POSTERIOR REVERSIBLE ENCEPHALOPATHY SYNDROME

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ABSTRACT

BACKGROUND

Posterior reversible encephalopathy is a proposed clinico-neuroradiological entity characterised by headache, altered mental status, cortical blindness, seizures, other focal neurological signs and a diagnostic magnetic resonance imaging. A variety of different aetiologies have been reported like hypertension, pre-eclampsia/eclampsia, uraemia and porphyria. With early diagnosis and prompt treatment, the syndrome is usually completely reversible. We report a case of PRES in a patient with elevated blood pressure. Clinicians as well as radiologists must be familiar with this clinically frightening, underdiagnosed condition to assure timely diagnosis and treatment to prevent persistent deficits.

KEYWORDS

PRES, Reversible Encephalopathy, Cortical Blindness.

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BACKGROUND

Posterior Reversible Encephalopathy Syndrome (PRES) is a proposed clinico-neuroradiological entity with a vast spectrum of different aetiologies. The clinical hallmark of this syndrome are headache, confusion, seizures, cortical visual disturbances or blindness and less common are other focal neurological signs. A variety of causes are known like hypertension, pre-eclampsia/eclampsia, uraemia and porphyria. We report a patient of PRES in whom Blood Pressure (BP) was elevated and no other aetiology of PRES could be unfolded.

CASE REPORT

A 17-year-male admitted with chief complaints of one episode of generalised tonic-clonic convulsion. Patient has history of headache since 3 days. On admission patient was drowsy, arousable and obeying commands. His pulse was 110/min and blood pressure was 160/100 mmHg. Patient do not have any history of being medically treated for hypertension previously. Patient was investigated with abdominal sonography to rule out renal artery stenosis and pheochromocytoma as the cause of persistent hypertension in this age group, 24-hour urinary Vanillylmandelic Acid (VMA) was sent and it was normal. Blood pressure was measured manually thrice a day and was persistently raised with the highest measurements of 170/110 mmHg. Echocardiography was normal. Magnetic Resonance Imaging (MRI) scan revealed high signal intensities on T2 weighted images, which were pronounced in the parieto-occipital cortices in the both hemispheres. The Electroencephalogram

(EEG) was done, which was suggestive of ongoing seizure disorder. The patient improved neurologically over a period of 4 days and his blood pressure settled to 140/80 mmHg with antihypertensives.

Figure 1. High Signal Intensities on T2 Weighted Images, which were Pronounced in the Parieto-Occipital Cortices in the both Hemispheres

DISCUSSION

PRES can be diagnosed with (massive) reversible hyperintensities on T2-weighted cranial MR-images. Predominantly, it affects the territory of the posterior circulation and the clinical hallmarks are headache, vomiting, confusion, seizures, cortical visual disturbances or blindness and less common are other focal neurological signs.^{1,2,3} Modern imaging modalities (Diffusion Weighted Imaging (DWI) with apparent diffusion coefficients (ADC-maps) could prove that most cases of PRES are caused by vasogenic oedema rather than cytotoxic oedema.^{4,5} The pathogenesis is

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attributed to a failure of cerebral autoregulation that probably is facilitated in posterior brain regions due to a sparse sympathetic innervation of the vertebrobasilar vascular system.^{4,6} Isolated brainstem involvement is rare, but has even been reported as a clinically silent reversible phenomenon.^{7,8,9} In any case, the absence of abnormalities on DWI (with increased ADC) can be interpreted as a favourable prognostic finding, whereas increased signal on DWI or reduced ADC point towards frank ischaemia with cytotoxicity and cell loss.¹⁰

The cause of PRES remains controversial, but the most popular theory is that severe hypertension causes interruption to brain autoregulation.¹¹ Cerebral blood flow is usually regulated by dilatation and constriction of vessels to maintain adequate tissue perfusion¹¹ and to simultaneously avoid excessive intracerebral hypertension. Breakdown in autoregulation occurs above a mean arterial blood pressure of 150 - 160 mmHg in chronic hypertension, it occurs at higher pressures.¹¹ Uncontrolled hypertension leads to hyperperfusion and cerebral vessel damage resulting in interstitial extravasation of proteins and fluids causing vasogenic oedema. Irreversible damage is seen at mean arterial pressure above 200 mmHg.¹¹

The key to diagnosis in PRES naturally is the Magnetic Resonance Imaging (MRI), but suspicion must be raised by the clinician who should be familiar with this underdiagnosed, clinically frightening syndrome to avoid persistent deficits.^{10,12} It mostly is a benign, reversible condition, especially once the causative factor (eg. hypertension) is eliminated.

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