Posterior reversible encephalopathy syndrome : Case Report

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Abstract :

Posterior reversible encephalopathy syndrome (PRES) is a neurological syndrome. Posterior reversible encephalopathy syndrome also known as reversible posterior leukoencephalopathy syndrome presents with rapid onset of symptoms including headache, seizures, vomiting, altered consciousness, and visual disturbance. It is associated with hypertension. The clinical syndrome usually resolves within a week and the changes seen in magnetic resonance imaging resolve over days to weeks. 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.

Introduction : Posterior reversible encephalopathy syndrome (PRES) was first described by Hinchey in 1996. It is characterised 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 vasogenicoedema which seems to be the crucial pathogenic mechanism. As the name suggests, it is typically reversible once the underlying cause is removed.[2] PRES isalso 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 presentaton to the emergency center. 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. Respiratory and cardiovascular examination was within normal limits. Abdominal examination was normal. Renal angles were non-tender. An ocular examination revealed a diminution of vision of bilateral eyes. Pupils were normally reactive to light and fundus examination was unremarkable. Rest of the cranial nerve examination was unremarkable. Power was 5/5 across all major joints and sensory function was intact all over the body. Cerebellar signs were intact and there was no evidence of meningeal signs such as nuchal rigidity or Kernig's/Brudzinski's sign. Plantars were downgoing bilaterally. Laboratory findings were significant for an elevated white cell count of 15400, haemoglobin 11.5gm/dl, platelets were 230000/cumm. Urinalysis was remarkable for 3+protein and RBC were present. PT, PTT, INR and liver profiles were within normal limits. T2-weighted images of brain MRI showed bilateral posterior parietooccipital hyper densities in the cortex and subcortical white matter consistent with posterior reversible leukoencephalopathysyndrome. 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, anticonvulsant and supportive treatment.

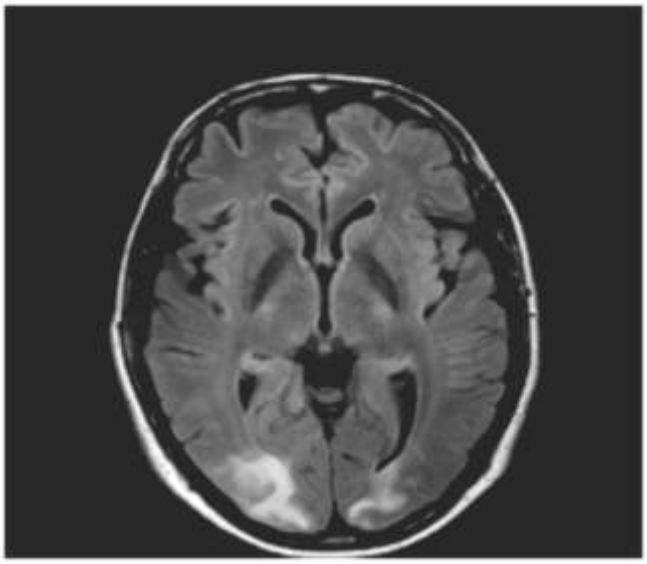

Fig :1 Bilateral, hyperintense subcortical matter lesions on MRI.

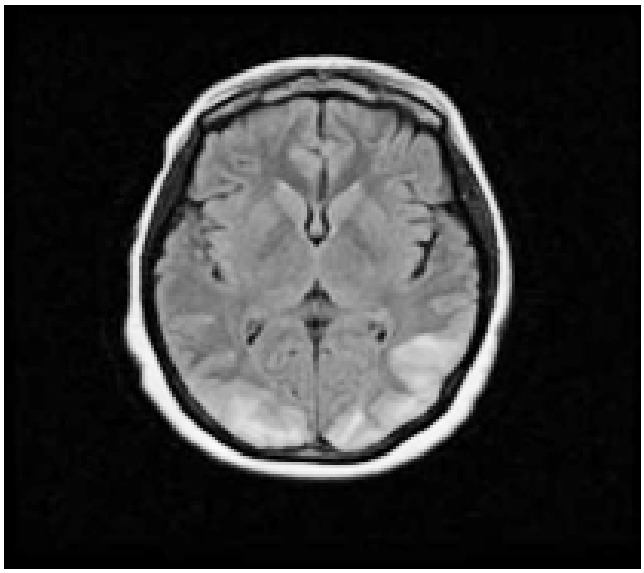

Fig : 2 Hyperintense lesion on T2 white-weighted MRI over parietal and occipital aear.

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.[4] The preferential involvement of the posterior circulation has been postulated as being due to the sympathetic innervation protecting the brain from sudden increase in blood pressure being relatively less in the arterioles supplied by the vertebrobasilar system than in the anterior circulation.[5]

One characteristic of this syndrome is that edema is present without infarction. Therefore, recognition and accurate treatment of the syndrome is imperative to halt progression, find and remove the cause, and prevent permanent damage or death.[6,7]

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. We maintain that should be kept as a possibility in children presenting with encephalopathy and seizures in the setting of raised blood pressure or renal disease as delay in diagnosis and treatment may result in permanent neurological deficit.

References

1. Hinchey J, Chaves C, Appignani B, Breen J, Pao L, Wang A, et al. Reversible posterior leukoencephalopathy syndrome. N Engl J Med. 1996;22:494–500.

2. Bartynski WS, Boardman JF. Distinct imaging patterns and lesion distribution in posterior reversible encephalopathy syndrome. AJNR Am J Neuroradiol. 2007;28:1320–7.

3. Ozcakar ZB, Ekim M, Fitoz S, Teber S, Hizel S, Acar B, et al. Hypertension induced reversible posterior leuoencephalopathy syndrome: A report of two cases. Eur J Pediatr. 2004;163:728–30.

4. Girişgen I, Tosun A, Sönmez F, Özsunar Y. Recurrent and atypical posterior reversible encephalopathy syndrome in a child with peritoneal dialysis. Turk J Pediatr. 2010;52:416–9.

5. Casey SO, Truwtt CL. Pontine reversible edema: A newly recognized imaging variant of hypertensive encephalopathy? AJNR Am J Neuroradiol. 2000;21:243–5.

6. Ahn KJ, You WJ, Jeong SL, Lee JW, Kim BS, Lee JH, et al. Atypical manifestations of reversible posterior leukoencephalopathy syndrome: Findings on diffusion imaging and ADC mapping. Neuroradiology. 2004;46:978–83.

7. Onder AM, Lopez R, Teomete U, Francoeur D, Bhatia R, Knowbi O, et al. Posterior reversible encephalopathy syndrome in the pediatric renal population. PediatrNephrol. 2007;22:1921–9.