Case Report

Posterior reversible encephalopathy syndrome and spontaneous spinal epidural hematoma with pregnancy induced hypertension : a rare association

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Posterior Reversible Encephalopathy Syndrome (PRES) is reversible condition characterised by typical neurological and radiological features. Spontaneous spinal epidural hematoma (SSEH) is a rare entity, mostly associated with bleeding disorders, anticoagulant therapy, arteriovenous malformation, arteritis, and sometimes iatrogenic. We report a young pregnant female with a rare association of atypical PRES and SSEH along with pregnancy induced hypertension.

[*J Indian Med Assoc* 2019; **117:** 30-1 & 33]

Key words **: Posterior Reversible Encephalopathy Syndrome, Spontaneous spinal epidural hematoma, Pregnancy induced hypertension.**

Posterior Reversible Encephalopathy Syndrome (PRES), a condition which is usually reversible, is characterised by typical neurological and radiological features associated primarily with severe hypertension, preeclampsia/eclampsia, treatment with immunosuppressive drugs and renal disease[1]. Spontaneous spinal epidural hematoma (SSEH) is a rare entity, associated with conditions like bleeding disorders, anticoagulant therapy, vascular malformation, and arteritis[2]. Hypertension has also been reported as a rare cause of SSEH[3]. We report an uncommon case with PRES and SSEH in the setting of pregnancy induced hypertension.

Case Report

A 32 year old female with 8 months pregnancy developed acute, severe back pain, weakness of both lower limbs along with decreased sensations below waist and retention of urine. Few hours later she developed headache, irrelevant talking and inability to recognise family members. This was not associated with fever or seizures. She was admitted to a nearby hospital, where her blood pressure was recorded 210/130 mm Hg and was treated with antihypertensives. MRI brain revealed T2 and FLAIR hyperintensities in bilateral fronto-parietal and periventricular white matter, bilateral basal ganglia, thalami, brainstem and cerebellum (Fig 1). MR angiogram of brain was normal. MRI dorsal spine showed epidural hematoma at D9 to D12 level, compressing the cord (Fig 1). There was no evidence of any well formed nidus, abnormal bunch of vessels, any prominent vein or arterial feeder which could have raised suspicion of arteriovenous malformation. Her coagulation profile was normal. After 3 days of treatment, her sensorium and headache improved but deficit in lower limbs persisted. Next day she delivered a still born child with caesarean section and was referred to our centre for further management, but she reported after 3 months. Her past history revealed recurrent neurological deficits during peripartum period in each of her previous three pregnancies which developed acutely and were accompanied by high blood pressure and moderate intensity headache. These deficits resolved completely over next 3-7 days with antihypertensive and supportive treatment. Previous treatment record or scans were not available. She was normotensive between her pregnancies and did not have history of foetal loss or still birth before the present illness. There was no past history of seizures, diminution of vision, prolonged or excessive bleeding or repeated blood transfusions, anticoagulant drug intake, orogenital ulcers, joint pain, weight loss, any spinal procedure or trauma.

Her general physical examination showed pallor, blood pressure was 130/80 mm Hg. She was conscious and oriented with normal higher mental functions. Cranial nerves' examination including fundus was normal. Motor system examination in upper limbs was normal, while she had hypotonia, power of MRC grade 0/5, absent deep tendon reflexes, and extensor plantar response in both lower limbs. All sensory modalities were impaired below waist.

She had microcytic hypochromic anemia (Hb- 7.2 gm %). Her coagulation profile was normal. Serum chemistry reports were within normal limits. CRP was positive, while tests for RF, ANA antiphospholipid antibodies, HIV and treponemal serology were negative.

Repeat MRI brain revealed near complete resolution of signal abnormalities except for few small circumscribed hyperintensities in periventricular white matter (Fig 2). Repeat MRI of dorsal spine showed myelomalacia at D10, D11 level (Fig 2). There was no evidence of any abnormal vessel in this scan too which could have suggested the possibility of spinal AVM. So CT angiogram of dorsal spine was not ordered. In view of the reversible clinical and radiologic affection of brain, associated spinal epidural hematoma and subsequent myelomalacia, and the presence of pregnancy induced hypertension, a diagnosis of PRES with SSEH was made.

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Poor prognosis regarding neurologic deficit in lower limbs was explained to the patient.

Discussion

PRES is a reversible clinico-radiological syndrome which is prevalent worldwide, and is seen in persons of all age and sex[4]. There are several hypotheses proposed to explain the poorly understood pathogenesis of PRES[4] which includes (1) breakdown of cerebral autoregulation due to a sudden rise in blood pressure leading to disruption of the blood–brain barrier, (2) endothelial dysfunction due to circulating toxins which affect the blood–brain barrier leading to subsequent extravasation, (3) focal vasospasm leading to decreased blood flow and ischemia with resultant edema.

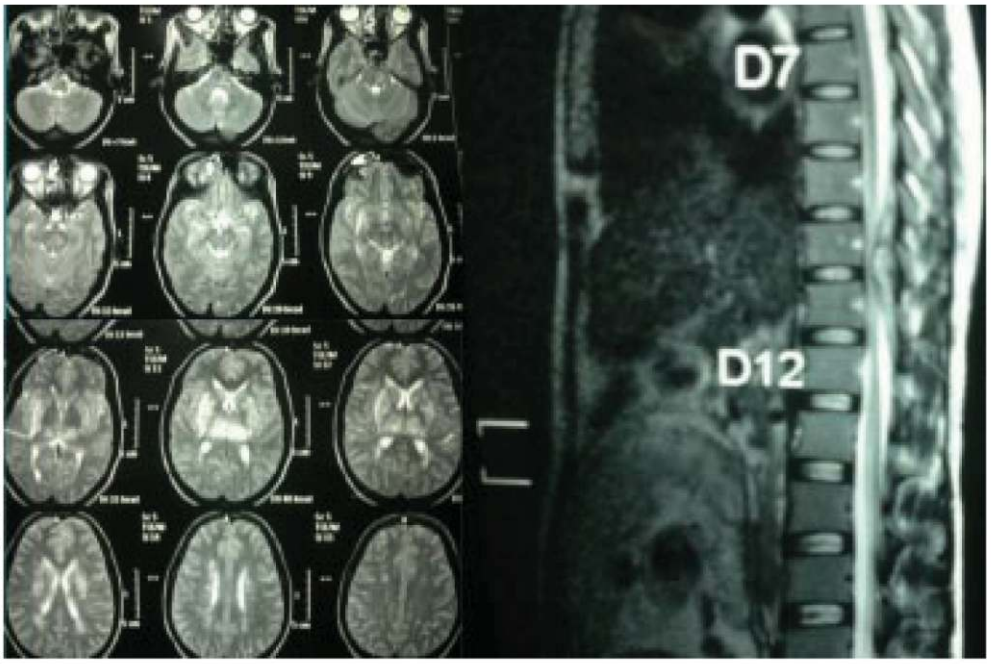

Fig 1 — MRI brain and dorsal spine at the onset of illness

Encephalopathy and seizures are the main presenting symptoms in PRES followed by headache, visual abnormalities and focal neurological deficits[5]. Acute hypertension is reported in more than 80% of patients with PRES[1]. It is classically associated with subcortical vasogenic edema, with preferential affection of the occipito-parietal regions of brain[5]. Frontal lobe involvement is seen in 51-77% of cases[5,6]. Lesions in temporal lobe, cerebellum, brainstem, basal ganglia and corpus callosum are also reported, though these locations are considered atypical for PRES[5,7]. Diagnosis of PRES in our patient was quite evident in the presence of reversible clinico-radiologic findings and pregnancy induced hypertension. Reversible cerebral vasoconstriction syndrome (RCVS) was also considered as a differential diagnosis, but ruled out on the basis of absence of characteristic thunderclap headache and normal brain angiogram. The recurrence of reversible focal neurologic deficits in our patient can be explained by recurrent PRES. Only few cases of recurrent PRES have been reported in literature, the possible triggers for recurrence being infection and hypertension[7].

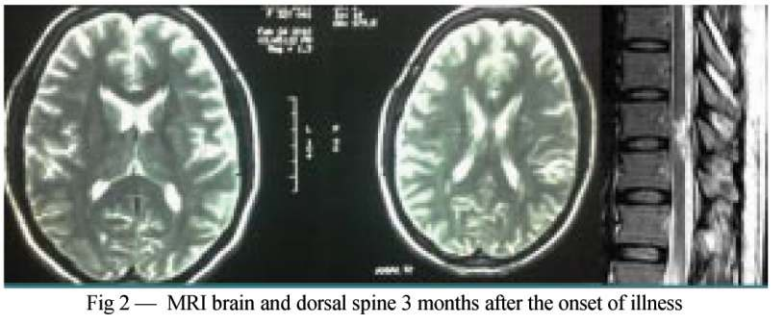

Fig 2 — MRI brain and dorsal spine 3 months after the onset of illness

SSEH is a rare condition, responsible for less than 1% of the spinal epidural lesions. Mostly it is associated with anticoagulant therapy, inherited or acquired bleeding disorders, arteritis, vascular malformation and rarely with hypertension[3]. Neurological signs and symptoms include sharp, radiating back or neck pain, progressive sensorimotor affection, and bowel-bladder disturbances, which develop acutely within minutes to hours, corresponding to the level of the spinal cord affection. The pathogenesis of SSEH is not clear. Most authors accept epidural venous plexus as the source of hematoma while others believe that the spinal epidural artery is the ruptured vessel[2]. The gold standard management is emergency evacuation of the hematoma and spinal cord decompression. Prompt surgical decompression is associated with favourable outcome. Conservative treatment may be sometimes successful in cases with minimal neurologic deficits or patients showing rapid spontaneous improvement[8,9]. In our case, the possibility of coagulation disorder and spinal vascular malformation was ruled out appropriately. The spinal epidural hematoma and the reversible cerebral lesions were probably the consequence of hypertension. Pregnancy induced hypertension overwhelmed the cerebral autoregulatory mechanisms leading to brain edema and similarly led to the extravasation of blood in spinal epidural space[3]. Functional deficit could not be improved because of the lack of surgical decompression at appropriate time.

This case is being reported due to the rare association of recurrent PRES with SSEH, in the setting of pregnancy induced hypertension.

Conflict of Interest : No financial interests, direct or indirect, exist for any of the individual contributor in connection with the content of this paper. There were no sources of outside support for the project.

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