Case Report

Atypical Presentation of Posterior Reversible Encephalopathy: In a Child with Bilateral Grade IV Vesicoureteric Reflux

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Summary

Posterior reversible encephalopathy, better known as potential reversible encephalopathy syndrome (PRES), is a clinicoradiological entity mostly described in adult populations manifesting predominantly as bilateral symmetrical diffuse white matter vasogenic oedema in parieto-occipital regions. Rarely it may also present as patchy reversible areas of involvement in the basal ganglia, brainstem and deep white matter. It is reported scarcely in childhood populations. Frequent association with acute hypertension (67–80%) is reported in many studies. Involvement of the brainstem and cervical cord (apart from the typical parieto-occipital lesions) is an extremely rare imaging manifestation of PRES and its recognition is important to avoid misdiagnosis as myelitis or acute disseminated encephalomyelitis by proper clinical correlation. We hereby report a case of PRES in a 7-year-old child showing an uncommon pattern on imaging study involving the brain as well as the brainstem and cervical spinal cord.

Key words: Brainstem, cervical cord, children, posterior reversible encephalopathy.

Case Summary

A 7-year-old boy presented to paediatric emergency with two episodes of generalized tonic–clonic seizures followed by altered sensorium. He had history of low-grade pyrexia off and on, headache mainly localized to the posterior nuchal region and occasional non-bilious non-projectile vomiting for 1 month.

On examination, child was found to be drowsy with Glasgow Coma Score (GCS) score of 9/15, pulse rate 90/min and respiratory rate of 20/min. He was afebrile and blood pressure recorded in right upper limb (supine) was 120/84 mm Hg. Central nervous system examination revealed neck stiffness with brisk deep tendon reflexes and bilateral extensor plantar. Fundus examination was normal. Contrast-enhanced computed tomography (CECT) of head was normal, and cerebrospinal fluid examination was also normal, except increased protein (200 mg%). Other supportive results suggestive of tubercular meningitis were also lacking, as child did not have history of contact with a patient with tuberculosis, had normal chest radiograph, had non-reactive Mantoux test, and had normal erythrocyte sedimentation rate. Child regained full consciousness within 24 h and blood pressure (BP) normalized with use of mannitol for 48 h, but neck pain persisted.

On Day 5, he was detected to have acute increase in BP to more than the 99th percentile, i.e. 190/100 mm Hg on repeated recordings. Magnetic resonance imaging (MRI) of the brain done at this stage showed multiple focal hyperintensities in the bilateral external capsule, lentiform nucleus, left caudate nucleus, parietal cortex, midbrain, pons and medulla and diffuse increase in intensity in cervical spinal cord on T2-weighted and fluid-attenuated inversion recovery (FLAIR) sequences (Fig. 1A and C). Initially, the possibility of acute disseminated encephalomyelitis was kept with less likelihood of atypical presentation of posterior reversible encephalopathy syndrome (PRES). Visual evoked potential testing revealed normal p100 latency bilaterally of 104 ms. Child was also investigated for cause of hypertension. Bilateral scarring of kidneys with bilateral vesicoureteric reflux grade IV was found on dimercaptosuccinic acid scan, micturating cystourethrogram and CECT abdomen. Patient was started on antihypertensive therapy with good control. Antiepileptics were continued and antibiotic prophylaxis was added. Kidney function tests were normal. Three weeks after this episode, MRI brain done was completely normal (Fig. 1B and D). A final diagnosis of atypical PRES was made.
Encephalopathy associated with acute increase in blood pressure is uncommon and described mainly in adults. Typical imaging findings are bilateral symmetrical subcortical and deep white matter vasogenic oedema limited to posterior circulation seen as hyperintensities on T2-weighted and FLAIR images. Atypical findings include focal areas of restricted diffusion (11–26%), haemorrhage in the form of focal haematoma and possible presence of subarachnoid or sulcal bleed (15%) [1]. Although reversible in most cases with resolution of cause, there is risk of neurological impairment and up to a 15% mortality rate. Hypertension is associated in 67–80% cases of PRES, but blood pressure may be normal in 20–30% cases. PRES has also been reported with chronic renal disease, chemotherapeutic agents, cyclosporine and Cushing’s disease. Exact pathophysiology of PRES is not known. Hypotheses involving impairment of cerebral autoregulation and cerebral blood perfusion resulting in blood–brain barrier dysfunction with cerebral vasogenic oedema are suggested [2].

Most of case series report seizures, altered sensorium and headache as most common manifestations that may develop over many days or recognized acutely. In a setting of hypertension with clinical symptoms and signs, typical imaging findings help
in diagnosis of PRES, as there are no consensus guidelines regarding its diagnosis. In presence of prolonged symptoms like headache, vomiting and positive neck stiffness, diagnosis can be confused, more so if imaging shows atypical involvement, as in our case. The involvement of basal ganglia, brainstem and spinal cord in PRES is extremely rare. There are only few case reports in children and adults showing this atypical presentation. Choh et al. [3] reported a case of a 17-year-old male with immunoglobulin A nephropathy presenting with PRES and showing hyperintense signals in medulla and cervical cord on T2 and FLAIR images, which is similar to the case reported here.

Unlike adults, in most (80–85%) pediatric patients, hypertension is secondary to a renal cause (parenchymal, renovascular). Most cases show complete resolution of symptoms, signs and imaging findings, as in the case reported here. In a case series of 12 children with hypertensive encephalopathy in Taiwan, the most common cause was acute glomerulonephritis and typical imaging studies resolved on follow-up [4].

In large case series by McKinney et al. [5], of 76 patients with PRES ranging from 5 to 80 years (mean, 33.5 years), most common involvement was of parieto-occipital region (98.7%). Less common areas of involvement included the cerebellum (34.2%), thalamus (30.3%), brainstem (18.4%) and basal ganglia (lentiform or caudate 11.8%), but none of the patients had spinal cord involvement.

To distinguish from demyelinating spinal cord syndromes, it is helpful to know that patients with myelitis have a deep and prolonged neurodeficit, whereas patients with spinal manifestations of PRES are either asymptomatic or recover completely.

Involvement of the brainstem and spinal cord in PRES has probably been an under-recognized manifestation, as spinal imaging is not routinely performed in PRES.

Conclusion

Atypical and rare neuroradiological presentation of PRES may pose a diagnostic dilemma, but associated clinical features like hypertension, complete diagnostic workup to find the cause and subsequent follow-up usually showing early and complete resolution may help in confirmation of diagnosis and management.

References