Posterior Reversible Encephalopathy Syndrome in a Postoperative Patient-A Case Report . Prof. R. M. Bhoopathy, S. Hariharan PG

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Abstract :
Posterior Reversible Encephalopathy Syndrome (here in after referred to as PRES) is classically characterized as parieto occipital edema but may occur in other distributions with varying imaging appearances. We report a case of 36yr old male who underwent hemorrhoidectomy and postoperatively presented with features of PRES.

Keyword : PRES, pseudonormal ADC, vasogenic edema

Introduction:
PRES is a neurotoxic state coupled with a unique CT (computedtomography) or MR (magnetic resonance) imaging appearance. PRES was commonly identified in the clinical setting of severe hypertension and eclampsia. Of late PRES is diagnosed in varying clinical settings with a background of hypertension and neurotoxic symptoms coupled with MR imaging of symmetric vasogenic whittematter edema in typical distribution patterns.

Case report:
Our patient was a 36 yr old male who underwent hemorrhoidectomy. On the 2nd POD (post operative day) he developed headache and vomiting. His sensorium was intact. There was no visual disturbances or focal neurological deficits. On the 6th POD he developed seizures - Left focal motor seizures with secondary generalization. The seizures controlled with antiepileptics (T.Carbamazepine,inj.Phenytoin and inj.Diazepam). In the postictal state visual disturbances were noted in the form of difficulty in identifying objects. Paucity of movements of the left side of the body was also observed. On examination, he was afebrile and the blood pressure was 140/90mmHg. There was no papilledema or facial asymmetry. Bilateral menace reflexes were absent. Bilateral plantar reflexes were absent. There was no neck stiffness and Kernig sign was negative. Sensory and cerebellar evaluation could not be performed as the patient was drowsy. Complete blood count, renal function tests, liver function tests and serum electrolytes were within.
normal limits. VDRL and HIV serology were negative. Chest X ray and electrocardiogram were normal. Echocardiography showed normal cardiac status. Blood and urine culture showed no growth. CSF analysis including pressure evaluation was normal. MRI revealed right frontal and bilateral parieto occipital subcortical white matter hyperintensities in T2WI, FLAIR sequences. Corresponding mild hyperintensity in the Diffusion weighted images were noticed. On ADC map there was no diffusion restrictions suggesting presence of vasogenic edema rather than cytotoxic edema in the brain. MR Venogram and MR Angiogram were normal.

In the repeat MRI (taken 20th POD) symmetric extensive, confluent white matter hyperintensities were observed throughout the bilateral cerebral hemispheres. Absence of diffusion restrictions suggested vasogenic edema. No infarcts or hemorrhage in the brain. No dural venous sinus thrombosis.

diffusion weighted image (taken on 6th POD) ADC (Apparent diffusion Coefficient) mapping showing Pseudonormalization (taken on 6th POD)
There were no further seizure episodes. His vision recovered completely over next 3 weeks. At discharge the patient had left spastic hemiparesis and upper motor neuron type of facial palsy.

On the 3rd month of follow up, MRI showed gliotic changes in the bilateral parietooccipital white matter – suggestive of chronic infarcts. He continues to be on Tab. Carbamazepine and Tab. Phenobarbitone. No further seizure episodes till date.

Discussion:

PRES is a clinico-radiological entity wherein the patient presents with clinical signs of neurotoxicity while MR imaging reveals symmetric subcortical whitematter hyperintensities in the bilateral cerebellar hemispheres. PRES is diagnosed in various clinical conditions. The clinical conditions predisposing to PRES include:

- Toxemia of pregnancy (preeclampsia/eclampsia)
- Post transplantation
- Allogenic-BMT
- Immune suppression drugs:
  - Cyclosporine
  - Tacrolimus (FK-506)
- Infection/sepsis/shock:
- Systemic inflammatory response syndrome
- Multiorgan dysfunction syndrome
- Autoimmune diseases:
  - Systemic lupus erythematosus
  - Systemic sclerosis (scleroderma)
  - Wegener’s granulomatosis
  - Polyarteritis nodosa
- Combination high-dose chemotherapY

The clinical signs of neurotoxicity include headache, nausea, altered mentation, cortical visual disturbances, and seizures later leading to coma. Symptoms may present acutely or present over several days. 70-80% of the patients in the neurotoxic state have elevated blood pressure. In the remaining 20-30% of patients the blood pressure is within normal limits or is only minimally elevated. The term PRES is adopted in these cases based on the similarity in imaging and predominance of posterior – parietooccipital location. The basic PRES pattern resembles the watershed zones, with the cortex and subcortical / deep white matter involved to varying degrees. Three imaging patterns may be encountered in similar frequencies (holohemispheric, superior frontal sulcal and parieto-occipital patterns). These patterns basically demarcate the watershed between the medial and lateral hemispheric blood supply. Lateral hemispheric supply from the middle cerebral artery, medial hemispheric supply from the anterior and posterior cerebral arteries. In holohemispheric watershed pattern vasogenic edema will be present in a linear pattern spanning the frontal, parietal and occipital lobes.
It demonstrates watershed between the entirelateral and medial hemispheric supply. In the superior frontal sulcus pattern distinct involvement of the frontal lobe along the superior frontal sulcus will be observed. The classical parieto-occipital involvement is the third pattern of involvement. Partial or asymmetric expression of the three patterns is also common. The pathogenesis of PRES is widely debated and controversial. Hypertension leading to failure of cerebral autoregulation and hyperperfusion is the commonly suggested theory. Hypertension leading to vasoconstriction and hypoperfusion is the alternative theory. The watershed distribution in imaging favors the second theory. Potential etiological factors include endothelial injury as is present in eclampsia or other causes such as conditioning regimens before transplantation, graft versus host effects, or the effects of the immunosuppressive drugs such as cyclosporine. PRES usually shows high signal-intensity lesions on FLAIR (fluid attenuated inversion recovery) images, predominantly localized in (but not limited to) the subcortical white matter (mainly association fibers) of bilateral occipital and parietal lobes. On DWI (diffusion weighted imaging), focal lesions of subtle bright signal-intensity lesions not enough to call acute infarct support the hypothesis of vasogenic edema of brain tissue. The concurrent increased apparent diffusion coefficient (ADC) value over the lesion site also proves this theory. A conundrum arises in the setting of DWI hyperintensity and "pseudonormal" ADC from intravoxel averaging of focal cytotoxic edema in larger areas of vasogenic edema; these lesions can progress to infarction. Several theories address this cytotoxic effect. First, hyperperfusion may cause severe mass effect from vasogenic edema compressing the local microcirculation, with pseudonormal or slightly elevated ADC values surrounded by larger areas of vasogenic edema. Second, vasoconstriction may be a response to the edema, eventually causing cytotoxicity if not reversed; accordingly, narrowed intracranial arteries have been noted in hypertensive human and animal models. Third, spasm could occur in response to the subarachnoid hemorrhage that uncommonly occurs in PRES. Also, aggressive correction of hypertension may induce ischemia. We present this case as it had few atypical manifestations not seen in the classical PRES cases. Our patient had no hypertension. Secondly imaging revealed a holohemispheric pattern. Thirdly even though the MRI consistently showed vasogenic edema the patient went on to develop infarcts highlighting the pseudonormalization of ADC map. Lastly the patient progressed to develop left spastic hemiparesis in contrast to absence of focal neurological deficits in classical PRES.

Conclusion:

PRES develops in patients with complex systemic conditions such as eclampsia, after transplantation, in infection/sepsis/shock and autoimmune disease, and after cancer chemotherapy. Hypertensions absent in some patients (upto 20% of pts) and, when present, does not typically reach the level of failed autoregulation. The imaging appearance typically demonstrates symmetric vasogenic edema with several characteristic patterns, generally representing distribution between lateral and medial cerebral arterial branches (i.e., a watershed distribution). Awareness of this variability in patterns is important in proper recognition of PRES, when present.
References:


