Abstract

We report a case of hypertensive brainstem encephalopathy (HBE) with unusual magnetic resonance imaging (MRI) findings. A 67-year-old woman presented with high blood pressure and stupor as the only symptoms. MRI revealed lesions localized in the area from the upper medulla oblongata to the lower pons with high fluid-attenuated inversion recovery (FLAIR) and T2-weighted signal intensity, but these were not seen in the whole brainstem and there were no accompanying occipital lobe changes. To our knowledge, no similar case has been reported. The lesions and symptoms dramatically improved after normalization of blood pressure. Severe hypertension that exceeded the range of autoregulation may have resulted in segmental vasodilatation and the increased vascular permeability may have lead to vasogenic edema in the localized areas of the brainstem.

Key words: hypertensive brainstem encephalopathy, magnetic resonance imaging

Introduction

Hypertensive encephalopathy (HTE) is included in reversible posterior leukoencephalopathy syndrome (RPLS) (1), and is characterized by acute onset, severe arterial hypertension, headache, confusion, convulsion, and symptoms of posterior circulation. The most common abnormality on magnetic resonance imaging (MRI) in HTE is found bilaterally in the white matter of the posterior cerebral hemisphere, which is presumed to be brain edema (2–9). Involvement of the brainstem in addition to supratentorial lesions has been rarely reported, and is termed hypertensive brainstem encephalopathy (HBE) with abnormal MRI findings usually in the whole brainstem (6–10). Here, we report an unusual case of HBE with MRI findings localized in the brainstem without accompanying occipital lobe changes.

Case Report

A 67-year-old woman was suddenly found unresponsive by her family. She had no complaint of headache or nausea. Her past medical history was significant for poorly controlled diabetes mellitus (DM). She had no history of hypertension and did not take a depressor medication. She was admitted to our hospital. Her blood pressure at admission was 204/106 mmHg, but her general physical examination showed no other abnormalities. She was stuporous, did not complain of headache or nausea, and did not develop seizure. She was not able to follow simple commands, but quickly responded to pain. Her cranial nerves, including visual function, conjugated deviation, nystagmus, and facial nerve palsy, were intact. She had no weakness. Her deep tendon reflexes were normal without pathological reflexes. Fundoscopy showed hypertensive retinopathy. Brain MRI (the GE Signa 1.0-T system was used) showed no abnormal areas on diffusion-weighted image (DWI) (Fig. 1A) and high intensity areas in the marginal areas (blood vessels thickly distributed areas) of the brainstem, especially the medulla oblongata in fluid-attenuated inversion recovery (FLAIR, 8002/136/1 [TR/TE/number of excitations]) (Figs. 1B–E) and T2-weighted image (T2WI, 4000/106/1) (Figs. 1G–J), without any accompanying occipital lobe changes (Fig. 1F). Laboratory tests showed a blood glucose level of 324 mg/dl and HbA1c 7.3%. Blood urea nitrogen (40.0 mg/dl) and creatine concentrations (2.97 mg/dl) were elevated, and the electrolytes were normal. At lumbar puncture, the cerebrospinal fluid (CSF) pressure was 130 mmHg. Cell count was
7/mm$^3$ (neutrophil 6, lymphocyte 1). The CSF protein concentration of 81 mg/dl and glucose level of 158 mg/dl (blood glucose 286 mg/dl) were elevated. We immediately started intravenous infusion of nicardipine (a calcium channel blocker) which lowered her systemic pressure to 150–160/80–90 mmHg, followed by amlodipine (an oral calcium channel blocker) and doxazosin (angiotensin-converting enzyme inhibitor) on day 1. On day 3 after the onset, synchronized with improvement of hypertension, she became alert. On day 9, lumbar puncture showed a normal cell count (0/mm$^3$) and the CSF protein level (35 mg/dl). But the CSF IgG index was slightly high at 0.77. On day 12, single
photon emission computed tomography (SPECT) using $^{123}$I-IMP showed hyperperfusion in the lower brainstem (Figs. 2A, B, arrows). On day 15, abnormal MRI findings in the brainstem had improved (Figs. 1K, L), but renal failure due to DM did not improve.

**Discussion**

The current case presented with high blood pressure levels and stupor as the only symptoms on admission. She promptly responded to antihypertensive therapy and her symptoms were resolved. Differential diagnosis included brainstem infarction, brainstem tumor, encephalitis, central pontine myelinolysis (CPM), and acute disseminated encephalomyelitis (ADEM). Brainstem infarction was ruled out by the lack of major brainstem signs, and the rapid clinical recovery. CPM should show a lesion in the central part of the brainstem, but the present case showed a marginal lesion and had no electrolyte abnormalities. Infectious brainstem encephalitis was unlikely the cause based on the normal CSF findings and rapid recovery without specific treatment. The rapidity of the clinical evolution helped differentiate the tumor. ADEM was the most difficult condition to exclude, but HBE is symmetric and unaccompanied by inflammatory CSF changes. HTE usually involves the posterior parieto-occipital structures, and HTE with involvement in only the brainstem regions, as in the present case, is rare. The characteristics of HBE are summarized as follows: severe arterial hypertension, symmetrical MRI lesions with poor focal signs, and no lesions in DWI, great recovery of clinical symptoms and MRI findings after successful lowering of blood pressure (2, 6–10). Thus, present patient was diagnosed as HBE.

MRI of HBE usually shows lesions in the whole brainstem, but the present case had only marginal involvement. To our knowledge, a similar case has not been reported. The mechanisms of HTE are postulated as follows: a rapid blood pressure rise over autoregulation, arteriole extension resembling sausage-string (Fig. 2C), endothelial dysfunction with increasing permeability of the blood brain barrier (BBB), which causes vasogenic edema (11). Thus, the essential mechanism of HTE is hyperperfusion and vasogenic edema due to severe hypertension. Reversible lesions in FLAIR and T2WI that are not generally seen in DWI support this hypothesis. Hyperperfusion of the brainstem in SPECT (Figs. 2A, B) and the increase of the IgG index in the present case support the diagnosis of HTE (12). Because the bloodstream enters the brainstem from marginal areas to the central areas
and also because white matter in the peripheral areas of the brainstem more easily collects extracellular water by vasogenic edema than gray matter, the marginal area is more hyperperfusive and vulnerable to formation of vasogenic edema than the central area (Fig. 2D).

The reason the present case did not show these MRI findings may have been a result of hypertension that was not severe. We hypothesize that mild hypertension caused minimum hyperperfusion in the blood vessels which had vulnerable autoregulation due to a disturbance of the sympathetic nerve caused by DM. In cases similar to ours, the marginal areas of the brainstem, which have vulnerable autoregulation compared to the central areas, may display high intensity areas on MRI.

**References**


