Sudden Bilateral Choroidal Detachment in a Patient with Posterior Reversible Encephalopathy Syndrome

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Abstract

BACKGROUND: Pre-eclampsia is one of the most important causes of a rare cliniconeuroradiological entity called posterior reversible encephalopathy syndrome. The syndrome is characterized by headache, visual disturbances, seizures, altered mental status and radiological findings of edema in the white matter of the brain areas perfused by the posterior brain circulation.

CASE REPORT: Here we present a patient with sudden bilateral visual loss and deafness in early postnatal period without any other neurological deficits, but with affiliated ophthalmological pathology. The correct diagnosis was made 3 days after the Cesarean section. The patient was diagnosed with posterior reversible encephalopathy syndrome and bilateral choroidal detachment.

CONCLUSION: Vascular changes in posterior reversible encephalopathy syndrome can cause visual disturbances not only by brain edema, but combined brain and ocular pathology.

Introduction

Posterior reversible encephalopathy syndrome (PRES) was first described in 1996 by Hinchey [1] as reversible posterior leukoencephalopathy syndrome. Except pre-eclampsia and eclampsia [2], most common causes of PRES are acute hypertensive encephalopathy[3], hemolysis [4], HELLP syndrome, immunosuppressive medication, infection or autoimmune diseases, cancer, toxic and metabolic disorders. Visual disturbances range from blurred vision to cortical blindness and even permanent visual field defects. The syndrome is also characterized by headache, visual disturbances, seizures, altered mental status and radiological findings of edema in the white matter of the brain. The precise pathogenic mechanism leading to the development of PRES has not yet been identified. There are two hypothesis for the causes of PRES [5, 6]. The first is that a rapid rise in blood pressure leads to altered autoregulation of cerebral blood flow producing dilatation of cerebral arterioles with opening up of endothelial tight junctions and leakage of plasma and red cells into the extracellular space, producing cerebral edema. The second hypothesis is that patients with PRES develop vasospasm secondary to sudden and severe rises in blood pressure and ischemia of brain tissue. Ischemic damage to brain tissue first produces cytotoxic edema and then extracellular edema. The pathophysiology of
PRES implicates endothelial dysfunction, especially in cases without severe hypertension, such as pre-eclampsia, for both the pregnancy and the postpartum period predispose the cerebral circulation to forced dilatation at lower pressures. Such response may lower cerebrovascular resistance and promote hyperperfusion when blood pressure is elevated, as occurs during eclampsia. The right diagnosis is typically made on magnetic resonance imaging of the brain and cerebral angiography. The treatment of PRES depends on the underlying cause.

**Case Report**

A 22 year-old pregnant woman was examined by a gynecologist and diagnosed with pre-eclampsia at the gestation age of 37 weeks + 4 days. She developed high blood pressure and generalized edema. Laboratory tests showed slight elevation of liver enzymes and mild proteinuria. She was admitted to the Gynaecology Department and treated with magnesium sulfate and parenteral antihypertensive therapy (labetalol), without effective response to treatment as her blood pressure remained elevated at 190/120 mmHg. Considering her gestation age, obstetric finding and hypertension unresponsive to treatment, Cesarean section was performed without any complications. After the procedure, the patient’s general condition improved, with lowering of the blood pressure. Ten hours after the Cesarean section, the patient started to complain of bilateral blurred vision, which rapidly deteriorated to light perception in both eyes. Another ten hours later, the patient developed bilateral deafness. However, her general condition and her postoperative course remained stable. The etiology of sudden bilateral visual loss and deafness was unclear. She was referred to the University department of Ophthalmology and diagnosed with massive bilateral choroidal detachment and bilateral macular edema. Clinical findings were confirmed with ultrasound imaging. Otorhinolaryngology examination did not reveal any abnormalities. Lab tests showed only mild elevation of liver enzymes and CRP (30.6 mg/L). Physical examination was also normal, including a full neurological status.

Magnetic resonance imaging (MRI) of the brain revealed bilateral symmetric vasogenic edema mainly involving cortical and subcortical white matter in the occipital lobes, but also periventricular white matter, basal ganglia and part of parieto-occipital sulcus on the right side. The finding was recognized as posterior reversible encephalopathy syndrome (PRES). Magnetic resonance angiography (MRA) showed spasm of intracranial blood vessels without any vascular abnormality. Her blood pressure was within normal limits under parenteral antihypertensive therapy. Her treatment included magnesium sulfate, infusion, sodium nitroprusside, diuretics (furosemide) and antiedema osmotherapy (Mannitol). She was under continuous monitoring by ophthalmologists and was treated with topical corticosteroid and cycloplegic drops. Six days after the development of visual disturbances there was a significant improvement of vision. Intraocular pressure was within normal limits (14/16 mmHg). Ophthalmoscopic exam showed a regression of choroidal detachment, with a shallow residual detachment in the peripheral lower parts and minor macular edema with folds of internal limiting membrane. Optical coherence tomography images of the macula confirmed intraretinal edema in both eyes. Her visual disturbances resolved after one month, and macular edema regressed as well. However, her best corrected visual acuity remained slightly diminished at 0.7 on both eyes. Patient’s hearing recovered completely and the blood pressure was within normal limits. Follow-up MRI demonstrated complete resolution of the edema six weeks after the delivery.

**Discussion**

PRES is a clinicoradiological syndrome precipitated by various clinical settings [7-13]. The exact pathogenesis of this neurotoxic state is not known, but is thought to be related to altered integrity of the blood brain barrier. Treatment of severe hypertension in pregnancy is important for the hypertension remains a major cause of maternal and fetal morbidity and mortality. It is recommended that blood pressures greater than 170/110 should be treated with urgency [14]. Our patient was treated with parenteral labetalol which is safe and effective initial treatment of severe hypertension in pregnancy [15, 16], and with anticonvulsant therapy (magnesium sulfate) prophylactically [17]. Magnesium therapy should be initiated as soon as eclampsia or PRES in pregnancy is suspected, as it treats both seizures and hypertension [18, 19]. Since the delivery presents an ultimate treatment for the pre-eclampsia, the Cesarean section in our patient was performed, considering patient’s gestation age and obstetric finding. Accelerated hypertension unresponsive to treatment in our patient may be related to altered integrity of the blood brain barrier and PRES. The treatment of PRES is based in the management of the triggering factor, and the reversibility of the neurological changes mainly depends on the rapid treatment of brain edema. Accelerated hypertension in pre-eclampsia has been commonly associated with retinopathy, optic neuropathy and choroidopathy [20, 21]. Our patient developed bilateral choroidal detachment and macular edema, the finding which has not yet been reported in association with PRES or accelerated hypertension. The exact pathogenesis of bilateral choroidal detachment within PRES requires additional research. We consider ocular changes to be partially responsible for incomplete visual recovery in our patient, since her follow-up brain MRI demonstrated no abnormalities.

In conclusion we suggest the importance of ophthalmologic examination and monitoring of...
patients with ocular symptoms connected with PRES since visual disturbances may not only be induced by brain edema but also by associated local ocular changes, as serous retinal detachment described previously in the literature [22-24], and bilateral choroidal detachment described in this case report.

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