Posterior Reversible Encephalopathy Syndrome: A Case Report

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Abstract :

Posterior reversible encephalopathy syndrome (PRES) is characterized by seizure activity, impaired consciousness, headaches, visual symptoms, nausea/vomiting and focal neurological signs. It presents with rapid onset of symptoms including headache, seizures, vomiting, altered consciousness, and visual disturbance. It is associated with hypertension. As the name suggests, it is typically reversible once the underlying cause is removed. PRES is also known as acute hypertensive encephalopathy or reversible posterior leukoencephalopathy. The diagnosis is typically made clinically with magnetic resonance imaging of the brain often revealing hyperintensities on T2-weighed imaging. The treatment of Posterior reversible encephalopathy syndrome dependent on its cause. Anti-epileptic medication may also be appropriate. We report a case of 15-year male child presented with headache, vomiting and one episode of generalized tonic–clonic seizure approximately 30 minutes prior to presentation to the emergency center. Physical examination revealed that he was fully alert and oriented. We made a diagnosis of Posterior reversible encephalopathy syndrome on further evaluation of history, laboratory findings and radiological investigations. The patient was managed on anti-hypertensive, anti-convulsant and supportive treatment.

Keywords: Posterior reversible encephalopathy syndrome, PRES, acute hypertensive encephalopathy.

Introduction:

Posterior reversible encephalopathy syndrome (PRES) was first described by Hinchey in 1996.

It is characterized by seizure activity, impaired consciousness, headaches, visual symptoms, nausea/vomiting and focal neurological signs.[(1)] PRES can be associated with a number of conditions, all of which result in cerebral vasogenic oedema which seems to be the crucial pathogenic mechanism. As the name suggests, it is typically reversible once the underlying cause is removed.[(2)] PRES is also known as acute hypertensive encephalopathy or reversible posterior leukoencephalopathy. It should not be confused with chronic hypertensive encephalopathy, also known as hypertensive microangiopathy, which results in microhemorrhages in the basal ganglia, pons, and cerebellum.[(3)]

Case report:

An 15-year male child presented with headache, vomiting and one episode of generalized tonic–clonic seizure approximately 30 minutes prior to presentation to the emergency.

Physical examination revealed that he was fully alert and oriented. He had a temperature of 99°F, blood pressure 186/100 mm Hg, pulse rate 94/min and respiratory rate 20/min. Renal angles were non-tender.

An ocular examination revealed a diminution of vision of bilateral eyes. Pupils were normally reactive with normal fundus. Power was 5/5 across all major joints and sensory function was intact all over the body. No any positive cerebellar or signs of meningeal involvement. Plantar were down going bilaterally.

Laboratory findings were significant for an elevated white cell count. Urinalysis was remarkable for +protein and RBC were present. T2-weighted images of brain MRI showed bilateral posterior parietooccipital hyper densities in the cortex and subcortical white matter consistent with posterior reversible leukoencephalopathy syndrome.**(Fig 1 & 2)**

We made a diagnosis of Posterior reversible encephalopathy syndrome on further evaluation of history, laboratory findings and radiological investigations. The patient was managed on antihypertensive, anti-convulsant and supportive treatment.

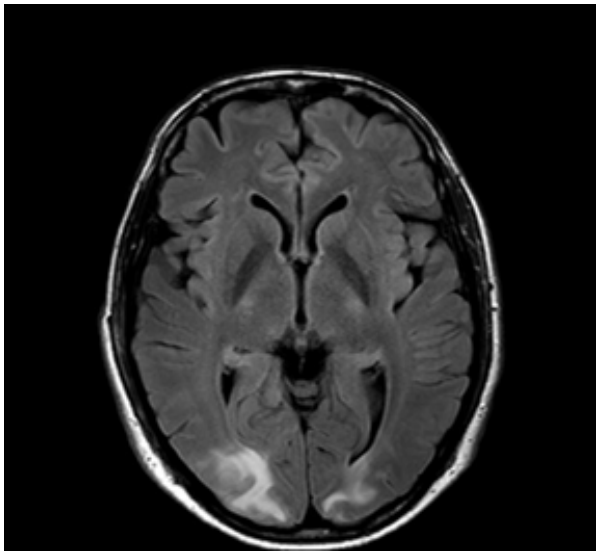

Fig.1: Bilateral posterior parietooccipital hyper densities in the cortex & subcortical white matter

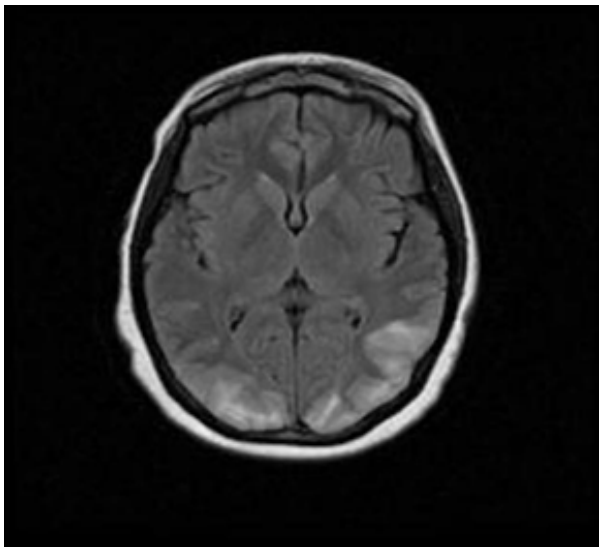

Fig.2 : Hyperintense lesion on T2 weighted MRI over parietal and occipital area

Discussion:

Although significant elevation of the blood pressure may not always be demonstrated, Posterior reversible encephalopathy syndrome is considered to be a variant of hypertensive encephalopathy.[3]

Two theories are considered in the pathophysiology of PRES, the first being sudden increase in blood pressure causing vasospasm and the other being failure of autoregulatory mechanism. With sudden elevation in systolic blood pressure, the autoregulatory capacity of brain vasculature is exceeded which results in a region of vasodilatation and vasoconstriction, especially in the arterial boundary zone. This causes breakdown of the blood–brain barrier with subsequent transudation of fluid along with hemorrhage.

Posterior reversible encephalopathy syndrome (PRES) is a rare disorder associated with acute hypertension; its exact incidence remains unknown. The pathogenesis of PRES is not clear; it seems to be associated with a rapid development of hypertension that leads to a malfunction of cerebral autoregulation; in particular in occipital lobe where the sympathetic innervation is less widespread, resulting in focal vasogenic edema.[4] Other conditions related to PRES are also chemotherapy, infection, sepsis, autoimmune diseases, and hypercalcemia (cytotoxic edema). Indeed, a leading hypothesis suggests a crucial role for endothelial dysfunction and activation in PRES pathogenesis.[1]

PRES is characterized by transient neurologic signs including headache, visual changes, seizures, and altered sensorium.[5] Cortical blindness is considered a typical and characteristic symptom of this syndrome.[6] PRES is reversible in a few days but if appropriate management is delayed there is high risk of permanent neurologic damage secondary to cerebral infarction or hemorrhage and transtentorial herniation resulting in death. Subjective cognitive problems, development of chronic epilepsy, and progress to irreversible (partial) blindness can be long-time consequences after years from acute episode. Early and late complication such as pulmonary edema, dissection of extracranial internal left carotid artery, cerebral herniation, short term memory loss, subarachnoid hemorrhage, permanent mild dysmetria, visual impairment, and death have been described.[7]

Early recognizing of symptoms is fundamental for a timely diagnosis. As reported in literature cerebral MRI is the gold standard diagnostic tool; neuroimaging performed shows diffuse edema of the white matter, which selectively involves the parieto-occipital regions of the brain; edema usually shows iso- or hypointensity in DWI.

In the differential diagnosis, cerebral venous thrombosis, bilateral posterior lobe infections, herpes virus and other viral encephalitis, and cerebral changes related with electrolyte disturbances should be considered.[8] In our patients, venous or ischemic infarction was excluded by way of diffusion-weighted MRI and encephalopathy related with infections and electrolyte disturbances was excluded by way of history, physical examination, clinical status, and laboratory tests.

There is no specific treatment for PRES. Supportive treatment directed to symptoms is the mainstay of treatment. Supportive treatment includes antihypertensives, antiepileptic drugs for seizure, discontinuation of drugs that may lead to the condition, and most importantly, controlling the underlying disease causing hypertension. Antiepileptic treatment is used in the emergency treatment of seizures related with PRES, but there is no need for long-term antiepileptic treatment.[9] The initial evaluation of patients with PRES should focus on a rapid correction of blood pressure, hydration using crystalloid fluids, and maintenance of adequate oxygenation.

A treatment focused on hypertension control; cerebral edema reduction is a successful therapy which allowed us to avoid neurological sequelae, early and late complications, and patient's death. Performing a cerebral MRI in the suspicious of PRES clinicians should be aware to detect signs of cytotoxic edema that is a sign of the development of the disease.[10]

The spread and the localization of edema are variable and could depend on the latency time between the seizure and the MRI. There is a large variability also in time of cerebral MRI normalization. According to the literature, despite the importance of cytotoxic edema, it is not linked to poor prognosis or to the development of early or late sequelae. Nowadays the hypothesis of endothelial dysfunction in the pathophysiology of PRES is also proposed. For this reason monitoring LDH serum level as marker of endothelial dysfunction could be useful.[11]

Conclusion:

The symptoms and lesions of Posterior reversible encephalopathy syndrome may resolve completely if the diagnosis and treatment is prompt, as was seen in our patient; however, failure to diagnose may lead to irreversible infarction and death. Recurrence of PRES is rare and may be associated with infections and rapid rise in blood pressure. The diagnosis may be overlooked, especially in children, unless a high index of suspicion and precise clinical history is maintained. It should be kept as a possibility in children presenting with encephalopathy and seizures with raised blood pressure as delay in diagnosis and treatment may result in permanent neurological deficit. In conclusion, PRES should be considered in the differential diagnosis of acute encephalopathy in patients who present with new-onset seizures, systemic hypertension, and clouding of consciousness. The clinical findings, underlying disease, history of drug use, and brain imaging should be evaluated together, and it should be kept in mind that complete clinical recovery can be provided with early diagnosis and appropriate therapeutic approaches.

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