The Posterior Reversible Encephalopathy Syndrome

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ABSTRACT

The posterior/potentially reversible encephalopathy syndrome is a unique syndrome encountered commonly in hypertensive encephalopathy. A 13-year-old boy presented with of intermittent high grade fever, throbbing headache and non-projectile vomiting for 5 days. The patient had a blood pressure of 120/80mmHg but fundoscopy documented grade 3 hypertensive retinopathy. The patient improved symptomatically following conservative management. However, on the 5th post-admission day headache reappeared, and blood pressure measured at that time was 240/120 mmHg. Neuroimaging suggested white matter abnormalities. Search for the etiology of secondary hypertension led to the diagnosis of pheochromocytoma. Repeat MRI after successful surgical excision of the tumor, patient showed reversal of white matter abnormalities. Reversible leucoencephalopathy due to pheochromocytoma have not been documented in literature previously.

Key words: The Posterior Reversible Encephalopathy Syndrome; Hypertensive encephalopathy; Metachromatic granules; Pheochromocytoma

The Posterior/potentially reversible encephalopathy syndrome (PRES) is a recently proposed clinico-neuroradiological entity. The common causes of PRES are hypertensive encephalopathy, eclampsia, cyclosporin-A neurotoxicity and uremic encephalopathy. A relatively symmetrical pattern of involvement, typically in the subcortical white matter and occasionally in the cortex of the posterior circulation area of the cerebrum is evident on magnetic resonance imaging (MRI) studies. Although most patients of PRES are markedly hypertensive at presentation, some have only mildly elevated or even normal blood pressure.

CASE REPORT

A 13-year-old boy was referred to us with a 5-day history of intermittent episodes of high grade fever associated with throbbing headache and two episodes of nonprojectile vomiting. There were no histories of cough, respiratory distress, dysuria, unconsciousness or convulsion. The patient was restless on presentation with a blood pressure of 120/80mm of Hg. Abdominal examination revealed a 2 cm splenomegaly. There were no signs of meningeal irritation, lymphadenopathy or sternal tenderness. Fundoscopic examination documented grade 3 hypertensive retinopathy. Neurological evaluation revealed normal tone in the limbs without any motor or sensory deficit. The patient had preserved reflexes with down going plantars. Examination of other systems did not reveal any abnormality. All the peripheral pulses were equally palpable without any carotid or renal bruit.

Complete Blood Count showed mild leucocytosis with ring forms of Plasmodium falciparum on peripheral smear. Blood biochemistry (e.g., glucose, urea, creatinine, liver function test, serum sodium, potassium) and urine analysis were within reference range. He was put on oral quinine sulphate and on the third day of therapy fever and headache subsided completely.

However, on the 5th post-admission day headache reappeared, though the patient remained afebrile. Blood pressure measured at that time was 240/120 mm of Hg. The patient also developed abnormal posture of his limbs. Neurological evaluation revealed increased tone in all limbs with extensor plantars. C.T. scan of brain performed at that time was suggestive of white matter demyelination (Fig. 1a). MRI of brain done subsequently also documented periventricular hyperintensities (T2W and FLAIR) (Fig. 1b). Analysis of cerebrospinal fluid by guarded lumbar puncture was normal. A thorough history from patient’s parents revealed that he had been suffering from episodic headache over the last 3 years and
he was diagnosed and being treated for childhood migraine by the family physician. Our search for the etiology of secondary hypertension led to the C.T. scan of abdomen and it showed a mass (5.3×6.5cm) over left adrenal gland (Fig.2). Further evaluation revealed elevated urinary epinephrine [7.78ìgm/day (Normal <11μg/day); 12.21μg/gm creatinine (Normal 1-10μg/g creatinine)] and nor-epinephrine [2120.22μg/day (Normal 12-88 μg/day); 3329.28μg/gm creatinine (Normal 15-58 μg/g creatinine)] with normal urinary dopamine [181.02μg/day; 284.25 μg/g creatinine (Normal <545 μg/g creatinine)]. Urinary metachromatic granules were absent. A diagnosis of pheochromocytoma was made. MIBG scan excluded other extra-adrenal localization of tumor. The blood pressure was controlled initially with alpha-receptor blocker (labetolol) and calcium channel blocker (Nifedipine) followed by nitroprusside infusion per-operatively. After successful surgical excision of the tumor patient became normotensive and a repeat MRI of brain after six months showed reversal of white matter abnormalities (Fig.3).

Our final diagnosis in this patient was PRES secondary to pheochromocytoma.

DISCUSSION

PRES was introduced into clinical practice in 1996 in order to describe a unique syndrome, expressed mainly during hypertensive encephalopathy.1

Hypertensive encephalopathy is characterised clinically by headache, vomiting, seizures, visual abnormalities, disturbances in cognition and alteration in the level of consciousness. Imaging studies often demonstrate edema of the white matter in the posterior parietal and occipital areas of the brain. This so-called PRES, which is the neuro-radiological correlate of hypertensive encephalopathy, is also associated with a number of conditions. There are several theories about the mechanism for PRES, but the most widely accepted hypotheses are the generation of vasogenic edema caused by failure of the mechanism that allows self regulation of the cerebral blood flow, and the production of cytotoxic edema due to ischemia.2 MRI with sequences that evaluate the diffusion of cerebral water is useful in distinguishing between the two mechanisms.

Pheochromocytoma occurs in near about 0.1% of the hypertensive population. It is usually curable if diagnosed and treated in time. In children, one fourth of tumors are bilateral and another fourth are extra adrenal.3 Solitary tumor favors the right side. However, in this child, the tumor was unilateral and was on the left side. No extra adrenal tumor was localized by MIBG scan. The initial diagnosis was cerebral malaria as signs and symptoms at that time resembled that of cerebral malaria which is very