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Dramatic Amelioration in Serial Magnetic Resonance Imaging in an "Isolated Brainstem" Reversible Encephalopathy Syndrome Case

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ABSTRACT

Posterior reversible encephalopathy syndrome (PRES) is characterized by transient vasogenic edema predominantly in supratentorial areas within the posterior circulation regions. Although PRES with only brainstem involvement is quite rare, accurate diagnosis is important because prompt antihypertensive therapy contributes to a favorable outcome. Herein, we report a case with isolated brainstem PRES showing dramatical improvement in an apparent diffusion coefficient (ADC) value of the lesion in magnetic resonance imaging (MRI) after clinical remission. The present case suggests the association between favorable clinical course and complete amelioration on MRI.

Key words diffusion-weighted magnetic resonance imaging; hypertensive encephalopathy; magnetic resonance imaging; posterior reversible encephalopathy syndrome

Posterior reversible encephalopathy syndrome (PRES) with only brainstem involvement is rare. As prompt antihypertensive therapy contributes to a favorable outcome, the accurate diagnosis of brainstem involved PRES is very important. Reversible hyperintensity on fluid-attenuated inversion recovery (FLAIR) imaging, and elevated apparent diffusion coefficient (ADC) value on brain magnetic resonance imaging (MRI) play crucial roles in the diagnosis of the isolated brainstem involved in PRES. Herein, we describe an isolated brainstem PRES case that showed dramatic improvement in both clinical symptoms and abnormalities on MRI after immediate antihypertensive therapy.

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Abbreviations: ADC, apparent diffusion coefficient; DWI, diffusion-weighted imaging; FLAIR, fluid-attenuated inversion recovery; MRI, magnetic resonance imaging; PRES, posterior reversible encephalopathy syndrome

PATIENT REPORT

A patient in her 70s with several months history of untreated hypertension had generalized weakness for 1 day. When she visited our hospital, she did not have symptoms of headache, nausea, fever, or convulsion. Initial examination was notable for elevated blood pressure (199/111 mmHg). In neurological examination, Glasgow Coma Scale score was 15/15, and the patient showed mild dysarthria and bradykinesia. She did not present gaze limitation, nystagmus, weakness or ataxia. Routine blood and biochemical analysis revealed mild hyponatremia (137 mmol/L), moderate hyperglycemia (265 mg/dL), and normal renal function (creatinine = 0.56 mg/dL; BUN = 17 mg/dL). Brain MRI showed mildly swollen pons, and inhomogenous hyperintensities in cerebral peduncle, pons, and pyramidal tracts in medulla oblongata, which mainly involved pontine longitudinal fasciculus and transverse pontine fibers on FLAIR imaging (Figs. 1A and B), but no abnormalities on diffusion- weighted imaging (DWI) (Figs. 1C and D). Magnetic resonance angiography was unremarkable. The patient was temporally diagnosed with hypertensive encephalopathy with chronic ischemic changes in the brainstem, and amlodipine was administered. Her blood pressure was low (130-140/60-70 mmHg), but her dysarthria and weakness improved in a few days. Two months later, brain MRI revealed that the hyperintense lesions in the brainstem were diminished compared to the initial findings on FLAIR imaging (Figs. 2A and B). Based on the above findings, a diagnosis of PRES was made. Ten months after the first MRI, the residual hyperintense lesions were completely resolved on FLAIR imaging (Fig. 2C). The ADC value of the lesional area was increased (1.03×10^{-3} mm^2/sec) on the first MRI (Fig. 2D), which gradually decreased to 0.83×10^{-3} mm^2/sec (Fig. 2E) and 0.79×10^{-3} mm^2/sec (Fig. 2F) 2 months and 10 months later, respectively. Changes of ADC value were easily recognized on the inverted images.

DISCUSSION

The isolated brainstem lesion in the present case should be differentiated from infarction, central pontine

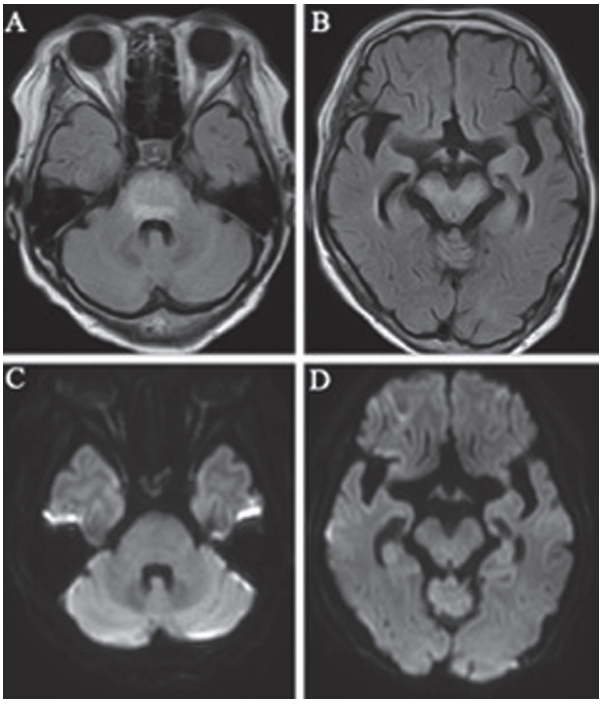

Fig. 1. (**A**) Brain MRI showing mildly swollen pons, and inhomogeneous hyperintensities that mainly involved pontine longitudinal fasciculus and transverse pontine fibers on FLAIR imaging. (**B**) Brain MRI showing inhomogeneous hyperintensities in cerebral peduncle, but no abnormalities in the supratentorial area on FLAIR imaging. (**C**) (**D**) No abnormalities are shown on DWI.

myelinolysis, inflammatory diseases, neoplasms, and PRES.[1] The patient's dysarthria and bradykinesia could have been explained by involvement of the nigrostriatal pathway in the midbrain; however, the patient did not present consciousness disturbance, impaired eye movements, motor paresis, or cerebellar ataxia, which are symptoms that are consistent with prominent lesions in pontine longitudinal fascicules and transverse pontine fibers. Clinicoradiological dissociation excluded infarction, inflammatory diseases, and neoplasms. Furthermore, the patient's hyponatremia and hyperglycemia were not severe enough to cause central pontine myelinolysis. A lack of prominent neurologic dysfunction despite the extensive brainstem abnormalities,[2] reversible hyperintensity on FLAIR imaging, and elevated ADC value on brain MRI reflecting vasogenic edema[1] were consistent with the characteristics of PRES. Thus, we diagnosed isolated brainstem PRES.

PRES is characterized by headache, confusion, visual disturbances, and seizure with transient vasogenic edema predominantly in supratentorial areas within the posterior circulation regions; furthermore, it is associated with brain-capillary leakage caused by various conditions such as hypertension, renal failure, and cytotoxic drugs.[3] Atypical lesions are occasionally observed in the brainstem following typical supratentorial lesions; however, isolated brainstem involvement occurs in only 1.5% of PRES patients.[4, 5] Although there

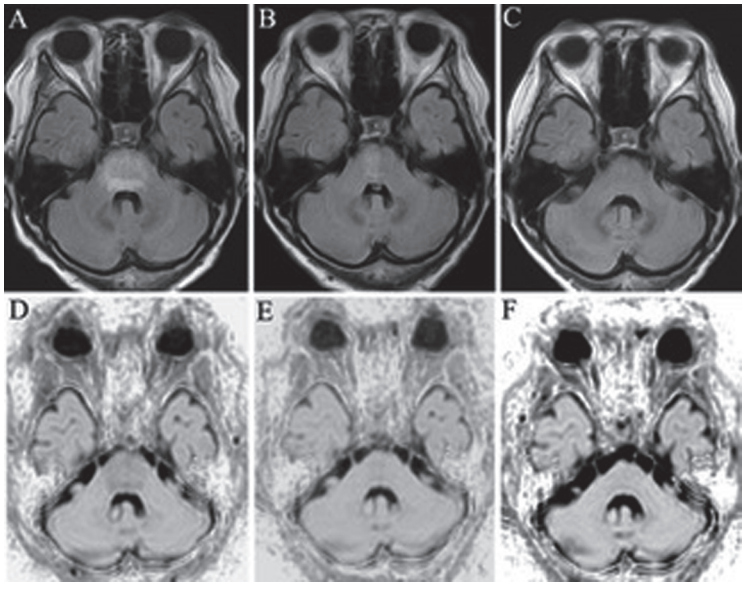

Fig. 2. (**A**) Brain MRI on the first visit, showing mildly swollen pons, and inhomogeneous hyperintensities that mainly involved pontine longitudinal fasciculus and transverse pontine fibers on FLAIR imaging. (**B**) Brain MRI 2 months later, showing that the hyperintense lesions in the brainstem had diminished on FLAIR imaging. (**C**) The residual hyperintense lesions are completely resolved on FLAIR imaging 10 months after the first MRI. (**D**) The ADC value of the lesional area on the first MRI is 1.03×10^{-3} mm^2/sec. (**E** and **F**) ADC values of the lesions on 2 months and 10 months after the first MRI are 0.83×10^{-3} mm^2/sec and 0.79×10^{-3} mm^2/sec, respectively. The changes of the ADC value can be easily recognized on the inverted images.

is no accepted explanation for isolated involvement of brainstem in PRES, previous studies have suggested that severe elevation of blood pressure is strongly related to infratentorial vasogenic edema lesions.[5, 6] One multicenter study reported that brainstem lesions showed less reversibility than typical cortical and subcortical PRES lesions,[7] and a previous study reported a case of isolated brainstem PRES which residual lesions on brain MRI observed over 3 months.[8] Furthermore, another previous study reported potential small vessel vulnerability caused by chronic hypertension may be related to the prolongation of vasogenic edema in the lesions.[7] Thus, potential vessel vulnerability due to hypertension may be related to the development of isolated brainstem PRES and prolonged elevation of ADC value reflecting the vasogenic edema of the lesional area in the present case.

The present case suggests that when extensive abnormalities in the brainstem on brain MRI without prominent neurologic dysfunction are observed in hypertensive patients, PRES should be considered as a differential diagnosis, as prompt antihypertensive therapy contributes to a favorable outcome.[6] Improvement in the ADC value of the residual lesion was observed after 10 months after clinical remission in this patient. Thus, a favorable clinical course may be associated to complete amelioration on MRI in the long term.

The authors declare no conflict of interest.

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