

Posterior Reversible Encephalopathy Syndrome with Involvement of the Cervical Cord and Medulla: a Case Report

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ABSTRACT

Posterior reversible encephalopathy syndrome (PRES) is a neurotoxic state, which is associated with symmetrical subcortical areas of vasogenic oedema that are preferentially parieto-occipital, and it typically resolves within a few weeks after appropriate treatment. We hereby report a case of a female with adrenal tumour presenting with PRES, who was featured by a very rare neuroimaging manifestation, the involvement of cervical cord and medulla.

Keywords: Adrenal tumour, Clinical features, Neuroimaging manifestations, Posterior reversible encephalopathy syndrome (PRES)

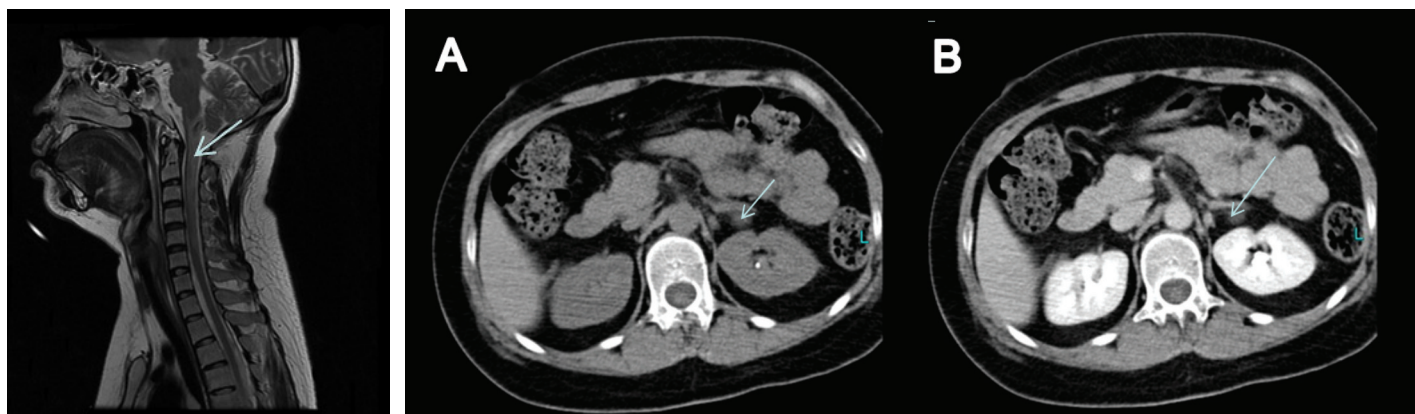
CASE REPORT

A 26-year-old women experiencing an epileptic seizure, abrupt headache, generalized body weakness, was admitted in the emergency, she was under excessive dieting followed by 15 kg increase of body weight since 3 months. On general medical examination, she was characterized by moon face, papulopustular rashes in the face and anterior of the chest, purple lines on her inner thighs, as well as high blood pressure ranging from 200/100 to 220/110 mmHg. On neurological examination, she was sober, could not move her right limb in the bed, but could lift her left limb. She had no past history of hypertension and did not take any medication before. Non-enhanced computed tomography (CT) showed no evidence of cerebral hemorrhage or subarachnoid hemorrhage (SAH). Magnetic Resonance Imaging (MRI) of brain and spinal cord was thus performed, which should hyperintense lesions involving the medulla and cervical spinal cord in T2-weighted images [Table/Fig-1]. The 24 h urinary vanillyl mandelic acid (VMA) was 11.2 μ mol/24 hrs, urinary 17-hydroxy corticosteroids was 39.4 μ mol/24 h, urinary 17-ketone corticosteroids was 88.7 μ mol/24 h, blood cortisol was 34.3 μ g/dl, blood cortisol (F-4pm) was 31.55 μ g/dl, blood adrenocorticotrophic hormone (ACTH) was 0.22pmol/l indicating ACTH-independent Cushing's syndrome. Abdomen CT of the patient showed a significantly enhancing mass in the left adrenal gland (size about 3.2cm x 2.6 cm) [Table/Fig-2A,B]. The patient was also subjected to adrenal lumpectomy, indicating the tumour of 2cm x 2cm x 3cm in size. The pathological findings showed adrenal adenoma. She was treated with levoamlodipine and prazosin for the hypertension, and 2 weeks later her muscle strength of all four limbs

recovered to normal. four weeks follow-up MRI of the spinal cord showed disappearance of previous imaging finding in T2-weighted image [Table/Fig-3]. Considering the characteristics of clinical and imaging performance, an atypical variant of PRES was the most plausible diagnosis. One year after being discharged from hospital, the patient was followed up and remained asymptomatic.

DISCUSSION

Recently, posterior reversible encephalopathy syndrome (PRES) has become increasingly recognized. Hypertensive encephalopathy, toxemia of pregnancy, cyclosporin A toxicity and uremic encephalopathy are the most common causes of PRES [1]. The mechanism of PRES is controversial. The present more popular theory suggests that severe hypertension exceeds the limits of autoregulation, leading to breakthrough brain oedema [2]. The patient whom we reported here, also suffered from severe hypertension because of adrenal tumour, which further confirmed the above theory. Another theory has also emerged after reports of PRES combined with regions of vasospasm, in which hypoperfusion is assumed to cause injury and subsequent oedema [3]. PRES is a clinic-radiologic syndrome with diverse symptoms. Clinically, headache, confusion, seizure and visual disturbance are common symptoms of PRES, yet muscle weakness of limbs, ataxia are rare [4,5]. The MRI characteristics of PRES are vasogenic oedema involving parieto-occipital lobes (cortical/subcortical white matter). The frontal and temporal lobes were common involved regions in PRES. Involvement of the basal ganglia, brain stem and deep white matter are the less common but were confidently recognized as part of the PRES process



[Table/Fig-1]: The cervical cord MRI of the case showed T2-weighted hyperintense involving the cervical cord and medulla **[Table/Fig-2]:** A: unenhanced CT of abdomen showed a mass (size about 3.2cm x 2.6 cm) in the left adrenal gland, the value of the CT was 21HU. B: enhanced CT of abdomen showed the mass was significantly enhanced, the value of the CT was 56HU



[Table/Fig-3]: 4 weeks follow-up MRI of the spinal cord showed disappearance of previous imaging finding in T2-weighted image

[6]. In recent years, focal hemorrhage, subtle subarachnoid hemorrhage was reported as uncommon imaging features of PRES [7-9]. Involvement of the cervical cord and medulla oblongata was indicated in our patient, which is a very rare neuroimaging manifestation of PRES and was reported in few cases [10-12]. Also from MRI of cervical cord, a high probability for infarction can be considered. But patients with massive brainstem infarction would have a sudden onset and more severe neurodeficit, as well as being comatose at presentation [13].

CONCLUSION

Base on the patient's clinical manifestation, MRI features, and the benign prognosis, diagnosis of PRES seems most reasonable. It

needs to be considered whether adrenal tumours or any particular etiology predisposes to cord involvement in PRES. Furthermore, In order to prevent misdiagnosis, the neurologists should recognize this atypical imaging of RPES.

ACKNOWLEDGEMENT

Xuan Hou, Jinfeng Xu contributed equally to this work.

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Date of Submission: **Aug 15, 2014**

Date of Peer Review: **Sep 21, 2014**

Date of Acceptance: **Oct 28, 2014**

Date of Publishing: **Jan 01, 2015**

FINANCIAL OR OTHER COMPETING INTERESTS: None.