses the management of PRES.

Case presentation

A 28-year-old multigravida female underwent lower segment caesarean section (LSCS) for her previous two deliveries at full term. She had no history of pre-eclampsia or eclampsia. She presented at our casualty department 11 days prior to the expected date of delivery, with abdominal pain for 3 hours. On examination, she was conscious, blood pressure was 140/90 mmHg and pulse rate was 90 beats per minute, regular. Abdominal examination revealed a full-term gravid uterus with scar tenderness. Sonography showed hypoechoic uterine defect at the site of previous caesarean. In view of impending scar rupture, she was taken up for emergency LSCS. Intraoperative course was uneventful and she delivered a full-term male baby. On postpartum day 2, she developed acute-onset vision loss in both the eyes. On examination, she was conscious and oriented. Blood pressure was 140/100 mmHg, pulse was 88 beats per minute. Both the pupils were 3 mm in size and reactive for direct and consensual light reflexes. She could not perceive light with both eyes. Extraocular movements were normal. Fundus examination was normal. Visual field could not be assessed. Rest of the nervous system and other systems examinations were normal. With acute vision loss in postpartum period, the differential diagnoses we considered were PRES and cerebral venous thrombosis. Blood tests showed total count of 13,100 cells/mm^3 with 84% neutrophils. Liver and renal function tests, coagulation profile, erythrocyte sedimentation rate, electrolytes and blood sugar values were normal. MRI of brain showed diffusion-weighted imaging (DWI) fluid attenuation inversion recovery sequence of T2-weighted imaging (T2WI-FLAIR) hyperintensities in bilateral lateral geniculate bodies, thalamus, hippocampus, midbrain and pons (Figures 1–3) without cortical and subcortical involvement.

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Figure 1 Axial DWI sequence of MRI brain showing diffusion restriction at bilateral lateral geniculate bodies. Cerebral cortex is spared

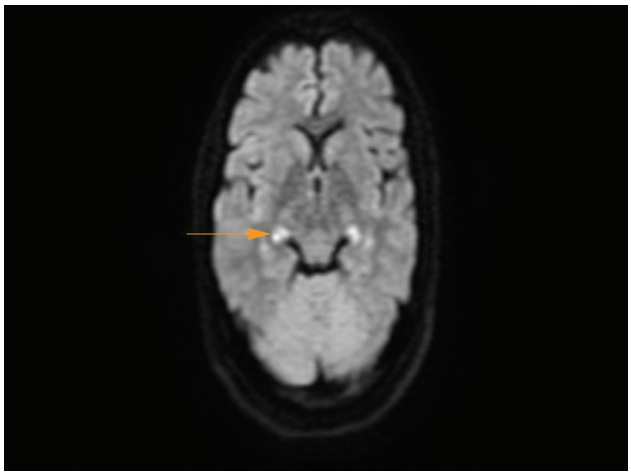
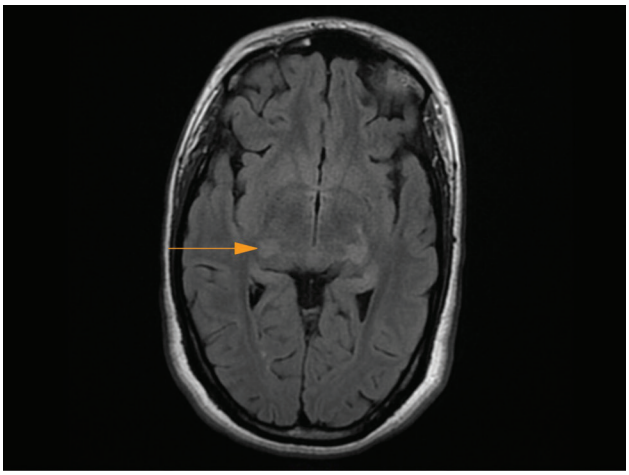


Figure 3 Axial T2WI-FLAIR sequence of MRI brain showing hyperintensities in the bilateral thalamus (lateral geniculate body) and hippocampus



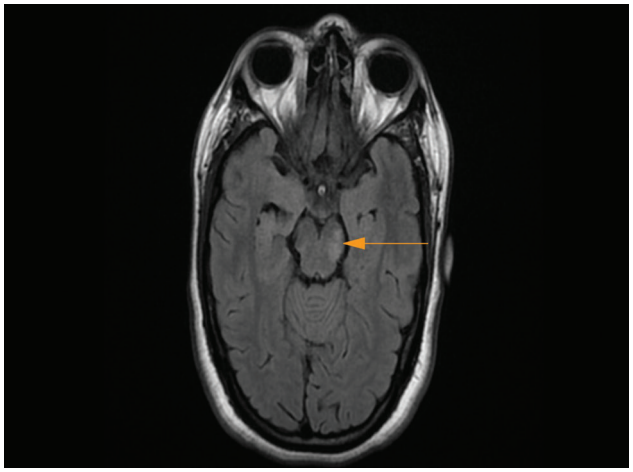
Time of flight sequence of MR venogram was normal. MRI findings were consistent with atypical PRES. She was managed with antihypertensives and intensive care unit care. Her vision improved to finger counting in both the eyes by the twelfth day. She was discharged on day 14, and on review after 1 month, her vision was normal in both the eyes.

Discussion

LGB is a relay centre in the thalamus that receives sensory input from the retina. LGB connects the optic nerve with the primary visual cortex in the occipital lobe. Malignant hypertension, acute on chronic kidney disease with accelerated hypertension, eclampsia, autoimmune vasculitides (systemic lupus erythematosus, Wegener's granulomatosis, systemic sclerosis) and immunosuppressive drugs are the risk factors for PRES. There are various theories explaining the pathophysiology of PRES:[4]

1. Uncontrolled hypertension causes failed autoregulation of cerebral blood flow resulting in hyperperfusion. This causes damage to the vessel wall leading to leakage of protein and fluid, implicating vasogenic oedema.
2. Vasoconstriction leads to hypoperfusion, cerebral ischaemia and infarction.
3. Systemic inflammatory states, such as sepsis and eclampsia, cause T-cell and cytokine-mediated endothelial damage followed by systemic vasoconstriction and hypoperfusion.

Figure 2 Axial T2WI-FLAIR sequence of imaging of MRI brain showing hyperintensities in the pons. There is no involvement of the cortex



PRES is characterised by acute-onset headache, visual disturbance, seizures and delirium. Between 20 and 30% of PRES cases have normal or mildly elevated blood pressure, as our patient had.[5] Clinical diagnostic criteria for PRES are still lacking, and in most cases the imaging is performed out of suspicion. Therefore, instant brain imaging is recommended in an appropriate clinical scenario. PRES typically involves the parieto-occipital lobes or 'posterior' cerebral cortex, hence the name. McKinney et al. studied 76 cases of PRES and described the patterns of involvement on MRI. The results showed involvement of the parieto-occipital lobes in 98.7%, posterior frontal lobe in 78.9%, temporal lobe in 68.4%, thalamus in 30.3%, cerebellum in 34.2%, brainstem in 18.4% and basal ganglia in 11.8% of cases.[6] Bartynski et al. described imaging patterns of PRES in 136 patients and the results showed lesions in parieto-occipital lobes in 98%, frontal lobes in 68%, inferotemporal lobes in 40% and cerebellum in 30% of patients. Involvement of other areas such as basal ganglia were noted in 14%, brainstem in 13% and deep white matter in 18% of the cases.[7] Loss of vision in PRES is commonly due to involvement of the visual pathway at the occipital lobe. Our patient had lesions involving LGB but sparing the retrogeniculate visual pathway, which is rare in PRES. Two similar cases have been previously reported. Stem et al. described a case of a young female with pre-eclampsia who developed PRES affecting bilateral LGB, but sparing the cerebral cortex.[2] A case of acute pancreatitis complicated by PRES with isolated LGB involvement was also reported.[3] Lesions involving basal ganglia, brainstem and deep white matter without affecting the cortical and subcortical grey matter are seen in 'atypical PRES'. A total of 12.5% patients had atypical PRES in the study by Bartynski et al.[7] Our case

had a similar pattern of involvement, thus fitting the picture of atypical PRES. The course of PRES is generally benign with good outcome in most cases. However, the outcome depends largely on the factor triggering PRES and timely management. Delay in management can lead to permanent neurological deficits.

Informed consent

Written informed consent for the paper to be published (including images, case history and data) was obtained from the patient/guardian for publication of this paper, including accompanying images.

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