Hypertensive Brainstem Encephalopathy

Yuzuru YASUDA, Ichiro AKIGUCHI, Tadahiko IMAI*,
Masanobu SONOBE** and Makoto KAGE*

Abstract

A 45-year-old man developed severe arterial hypertension associated with unusual hyperintensity in the brainstem, around the right internal capsule and in the deep white matter around the bilateral anterior horn of the lateral ventricle on T2-weighted and fluid-attenuated inversion-recovery images. The characteristic clinical findings were mild left hemiparesis and altered mental status which corresponded to the lesions of MR imagings. The lesions improved gradually with improvements in hypertension, which suggested that edema could be the principal cause of the unusual hyperintensity on MR images.

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Key words: hypertensive encephalopathy, brainstem, MR imaging, hypertension, edema

Introduction

Hypertensive encephalopathy which is caused by a dramatic rise in blood pressure, is characterized by altered mental status, seizures, headache and visual disturbances (1, 2). Diffuse or focal hyperintensity on T2-weighted images in posterior supratentorial white matter (3-5) have been reported in hypertensive encephalopathy and minor cerebellar and brainstem involvement is not unusual (3, 5). However, lesions that are predominantly in the brainstem are rare as causes of hypertensive encephalopathy (6-11). We encountered a patient with hypertensive encephalopathy whose main lesion existed in the brainstem; here, we discuss the pathogenesis of hypertensive brainstem encephalopathy (HBE).

Case Report

A 45-year-old man presented with dizziness and blurred vision of 2 days duration. His neurologic history was unremarkable. He had a 7-year history of hypertension which was untreated, but no history of palpitations or sweating. He had no family history of hypertension. Neurologically he was alert and neither disorientation or paresis were observed. A general examination showed a blood pressure of 250/160 mmHg, a pulse rate 100 beats/min and a body temperature of 36.5°C. A chest roentgenogram showed cardiomegaly (CTR=57%) and an electrocardiogram showed left ventricular hypertrophy. An ophthalmologic examination showed bilateral retinal hemorrhage with severe hypertensive retinopathy. Laboratory investigation gave the following results: white blood cell count 16.1x10³/ml, red blood cell count 5.32x10¹²/ml, hemoglobin 16.6 g/dl, hematocrit 49.8%. C-reactive protein was 0.2 mg/dl. Urinalysis revealed slight proteinuria, but occult blood was negative. Serum electrolyte levels were Na 138 mEq/l, K 3.7 mEq/l, and Cl 99 mEq/l. Liver function was normal, but renal function was disturbed; blood urea nitrogen 44.5 mg/dl, creatinine 2.53 mg/dl, creatinine clearance 33.3 ml/min, uric acid 8.9 mg/dl, renin was 46.5 ng/ml • h (normal 0.3-2.9), and aldosterone was 203 pg/ml (normal 36-240). Serum catecholamine levels were as follows; epinephrine 85 pg/ml (normal <100) and norepinephrine 440 pg/ml (normal 100-450). Urinary 17-OHCS was 11.8 mg/day (normal 3.4-12.0), and 17-KS was 10.4 mg/day (normal 4.6-18.0). Thyroid function was normal. Abdominal echography showed no stenosis of the renal artery and CT scan of the abdomen showed no evidence of pheochromocytoma. During the first 3 days of admission, he was treated with continuous infusion of nicardipine hydrochloride and his blood pressure fell to between 200/110 and 180/100 mmHg, and his pulse rate became 80 beats/min. However, on the third day of admission, he became drowsy and was able to follow simple commands and was transferred to neurologic ward.

From the Center of Neurological and Cerebrovascular Diseases, Takeda Hospital, Kyoto, *Department of Neurology, Otsu Red Cross Hospital, Otsu and **Department of Neurology, Kyoto City Hospital, Kyoto

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Reprint requests should be addressed to Dr. Yuzuru Yasuda, the Center of Neurological and Cerebrovascular Diseases, Takeda Hospital, 841-5 Nishinotoin Shiokoji Dori, Shimogyo-ku, Kyoto 600-8558

Internal Medicine Vol. 42, No. 11 (November 2003)
Figure 1. MR images in the acute stage. A: T1-weighted (500/18) image showed the swelling and low signal in the brainstem (arrow). B: T2-weighted (2,300/90) and C: FLAIR (6,856/110; inversion time, 1,700) image showed hyperintense signals in the brainstem (arrow). D: FLAIR (6,856/110; inversion time, 1,700) image showed hyperintense signals around the right internal capsule (arrow) and in the deep white matter around the bilateral anterior horn of the lateral ventricle.

The patient was lethargic and confused, but could respond to simple questions. He showed mild left hemiparesis and his deep tendon reflex was exaggerated on the left side without a pathologic reflex. An electroencephalogram showed no spikes or laterality and auditory evoked responses showed no abnormalities. A CT scan showed low density and moderate swelling in the brainstem and a low density area was also seen around the right internal capsule. T2-weighted and fluid-attenuated inversion-recovery (FLAIR) images showed hyperintense signals in the medulla, pons and midbrain (Fig. 1B, C), around the right internal capsule (Fig. 1D), and in the deep white matter around the bilateral anterior horn of the lateral ventricle. A T1-weighted image confirmed the swelling and low signal in the medulla, pons and midbrain (Fig. 1A) and around the right internal capsule. The cerebellum was not involved. A diffusion-weighted image (DWI) was not available. MR angiography showed no variation or vasospasm of the posterior cerebral artery. Lumbar puncture was not performed.

The patient was treated with amlodipine besilate (10 mg/day) and amosulalol hydrochloride (40 mg/day). His blood pressure dropped to between 210/120 and 190/100 mmHg, and he became alert at the eighth day of admission (Fig. 2). His left hemiparesis disappeared, but deep tendon reflexes on the left side remained exaggerated. T2-weighted and FLAIR images at the 12th day of admission showed that hyperintense signals remained in the brainstem, but the hyperintense signal around the right internal capsule and in the deep white matter around the bilateral anterior horn of the lateral ventricle showed improvement. The low signal in the brainstem and around the right internal capsule on the T1-weighted image also showed improvement. At the 16th day of admission, as his blood pressure was still high, methyldopa was added. At the 19th day of admission, his blood pressure was reduced to between 170/100 and 150/90 mmHg, and the hyperintense signals on T2-weighted and FLAIR images in the brainstem, around the right internal capsule, and in the deep white matter around the bilateral anterior horn of the lateral ventricle decreased markedly (Fig. 3B, C, D). The exaggerated deep tendon reflex on the left side returned to normal, and renal function showed improvement (blood urea nitrogen 24.5 mg/dl, creatinine 2.11 mg/dl). The patient’s blurred vision gradually improved with improvements in the retinal hemorrhage.

Discussion

The present patient presented with dizziness and blurred vision followed by lethargy and left mild hemiparesis. Blurred vision was due to retinal hemorrhage, because MR angiography showed no vasospasm of the posterior cerebral artery. Secondary hypertension was ruled out by laboratory tests and image studies. Blood pressure did not normalize.
Hypertensive Brainstem Encephalopathy

Figure 3. MR images in the recovery stage. A: T1-weighted (500/18), B: T2-weighted (2,300/90), C: and D: FLAIR (6,856/110; inversion time, 1,700) images. The lesion in the brainstem, around the right internal capsule and in the deep white matter around the bilateral anterior horn of the lateral ventricle showed marked improvement.

quickly due to renal failure and methyldopa added to amlodipine besilate plus amosulalol hydrochloride therapy was effective. The main lesion was clearly demonstrated in the brainstem by T2-weighted and FLAIR images, but clear-cut brainstem signs were absent. The left mild hemiparesis was caused by a lesion around the right internal capsule. As the main lesion of hypertensive encephalopathy typically exists in the posterior supratentorial white matter (3-5), our patient differed from the typical cases. To date, six reports of hypertensive encephalopathy with predominant brainstem involvement as seen in our case, have been presented in the literature (6-11). As this type of hypertensive encephalopathy is rare, the terms used to describe it vary. The most appropriate name seems to be hypertensive brainstem encephalopathy (HBE) (8).

Two reported cases with HBE showed brainstem swelling, with compression of the adjacent cisterns and fourth ventricle and both resulted in obstructive hydrocephalus (7, 9). One patient required a shunt operation (7) and another case recovered with aggressive antihypertension drugs (9). Six cases with HBE recovered with antihypertensive drugs only (6, 8, 10, 11) and one with the surgical removal of a pheochromocytoma (10). In cases with predominant brainstem involvement, HBE should be differentiated from central pontine myelinolysis, acute disseminated encephalomyelopathy, brainstem infarction and brainstem glioma. The former three could be differentiated on the basis of laboratory examinations and the clinical course. It has been reported that MR findings with the alternating bright and low signal intensity in HBE suggest edema or fluid between the transverse pontine bundle, and are proposed to be used to distinguish HBE from glioma (10). Such a finding was also seen in the acute stage of our case on T2-weighted images (Fig. 1).

The mechanism of hypertensive encephalopathy remains unclear. However, two theories have been proposed. One is that hypertensive encephalopathy results from autoregulatory vasoconstriction (overregulation) in the brain in response to severe hypertension and could cause hypoperfusion, resulting in ischemia and cytotoxic edema (4, 12). An alternative theory is a vasodilatatory mechanism which results from a breakthrough of autoregulation, with passive overdistension of cerebral arterioles. This results in extravasation of protein and fluid, producing focal vasogenic edema in the involved peripheral vasculature (13-15). If vasospasm is an important mechanism, ischemia with resultant infarction would occur in some patients. However, this is infrequent in both clinical (1, 5, 12, 13) and experimental cases (15). Focal enhancement on MR with gadopentetate dimeglumine in the acute stage suggests that hypertensive encephalopathy could be caused by a breakthrough of autoregulation with focal vasogenic edema (5).

DWI in a patient with HBE (11) showed findings of both vasogenic and cytotoxic edema in the brainstem; T2-weighted and FLAIR images and DWI showed high signal intensities in the pons, midbrain and bilateral middle cerebellar peduncle, and apparent diffusion coefficient (ADC) values were low in the right pons and midbrain but high in the remaining brainstem. It is reported that hyperintense T2 and FLAIR changes together with hyperintense DWI and normal to increased ADC indicate vasogenic edema, and together with hyperintense DWI and reduced ADC, lead to cytotoxic edema (16). Follow-up MR in this patient showed some lacunae in the pons which indicated that only part of the cytotoxic edema might progress to infarction.

It is well known that posterior circulation is susceptible to the lesions of hypertensive encephalopathy (13, 17). This observation could be explained by regional heterogeneity in the sympathetic vascular innervation which protects the brain from marked increases in blood pressure (18). Moreover, the internal carotid system has a more extensive sympathetic nerve supply than the vertebrobasilar system (19). Therefore, the posterior circulation is vulnerable to high blood pressure.

To date, six cases of HBE have been reported (6-11). Why HBE occurs rarely compared to involvement of the posterior supratentorial deep white matter remains unclear. It has been reported that the infratentorial structures could also be affected when severe hypertension continues for a long

Internal Medicine Vol. 42, No. 11 (November 2003)
time (20) As this patient had severe hypertension and his blood pressure did not return to normal rapidly, the brainstem may have become involved. Hypertension should be normalized as soon as possible to prevent the occurrence of HBE.

References