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CASE REPORT

Posterior Reversible Leukoencephalopathy Syndrome: A Rare Complication with Gestational Hypertension and Pre Eclampsia

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ABSTRACT

Incidence of pre eclampsia is 6-8% of all pregnancies worldwide. Two major complications of pre eclampsia include eclampsia and HELLP syndrome. In United kingdom Eclampsia occurs in 1 in 2000 deliveries and has a mortality of 1.8%. HELLP syndrome occurs in 1 in 500 deliveries and can be as dangerous as eclampsia. In India the incidence of eclampsia is 3-4% with maternal mortality ratio of 10.44% and incidence of HELLP syndrome is 10-12% with a maternal mortality of 18%. In very rare cases, temporary vision loss can occur due to partial or complete cortical blindness due to petechial hemorrhages and focal vasogenic edema in the occipital cortex. The reported incidence of posterior reversible encephalopathy syndrome (PRES) is 0.01%. We are reporting a case of PRES that was a cause of total cortical blindness in a patient with pre eclampsia which was completely reversed with timely intervention and supportive treatment.

INTRODUCTION

First reported case of reversible cortical blindness was in 1996 by Hinchey et al.[1]In 2000 this condition was called posterior reversible encephalopathy syndrome (PRES) It is also called reversible posterior leukoencephalopathy syndrome (RPLS). Before 1996 there were several cases reports describing the CT and MRI findings similar to that of PRES as an interesting finding of eclampsia.[2] This is a very rare complication of pre eclampsia with an incidence of 0.01%.[3] In all these cases the PRES was preceded by severe hypertension, headache (thunderclap) and convulsions. In our case only THUNDER CLAP head ache was present followed by complete loss of vision which is an alarming complication in the post natal period. It is a self limiting condition when blood pressure is controlled. Prompt control of blood pressure is warranted to prevent further cortical damage and neurological deficit.[4,5]

CASE HISTORY

A 32 year old gravida 3 with 1 live child and 1 abortion was detected to have gestational hypertension at 35 weeks of gestation with a blood pressure (BP) of 140/100. She was a booked patient with regular antenatal checkups. Physical examination was unremarkable and her routine investigations, renal and liver function test were within normal limits. She was started on antihypertensive α methyl dopa at 250 mg thrice daily and her blood pressure was maintained at 130/90 mm of Hg.

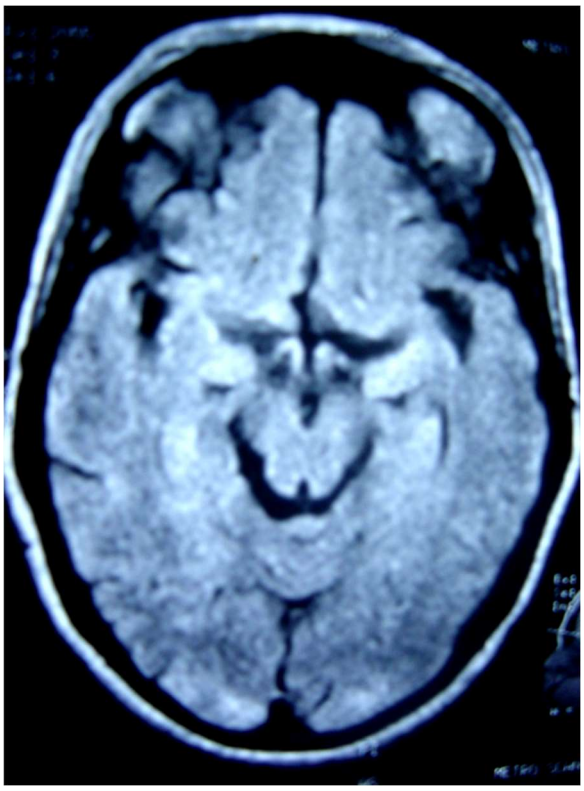

Figure 1: MRI: flair axial section: hyper intense lesion in both right and left occipital lobe

Elective LSCS was done at 37 completed weeks of pregnancy as she had a previous LSCS due to contracted pelvis. Preoperative and intra operative blood pressures were well controlled. Immediate post operative period was uneventful. As her blood pressure was normal postoperatively anti hypertensive were with held. On post-op day 4, BP was found to be 170/120 mm of Hg. Antihypertensive atenolol 25 mg daily was started. On post operative day 6, she developed sleeplessness; headache and sudden loss of vision both eyes with a BP of 180/120 and no Perception of light both eyes. The anterior segment was within normal limits. Pupil both direct and consensual reaction was normal in both eyes. Dilated fundus examination was within normal limits in both eyes. Neurological examination revealed absence of long tract signs, absence of neck stiffness, normal pupillary reflex and preservation of higher mental functions with signs of cortical blindness. Emergency MRI revealed high intensity signals involving sub cortical white matter in both occipital lobes, diagnostic of PRES.

Blood pressure was controlled with atenolol and amlodipine 5 mg daily. She was started on IV mannitol and other symptomatic measures Clinical improvement was noticed within 48 hours and her vision improved to 6/6 by the 4th day of diagnosis. On follow up after six weeks, she has no neurological sequelae and has normal fields of vision.

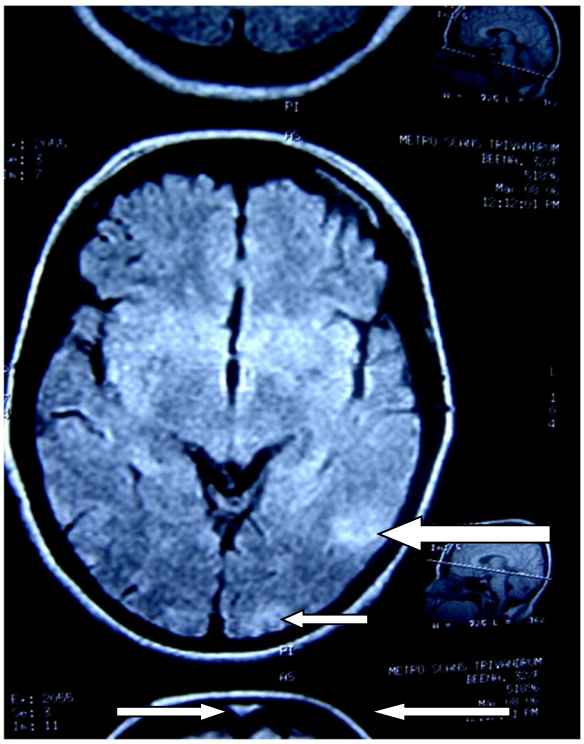

Figure 2: MRI: flair axial section: Two hyper intense lesion in the left occipital lobe

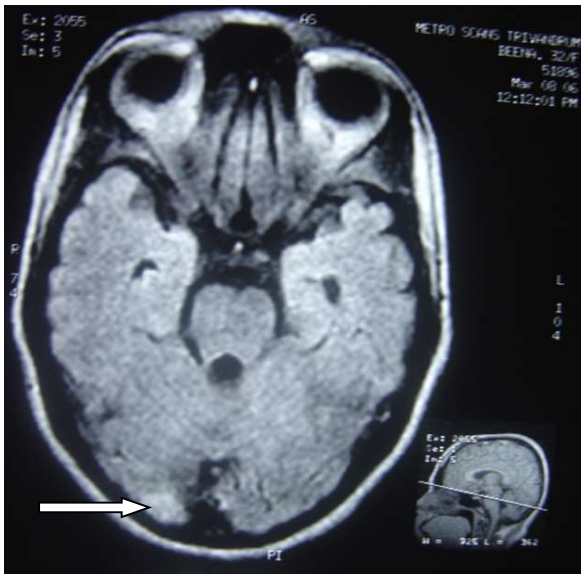

Figure 3: MRI Flair axial section - Hyper intense lesion in the right occipital lobe

DISCUSSION

Posterior reversible encephalopathy syndrome (PRES) is a recently proposed clinic neuro radiologic syndrome.[6, 7] It is characterized by seizures, disorders of consciousness, visual abnormalities and headaches associated with predominantly posterior white matter abnormalities on CT and MRI examinations. Common inciting factor of PRES are acute elevations

of blood pressure, renal decompensation, fluid retention, and treatment with immunosuppressive drugs.[1]

Table 1: Inciting factors of PRES

Acute elevation of Blood Pressure
Renal decompensation
Fluid Retention
Treatment with immunosuppressive drugs

The most common clinical symptoms and signs are headache, altered alertness and behavior ranging from drowsiness to stupor, seizures, vomiting, mental abnormalities including confusion and diminished spontaneity of speech, and abnormalities of visual perception. The onset of symptoms may be sub acute but it may be heralded by occurrence of a sizure. Seizures can occur at the onset or at a later stage. Usually signs with which the patient presents with are lethargy and somnolesence but stupor and frank coma may also be the presenting signs some patients have in coordination and weakness of limbs with brisk tendon reflexes. Signs of visual abnormalities may include visual neglect Hemianopia and frank cortical blindness.[1]

Table 2: Symptoms and signs in PRES

Symptoms	Signs
Thunder Clap Headache	Altered alertness
Vomiting	Drowsiness
Mental abnormalities	Stupor
Decreased spontaneity of speech	Seizures
	Visual abnormalities

Neuro imaging is indicated in patients presenting with altered consciousness in pregnancy and if there are atypical features or visual disturbance in patients with eclampsia in both antenatal and postnatal period[8] MRI is the imaging modality of choice.[9]

It is important to distinguish between PRES from acute ischemic stroke. Hypertension in ischemic stroke should not be managed aggressively but in PRES active management and control of hypertension is the treatment of choice.[5]

During the acute phase neuro imaging would reveal edema involving the white matter in the posterior portion of the cerebral hemisphere, especially bilaterally in the parieto-occipatal regions. The calacarine and paramedian occipital lobe structures are usually spared, a fact that distinguishes reversible posterior leukoencephalopathy from bilateral infarction of the posterior – cerebral artery territory[10, 11]. Hypertensive encephalopathy is the cause of this condition.

Hypertensive encephalopathy and preeclampsia-eclampsia share similar pathophysiological mechanism.[12] Sudden elevations in the systemic blood pressure exceed the auto regulatory capability of the brain vasculature and there is break down of the blood brain barrier with focal transduction of fluid and petechial hemorrhages. Disappearance of the clinical signs and imaging abnormalities after control of hypertension suggests towards edema as the cause.[13] Microscopically, these petechiae are ring hemorrhages around capillaries and precapillaries, which are occluded by fibrinoid material. The susceptibility of the posterior portion of the brain to the lesions seen in hypertensive encephalopathy and eclampsia is recognized. This is probably because the vertebrobasilar vessels are relatively devoid of sympathetic innervations and these results in the loss of auto regulation and forced arteriolar dilatation predominantly in watershed occipital lobe.

The differential diagnosis considered:[14]

Infarcts including "top of basilar syndrome"
Venous thrombosis
Infections-meningitis encephalitis
Post infectious encephalomyelitis
Vasculitis
Epinephrine induced [2015]

PRES is a devastating uncommon condition which has excellent prognosis if treated actively. In patients with Eclampsia and severe pre eclampsia magnesium sulphate treatment is started either at onset of seizure or prophylactically. As pre eclampsia is a progressive condition, the prognosis will be poor if pregnancy continues. Hence delivery of the fetus is the definitive management along with control of blood pressure and control of seizures. If the patient presents before 34 weeks of pregnancy the delivery may be delayed for 24-48 hrs for the action of corticosteroids to set in[1716]With these management the morbidity of PRES can be brought down.

Aggressive anti hypertensive treatment is started in all patients with systolic blood pressure of 170 mm of Hg or a diastolic blood pressure of over 110 mm of Hg[16] Drugs for treatment of hypertension in pregnancy has been subjected to Cochrane review. This shows that there is no advantage of one drug over the other[17]. The aim of pharmacological treatment is to maintain the blood pressure systolic between 140 and 160mm of Hg and diastolic between 90 -105 mm of Hg

Three cases where reported by magi et al in 2013[3] The first case of 29 years had seizures at 35 weeks of gestation and pregnancy was terminated by caesarean section due to eclampsia and she developed PRES after that. The second patient was 45 year old patient at 39 weeks of gestation who had severe oedema antenataly with no prior blood pressure elevation She

developed seizures and her pregnancy was terminated by caesarean section. 6 hours after Caesarean section she had Focal changes in the MRI diagnostic of PRES-. The third patient was a 35 year old second gravida with one live child who had no prior elevated blood pressure. She developed elevated blood pressure during labor and she had seizures 2 hour post delivery. She had focal neurological deficit and mild limb edema.[3] In all these cases the presenting feature was seizure

Our patient developed PRES in the puerperium, rather than during pregnancy. There is massive fluid accumulation in extracellular spaces and hemoconcentration in pregnancy complicated by preeclampsia. During puerperium, there is fluid shift back to intravascular space; this may have accentuated the tendency for brain edema to develop.

As our patient presented post natally she was treated with amlodipine and atenolol for control of blood pressure. Prompt control of blood pressure is essential for the treatment of this condition.

CONCLUSION

RPLS or PRES though reversible, if left untreated the arterial hypertension can lead to progressive neurological deterioration with infarction, hemorrhage and possible irreversible neurologic deficit.[4] The exact time interval between the diagnosis, control of blood pressure and the permanent damage has not been studied. Therefore, PRES is added to the list of indication for termination of pregnancy in patients with preeclampsia.

Prompt diagnosis and strict control of blood pressure is paramount importance in this condition. Early recognition and treatment can save patient's vision and avoid morbidity due to neurological deficits

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