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Case Report

Posterior Reversible Encephalopathy Syndrome with Spinal Cord Involvement –A Case Report

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Introduction

Posterior reversible encephalopathy syndrome also known as reversible posterior leukoencephalopathy is a neurotoxic state that occurs secondary to inability of posterior circulation to auto regulate in response to acute changes in blood pressure. It is a clinic-radiological entity with white matter vasogenic edema predominantly affecting posterior brain in relatively symmetric pattern. Involvement of the cord in PRES has probably been an under recognized entity as spinal imaging is not routinely performed in posterior reversible encephalopathy syndrome .Involvement of spinal cord is extremely rare with about 21 case reports described in literature. We present one such interesting case of PRES with spinal cord involvement (PRES-SCI).

Case Report

A previously healthy 23 years old female presented with severe bifrontal headache associated with recurrent vomiting and transient visual obscurations for 3 days with background history of headache for past 2 months. There was no preceding history of fever, rashes, cough and diarrhoea. There was no alteration in the level of consciousness, seizures and weakness of limbs. On examination her blood pressure was high (190/100 mm of hg). A complete neurological evaluation revealed no focal motor, sensory and bladder deficits. Fundus examination revealed bilateral papilloedema with grade 4 hypertensive retinopathy.

On investigating, she was found to have persistent hypokalemia with normal serum urea and creatinine levels .USG abdomen and renal artery doppler was normal. Aldosterone levels were elevated (51.4ng/dl) with low plasma renin activity (0.81ng/ml/hr). MRI brain and whole spine showed confluent areas of T2 FLAIR hyper intensity in pons, medulla, bilateral cerebellar hemisphere, occipital and temporal white matter. Spine imaging also revealed long segment signal alteration involving cervical cord from cervicomedullary junction. CSF analysis was normal with negative oligoclonal bands .Her visual evoked potential was normal. Her ENA profile was also within normal levels. CT imaging of abdomen was normal.

Her symptoms subsided with anti-edema measures and antihypertensive. Patient was put on aldosterone antagonist with strict salt restriction. Follow up imaging revealed complete resolution of lesions.

Discussion

PRES is a clinico radiological syndrome with constellation of clinical finding of hypertensive encephalopathy presenting with headache, vomiting, seizure, altered level of consciousness, visual impairment and papilloedema with reversible vasogenic subcortical edema on imaging. PRES typically involves parieto occipital region.

Acute rise in BP causes down regulation of cerebral auto regulation leading to hyper perfusion. This hyper perfusion causes surge of pro inflammatory cytokines which induces endothelial damage and there by vasogenic edema. The other mechanism is related to cytotoxicity and involves endothelial dysfunction, blood brain barrier alteration and cerebral hypo perfusion resulting in vasogenic edema [1]. Posterior cortical dominance is due to poor sympathetic innervation of vertebrobasilar system. Anterior spinal artery arising from posterior circulation explains the cord lesions in the cervical region [2].

MRI is more sensitive displaying hyperintense lesions in T2 and FLAIR sequences with increased ADC values which are characteristic of vasogenic edema .Subcortical areas are predominantly involved due to lower density of these areas. ADC values are of prognostic significance. Higher ADC values have been associated with reversibility of lesions. Functional imaging may be helpful in viewing the perfusion status of brain and the resultant pathology causing PRES [3].

Schwatz et al [4] in their CT, MR and SPECT imaging series of 14 cases of hypertensive encephalopathy had not described spinal cord involvement.

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Bartynski et al. [5] reviewed the MRI imaging of 136 cases of PRES and described three common hemispheric patterns of involvement: holohemispheric, parietal–occipital and superior frontal sulcal patterns, with overall parietal–occipital involvement found in 98% of their cases. These typical patterns can have variable expression or asymmetry in an individual patient. They also found involvement of the cerebellum, brainstem, temporal lobes, splenium and basal ganglia in atypical cases. Isolated involvement of these structures without parietal–occipital involvement is, however, uncommon. Spinal cord involvement was not described by Bartynski.

Choh et al. [6] had reported a case of IgA nephropathy with high BP and similar clinical features with T2 hyperintensity in medulla and cervical cord.

Haven et al. [7] have analyzed about 8 cases reported in literature then until 2014, of which 7 had hypertensive retinopathy , 4 patients had symptoms referable to cord and only 1 patient had seizure. All 8 patients had severe acute hypertension and confluent expansile central spinal cord T2 hyper intensity spanning at least 3 to 4 segments. 4 of these 8 patients with spinal cord lesions showed no contrast enhancement. Follow up imaging was normal in 7 out of 8 patients.

KoKhar et al. [8] Reported a case of Henoch schonlein purpura with nephropathy which had grade 4 retinopathy changes. MRI showed altered signal intensity areas involving bilateral cortical and subcortical parieto-occipital regions, bilateral cerebellar hemispheres, medulla, cervicomedullary junction, and cervical spinal cord until C6. Follow-up MRI after 25 days, showed complete resolution of the lesions of brain and cervical spinal cord.

Agarwal et al. [9] reported a case of aortoarteritis involving descending aorta with renal artery stenosis who presented with headache ,seizures, grade 4 retinopathy changes and long segment T2 hyperintensity with minimal cord expansion involving the

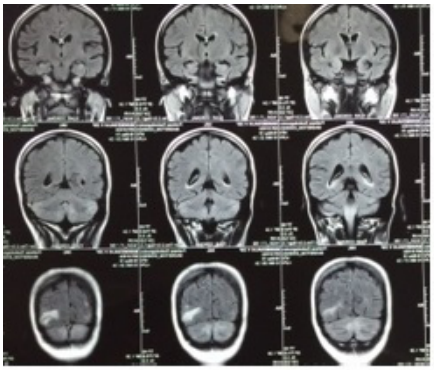

Fig 1: T2 Flair MRI showing hyperintensities in posterior brain regions

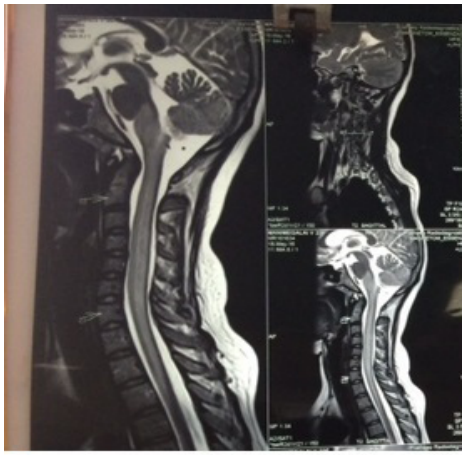

Fig 2: T2 MRI showing hyperintensity in brain stem extending to the spinal cord

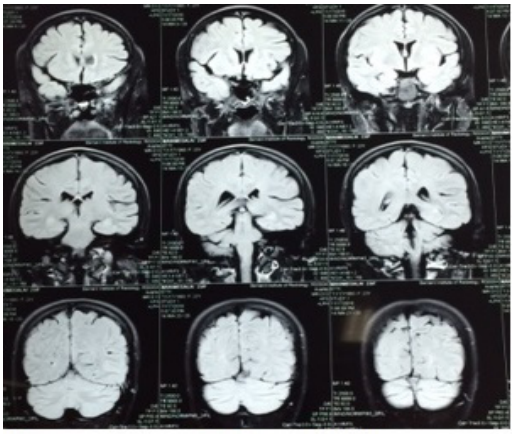

Fig 3 : Repeat MRI Brain T2 image showing resolution of hyperintensities in posterior brain regions

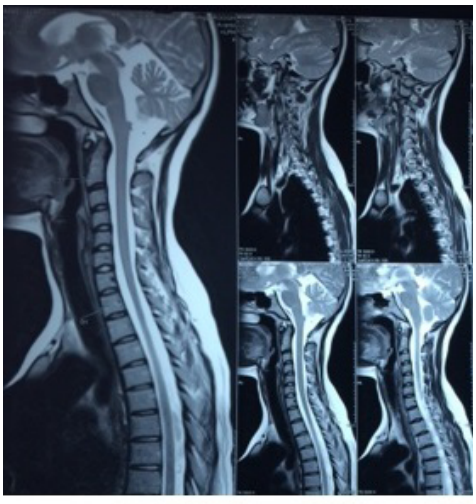

Fig 4: Repeat MRI T2 showing complete resolution of hyperintensities in spinal cord.

cervicodorsal spinal cord with complete regression of all changes in the brain and spinal cord in repeat MRI after 28 days .

Tai- Heng et al. [10] reported 2 cases : a 16 yr. old MPGN and an 8 yr. old with nephrotic proteinuria .In addition to the hypertensive encephalopathy features one presented with seizure and both had lower limb weakness with sensory involvement of which one had bladder and bowel incontinence. Both had elevated CSF protein. Both had T2 hyper intensity in cervical cord extending up to thoracic segments in addition to posterior brain regions. All neurologic symptoms subsided dramatically after blood pressure normalization in both the patients. Follow-up brain- and spinal MRI 2 months later showed resolution of the lesions.

Tai- Heng et al. [10] also analyzed 21 patients (8 were children) reported in literature before 2016. Regardless of age, the most common clinical symptom was headache and the least common clinical symptom was seizures. Atypical neuroimaging was more common in children (63%) than in adults (8%). All children recovered uneventfully, but 3 adults had sequelae.

Our patient did not have seizure, but presented similarly with headache, vomiting, blurred vision, papilledema with grade 4 hypertensive retinopathy and long segment T2 hyper intensity in cervical cord in addition to posterior brain regions and brain stem with no signs and symptoms referable to cord which completely normalized in follow up imaging.

Differential diagnosis of longitudinally extensive spinal T2 hyper intensity includes myelitis secondary to infection, autoimmune diseases, malignancy and vascular myelopathy from dural arteriovenous fistula. Typically patients with myelopathy have a profound neuro deficit, while patients with spinal manifestations of PRES are generally asymptomatic.

When compared to PRES, PRES-SCI presents at younger age with severe acute hypertension with high rates of headache, vomiting, visual disturbance, hypertensive retinopathy and very low rates of seizures and encephalopathy and involves central cervical spinal cord reversibly with or without cord symptoms [6,9-12].

Conclusion

Clinicians should be aware of this rare presentation PRES-SCI and thereby suspect this scenario in patients with extreme elevation of blood pressure and its related symptomatology and MRI lesions in brainstem and spinal cord with grade 4 hypertensive retinopathy with or without neurological signs referable to cord. Therefore, it is necessary to consider PRES-SCI as a differential diagnosis in situations having T2 hyper intensity in spinal cord implying longitudinal extensive transverse myelitis in appropriate clinical setting of high blood pressure related manifestations.

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