POSTPARTUM POSTERIOR REVERSIBLE ENCEPHALOPATHY SYNDROME (PRES)

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Abstract: *Posterior reversible leukoencephalopathy syndrome (PRES) is a rare neurological disease characterized by elevated blood pressure, headaches, consciousness impairment, seizures, focal neurological signs and visual abnormalities. The risk factors are: malignant hypertension, eclampsia, chemotherapy agents, chronic renal failure, bone marrow and solid organ transplantations. The vasogenic oedema in PRES usually resolves completely within a week. If PRES occurs during the pregnancy, delivery of the baby is almost always curative. If it occurs after the birth of the child, retained placental products are often found and removal seems to be curative. We present the case of a 19-year-old woman, primi-parous, who developed PRES in the post- partum period, without evidence of preeclampsia-eclampsia or hypertension. Two days after spontaneous vaginal delivery, the patient presented tonic-clonic seizures, hypertension and leg edema. Cerebral computed tomography (CT) scan revealed temporal and capsular bilateral hypodensities areas. Since depletive and anti-hypertensive treatment has been introduced, the patient has not had any seizure and the blood pressure has reached a normal level. Also, the leg edema has remitted. In conclusion, our experience with this case has shown the importance of an early imaging monitoring of patients who develop post-partum seizures so that they can be diagnosed with PRES. Early recognition of the signs and symptoms of PRES, particularly identifying and treating high blood pressure, can prevent permanent neurologic disability.*

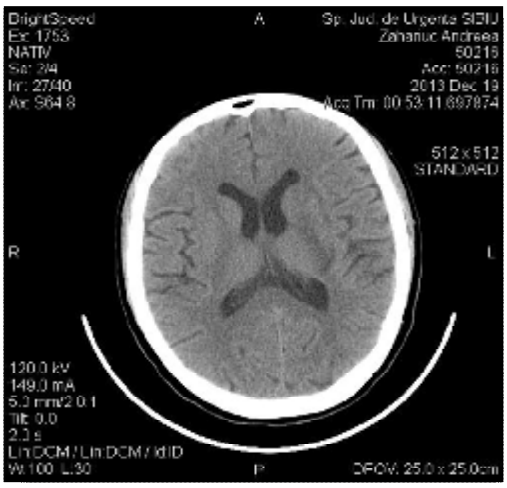
INTRODUCTION

Posterior Reversible Encephalopathy Syndrome (PRES) is a clinico-radiological syndrome, with diverse clinical presentation and characteristic Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) features. It describes a usually reversible non-specific neurologic symptoms include headache, confusion, visual disturbances, seizures, altered mental status, paresis, generalized seizures and coma.(1) The lesions in PRES are thought to be due to vasogenic edema, predominantly in the posterior cerebral hemispheres and are reversible with appropriate management. It may accrue in diverse situations, including hypertension, eclampsia, pre-eclampsia, allogenic bone marrow transplantation, in patients taking cyclosporine (and occasionally tacrolimus), in patients with autoimmune disease, renal failure, infection, sepsis (gram positive organisms predominate) and shock.(2,3) The imaging of PRES, CT or MRI shows occipital and parietal bilateral edema affecting both the cortex and subcortical white matter of the brain. Other common sites for PRES include the frontal lobes, the inferior temporal-occipital junction, and the cerebellum.(4) The treatment of PRES depends on the underlying cause. For instance, if the main problem is high blood pressure, blood pressure control will accelerate the resolution of the abnormalities. If the likely cause is medication, the withdrawal of the drug in question is needed.(5)

CASE REPORT

A 19-year-old female, primaparous, with negative past medical history, admitted in intensive care department for two episodes of generalized tonic-clonic seizures, accompanied by sphincter relaxation, vomiting, tongue bitten, hypertension and leg edema. She presented these symptoms after spontaneous vaginal delivery, two days before. Clinical examination: paleness of skin, afebrile, legs edema; blood pressure was– 168/110 mmHg. Neurologic examination – revealed absence of neck stiffness, absence motor deficits, normal pupillary reflex, deep tendons reflexes brisk, bilateral Babinsky sign, she was drowsy and confused. Emergency cerebral CT scan– revealed temporal and capsular bilateral hypo densities areas (figure no.1). All these changes are suggestive of posterior reversible encephalopathy syndrome (PRES).

Figure no. 1. Native cerebral CT scan, revealed bilateral hypodense lesions in temporal and capsular areas, suggestive of oedema



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The patient had no known history of migraine, or seizures.

At the admission in emergency unit, the laboratory examination (complete blood picture, liver enzymes, kidney function tests, clotting parameters, serum electrolytes-sodium, potassium) were in normal range. There was no evidence of urinary proteinuria.

The chest X-ray examination was normal. Abdominal examination was normal.

A lumbar puncture for CSF analysis to rule out meningoencephalitis was not performed as the patient was afebrile with no signs of meningeal irritation.

Electrocardiographic (ECG) study was unremarkable.

Although cerebral CT scan may show abnormalities in 50% of patients, MRI is the imaging modalities of choice fore the diagnosis of PRES.(7) Non-enhanced head CT (NECT) can detect PRES when subcortical areas of low density are visible, however areas of limited can be missed.(7)

In our case report, images presented at non-enhanced head CT, (figure no 1.) was suggestive for PRESS in clinic context.

In the pregnancy period, there was a fluid accumulation in extracellular spaces and this generated hemoconcentation condition. During puerperium, there was fluid shift back to intravascular space; this has accentuated the tendency to develop brain oedema.

Our patient was treated in intensive care unit, with magnesium sulphate, intravenous infusion, 2 g/hour (3 days), calcium channel blockers (amlodipine 5 mg daily, 5 days), benzodiazepines (midazolam, 2.5 mg-intravenous /one dose, at admission) and cerebral depletives IV mannitol 20%, (100 ml intravenous infusion every 6 hours, 5 days).

In intensive care unit, the patient had blood pressure controlled permanently.

Since depletive and anti-hypertensive treatment has been introduced, the patient has not had any seizure and the blood pressure has reached a normal level. Also, the leg edema has remitted.

10^{th} days after admission, the patient was discharged with a blood pressure of 110/70 mmHg and seizure free, in stable condition, with normal general and neurologic examination. Follow-up, non-enhanced head CT imaging also supported the resolution of abnormal signals.

Clinical signs, radiologic imaging and evolution sustained diagnostic of PRES in our case.

DISCUSSIONS

Rapid diagnosis of PRES is essential for initiating treatment, to prevent complications such as infarction and haemorragic stroke.(8) PRES was originally thought to only present in the occipital lobes. Although a significant number of patients have some occipital lobe involvement, lesions can appear anywhere in the white matter with edema extending to the cortex. The pathogenesis of PRES is not completely understood. Vasogenic theory i.e. hypertension with loss of autoregulation remains a widely accepted theory for the development of brain oedema.(6)

Endothelial dysfunction/injury, hypoperfusion, and vasoconstriction may lead to altered integrity of the blood-brain barrier (BBB). Although water has previously been thought to transit across BBB easily, this is now known to be mediated by specific cannel, aquaporin-4. Work in animals suggests that water transport across the BBB is regulated in large part by aquaporin-4 and recent work has identified aquaporin-4 as a possible agent in development of PRES.(4) In most cases, vasogenic oedema in PRES usually resolves completely within a week.

CSF usually normal or may be elevated protein. The differential diagnosis of PRES includes, infarcts including “top of basilar syndrome”, venous thrombosis, infections-meningitis, encephalitis, post infectious encephalomyelitis and vasculitis. Management is supportive and includes control of blood pressure, seizure and to reduce brain oedema. The aim of pharmacological treatment is to maintain the blood pressure systolic between 140 and 160 mmHg and the diastolic one between 90-105 mmHg.

CONCLUSIONS

We have presented the case of a young primi-parous female, with negative medical history, who developed early post-partum, recurrent seizures and hypertension. Prompt recognition of the signs and symptoms of PRES, particularly identifying and treating high blood pressure, can prevent permanent neurologic disability.

PRES is a reversible condition, if the arterial hypertension is promptly treated. The exact time interval between the diagnosis, control blood pressure and permanent damage has not been studied.

Imagistic particularity in this case refer to the damage of the subcortical brain regions (temporal and capsular area), presented at non-enhanced head CT, which was complete reversible after treatment.

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