Brain stem Posterior reversible encephalopathy-A study of four cases

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Abstract:
INTRODUCTION Posterior reversible encephalopathy syndrome is characterized by clinical feature of headache, visual disturbance, altered sensorium, seizure and focal neurological signs with typical neuroimaging feature of involvement of bilateral parieto-occipital lobe and subcortical white matter. However atypical involvement of other regions like frontal lobe, basal ganglia, thalamus and brain stem are less frequently reported. We report here four cases of atypical Posterior reversible encephalopathy with predominant brain stem involvement.

Methods Four patients of brain stem posterior reversible encephalopathy from institute of neurology, madras medical college during the year 2010-2011 were studied.

Diagnosis was based on history, clinical examination and neuroimaging findings. Other basic investigation including renal function were done.

Results The patients were between 34-54 years of age. Three were males and one was female. The onset was acute in two and subacute in two patients.

All of them presented with hypertension, altered sensorium, and blurring of vision. Two were know hypertensives. Unsteadiness was noted in two patients and one had lower motor neuron facial palsy. None of them had seizure. Two patients had renal failure in whom hypertensive retinopathy changes was noted in one patient. Neuroimaging of all four patients showed predominant involvement of brain stem. Two patients in addition had thalamic and parieto-occipital region involvement. Antihypertensives were administered for hypertension control. Complete resolution of clinical symptoms and radiological lesions occurred in all four patients within few days to two weeks.

Conclusion This paper is presented to highlight atypical clinical and neuroimaging findings of posterior reversible encephalopathy. Posterior reversible encephalopathy should be considered in the differential diagnosis of brain stem lesions with background history of hypertension in addition to other causes.
**Introduction:**
Posterior reversible encephalopathy syndrome is characterized by clinical feature of headache, visual disturbance, altered sensorium, seizure and focal neurological signs with typical neuroimaging feature of involvement of bilateral parieto-occipital lobe and subcortical white matter. However atypical involvement of other regions like frontal lobe, basal ganglia, thalamus and brain stem are less frequently reported. We report here four cases of atypical Posterior reversible encephalopathy with predominant brain stem involvement.

**Methods:** Four patients of brain stem Posterior reversible encephalopathy from institute of neurology, madras medical college during the year 2011-2012 were studied. Diagnosis was based on history, clinical examination and neuroimaging findings. Other basic investigation including renal function were done.

**CASE 1:**
A 50 year old male presented with acute onset of headache, vomiting, blurring of vision and altered sensorium with unsteadiness of gait 3 days after onset of headache. His blood pressure was 210/110 mm Hg. He was initially drowsy, after improvement in sensorium his visual acuity was 20/200 in both eyes. He had stance and gait ataxia. His renal parameters were blood urea – 92mg% and creatinine -8mg%. MRI - FLAIR sequences showed marked high-intensity signal changes of the brainstem and cerebellum with periventricular lesions. DWI showed iso-intensity in the brain stem. Corresponding ADC map showed increased diffusion in the brain stem lesions.

**CASE 1 FOLLOW UP:**

**CASE 2:**
A 57 year old non hypertensive male presented with history of difficulty in closing right eye, blurring of vision in both eyes and deviation of angle of mouth to left side. His blood pressure was 260/130 mmHg. He was conscious, oriented, visual acuity– 20/100. Fundus examination showed hyperemic disc, margins blurred, peri papillary hemorrhages, AV crossing changes and bilateral exudates suggestive of grade 4 hypertensive retinopathy. He had right lower motor neuron facial nerve palsy, spinomotor system and sensory examination were normal. His blood urea was 71mg% and creatinine was 2.5 mg%. MRI - FLAIR sequences showed marked high-intensity signal changes of the brainstem and periventricular region. The brain stem and cerebellar lesions.

**Keyword:** Brain stem PRES, PRES, ATYPICAL PRES
Case 3: A 44-year-old hypertensive male had headache, vomiting, blurring of vision, unsteadiness, and altered sensorium. His blood pressure was 210/120 mmHg. He was drowsy but able to follow simple commands. No other focal neurological deficits were present. His renal parameters were normal.

Case 4: A 54-year-old woman with a history of poorly controlled hypertension presented with headache, visual blurring, and altered sensorium. Her blood pressure was 220/114 mmHg. She was drowsy and no focal neurological deficits were detected. Her renal parameters were normal.

DISCUSSION:
The age group of our patients was 35-55 yrs. Three of them were males. Two patients had an acute onset and the other two had subacute onset. All patients presented with hypertension, visual blurring and...
altered sensorium with two of them had a history hypertension previously, gait ataxia was noted in two and one had a lower motor neuron facial palsy. None of them had seizure. Two patients had renal failure of which one had hypertensive retinopathy. Neuroimaging of all four patients showed predominant involvement of brain stem. Two patients in addition had thalamic and parieto-occipital region involvement. Complete resolution of clinical symptoms and radiological lesions occurred in all four patients within few days to two weeks following control of hypertension. Reports of predominant brain stem posterior reversible encephalopathy are less frequently reported. Salvador cruz et al studied 23 patients of brain stem posterior reversible encephalopathy. Mean age at onset was 42 years; 12 were men. All patients had hypertension. Other comorbidities were renal failure, eclampsia, pheochromocytoma, systemic lupus erythematosus. Headache, vomiting, blurred vision, abnormal gait, and paresis were the most frequently reported similar to our cases but seizure was not noted in any of our patients. Hypertension and renal failure were found in our patients also. Two hypotheses may explain the abnormalities in hypertensive encephalopathy. One proposes that infarction caused by fibrinoid necrosis and thrombosis of arterioles results in cytotoxic edema appearing as high signal in T2-weighted, FLAIR, and DWI images and as low signal in the ADC map. \(^2,^3\) The other proposes that severe hypertension that exceeds autoregulation results in segmental vasodilatation and increased vascular permeability leading to vasogenic edema that appears as high signal in FLAIR images and the ADC map and as low or normal signal in the DWI. \(^2,^3\) Reversibility of lesions supports the second hypothesis. Brain stem variants \(^5\) of posterior reversible encephalopathy should be differentiated from brain stem infarction, pontine glioma, central pontine myelinolysis, and infective encephalitis because the neurologic symptoms are reversible with prompt treatment in posterior reversible encephalopathy. Despite the presence of extensive lesions in neuroimaging, there are few symptoms or signs of brainstem or cerebellar dysfunction, a typical “clinical radiologic dissociation” present in posterior reversible encephalopathy.

**Conclusion:** This paper is presented to highlight atypical clinical and neuroimaging findings of posterior reversible encephalopathy. Posterior reversible encephalopathy should be considered in the differential diagnosis of brain stem lesions with background history of hypertension in addition to other causes.

**References:**


