

Case Report

Hypertensive Encephalopathy with Reversible Brainstem Edema

Sungjoon Lee, M.D.,¹ Byung-Kyu Cho, M.D., Ph.D.,² Hoon Kim, M.D.³

Department of Neurosurgery,¹ Seoul National University College of Medicine, Seoul, Korea

Department of Neurosurgery,² The Armed Forces Capital Hospital, Seongnam, Korea

Department of Neurosurgery,³ The Catholic University of Korea, Bucheon St. Mary's Hospital, Bucheon, Korea

Presented here is a 36-year-old male with arterial hypertension who developed brainstem edema and intracranial hemorrhage. Magnetic resonance scan revealed diffuse brainstem hyperintensity in T2-weighted and fluid-attenuated inversion-recovery images, with an increase in apparent diffusion coefficient values. After a reduction in blood pressure, rapid resolution of the brainstem edema was observed on follow-up. The patient's condition was thus interpreted as hypertensive brainstem encephalopathy. While many consider this a vasogenic phenomenon, induced by sudden, severe hypertension, the precise mechanism remains unclear. Prompt recognition and aggressive antihypertensive treatment in such patients are essential to prevent permanent or life-threatening neurologic injury.

Key Words : Brain stem · Edema · Encephalopathy · Hypertension.

INTRODUCTION

Hypertensive encephalopathy (HE) is heralded by headache, visual disturbance, seizures, and altered mental status, all associated with severe hypertension⁶. Without prompt lowering of blood pressure (BP), neurologic sequelae may ensue. The most common feature of HE by magnetic resonance imaging (MRI) is hyperintensity of subcortical parietal and occipital white matter on T2-weighted views, suggestive of vasogenic edema. Because such changes usually disappear upon BP stabilization, many authors refer to this state as reversible posterior leukoencephalopathy syndrome or posterior reversible encephalopathy syndrome (PRES)^{3,6,8,9}. Involvement of the brainstem and cerebellum in PRES has rarely been described. In this report, however, we present a case of PRES where the brainstem alone is affected, accompanied by intracranial hemorrhage.

CASE REPORT

A 36-year-old male with idiopathic hypertension (duration >6 yrs) was admitted to our hospital for abrupt-onset headache,

right facial palsy, dysarthria, and right hemiparesis that had developed three hours earlier. Two weeks prior to admission, he stopped his antihypertensive medication. BP recorded at the time of presentation was 230/160 mm Hg.

Laboratory studies were within normal range and ophthalmologic evaluation was unremarkable. Specifically, no papilledema was evident. Routine chest X-ray and echocardiogram suggested severe concentric left ventricular hypertrophy, later confirmed by transthoracic echocardiography. Emergency computed tomographic (CT) scan of the brain revealed small amount (7 cc) of intracranial hemorrhage involving the left basal ganglia, and there was relative hypodensity of the brainstem (Fig. 1). CT angiography demonstrated no cerebral vascular abnormalities, including vertebrobasilar circulation. Brain MRI showed diffuse high-signal intensities on T2-weighted images and fluid-attenuated inversion recovery images without gadolinium enhancement in the pons and midbrain. No high signal intensities in diffusion-weighted images (DWI) and high signal intensities in apparent diffusion coefficient (ADC) images, implying that the changes were not due to cerebral infarction (Fig. 2A-C). There were also no abnormal findings other than the small amount of

• Received : March 23, 2013 • Revised : May 22, 2013 • Accepted : August 5, 2013

• Address for reprints : Hoon Kim, M.D.

Department of Neurosurgery, The Catholic University of Korea, Bucheon St. Mary's Hospital, 327 Sosa-ro, Wonmi-gu, Bucheon 420-717, Korea
Tel : +82-32-340-2259, Fax : +82-32-340-7391, E-mail : armada1997@naver.com

• This is an Open Access article distributed under the terms of the Creative Commons Attribution Non-Commercial License (<http://creativecommons.org/licenses/by-nc/3.0>) which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

hemorrhage in basal ganglia, which was confirmed already in CT (Fig. 2D).

Based on these findings, a diagnosis of acute hypertensive encephalopathy with intracranial hemorrhage was made. We immediately initiated intravenous antihypertensive therapy. Ten days later, systolic and diastolic pressures were successfully stabilized. Follow-up MRI (including DWI) on Day 16 of hospitalization revealed complete resolution of prior imaging aberrations involving the brainstem (Fig. 2E, F).

DISCUSSION

As a distinct clinical entity, PRES is characterized by progressive headache, visual changes, altered mental status, and seizures. Most commonly PRES is secondary event, triggered by paroxysmal severe hypertension of various etiologies. The spectrum of predisposing factors/conditions includes renal disease, immunosuppressive and cytotoxic drugs, collagen vascular disorders

(such as systemic lupus erythematosus), eclampsia, and hematologic conditions (typically, thrombotic thrombocytopenic purpura and hemolytic-uremic syndrome)⁵. However, regardless of the underlying cause, the pathogenesis of the primary abnormality, cerebral vasogenic edema, is still under debate. If the main cause is corrected, PRES can be reversed. However, if patients with PRES present with coma and/or status epilepticus, permanent neurological impairment or death may occur in some cases⁸. Generally, subcortical white matter of the parietal and occipital lobes is affected, while involvement of brainstem is rare. In our patient, MRI changes were confined to brainstem with cerebral sparing.

Reversible brainstem hypertensive encephalopathy (RBHE) was first recognized by Chang and Keane² in 1999. When the brainstem alone is affected, most patients do not exhibit focal signs or symptoms. Intracerebral hemorrhage of the left basal ganglia, rather than the brainstem manifestations, was held responsible for the facial palsy, dysarthria and right-sided hemiparesis displayed in this instance. Because the neurologic effects of RBHE are reversible with prompt treatment, this syndrome must be differentiated from brainstem infarction, pontine glioma, central pontine myelinolysis, and infectious encephalitis. Severe hypertension, so-called “clinical-radiologic dissociation” (signaling brainstem involvement), and rapid resolution of MRI changes after antihypertensive treatment are pathognomonic of RBHE¹⁰.

The suggested etiology of RBHE is a failure of cerebrovascular autoregulation in the face of high BP, resulting in cerebral hyperperfusion, disruption of the blood-brain barrier (BBB), and vasogenic edema¹. While physiologic hypertension usually activates sympathetic pathways to maintain the BBB via cerebral vasoconstriction, experimentally induced hypertension in animal models has shown that transient reflex sympathetic hypertension can override the BBB. At breakthrough, constricted cerebral arterioles dilate under hypertensive force, allowing parenchymal extravasation of fluid, macromolecules, and red blood cells⁷. The posterior circulatory network, in particular, is less endowed with sympathetic innervation than anterior vessels, and thus is less capable of a protective vasoconstrictive response to a sudden increase in arterial BP. A disruption in BBB therefore results. In RBHE, this failure of autoregulation is marked by vasogenic (as opposed to cytotoxic) edema, confirmed by imaging studies. Consistent with other reports of RBHE, demonstrable hyperintensity on T2-weighted images and normal DWI with increased ADC values were indicative of vasogenic edema in our patient.

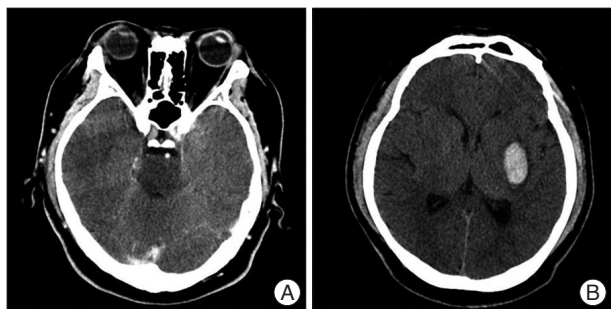


Fig. 1. Axial CT image of brain showing intracranial hemorrhage of left basal ganglia with brainstem hypodensity (A and B).

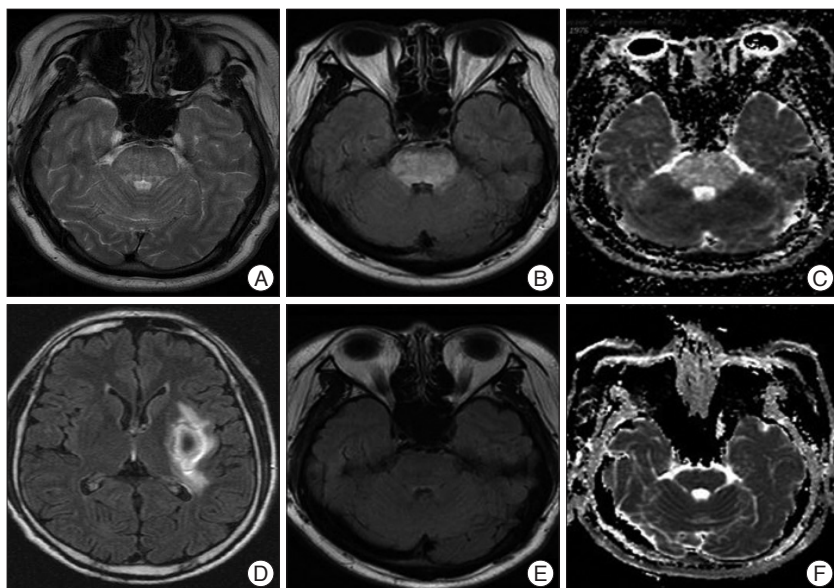


Fig. 2. Axial MRI T2 and fluid-attenuated inversion recovery image showing increased signal intensity of pons and midbrain (A and B) : increase in apparent diffusion coefficient values for same area (C) and there are no abnormal findings in subcortical white matter of the parietal and occipital lobes other than the small amount of basal ganglia (D). MRI and diffusion-weighted images after blood pressure stabilization; note resolution of prior brainstem abnormalities (E and F).

What has yet to be explained is the predilection for brainstem (versus cerebral) involvement with RBHE. Because the parietal and occipital lobes, as well as the brainstem, are within distribution of the vertebrobasilar artery and its branches, two tentative explanations have emerged⁴⁾. By allowing fluid to accumulate, the brainstem may simply absorb much of the hypertensive “tidal wave,” dissipating pressure before other parts of the brain are reached. In essence, the brainstem serves as a buffer to

protecting cerebrum and the distal parieto-occipital region in the vertebrobasilar system, more distal in blood supply. Another explanation is that the parietal and occipital lobes may be resistant to relatively rich sympathetic innervation via the posterior communicating artery (PCoA). A well-developed PCoA, also known as fetal-type PCoA, is thought to possess the same degree of sympathetic innervation as the anterior circulation. In this manner, individuals with a fetal-type PCoA may be protected from paroxysmal malignant hypertension.

Our patient, however, did not have a fetal-type PCoA. Consequently, it is clear that at least one of these theories is flawed and that more effort is needed to clarify the pathophysiologic mechanisms of RBHE.

CONCLUSION

RBHE may occur as a consequence of paroxysmal malignant hypertension. Thorough neurologic examination, in conjunction with MRI with DWI, is essential for the correct diagnosis. Clinical-radiologic dissociation and rapid improvement of MRI findings after antihypertensive treatment are features pathognomonic of RBHE. With prompt and proper management, this condition may be completely reversed, without any neurologic

deficit. However, physicians must cognizant of the potential for fatality and approach it as a medical emergency.

References

1. Bartynski WS : Posterior reversible encephalopathy syndrome, part 2 : controversies surrounding pathophysiology of vasogenic edema. *AJNR Am J Neuroradiol* 29 : 1043-1049, 2008
2. Chang GY, Keane JR : Hypertensive brainstem encephalopathy : three cases presenting with severe brainstem edema. *Neurology* 53 : 652-654, 1999
3. Chen TY, Lee HJ, Wu TC, Tsui YK : MR imaging findings of medulla oblongata involvement in posterior reversible encephalopathy syndrome secondary to hypertension. *AJNR Am J Neuroradiol* 30 : 755-757, 2009
4. Doi Y, Kimura F, Fujiyama T, Fujimura C, Nishina T, Sato T, et al. : Hypertensive brainstem encephalopathy without parieto-occipital lesion--two case reports. *Neurol Med Chir (Tokyo)* 46 : 75-79, 2006
5. Garg RK : Posterior leukoencephalopathy syndrome. *Postgrad Med J* 77 : 24-28, 2001
6. Hinchey J, Chaves C, Appignani B, Breen J, Pao L, Wang A, et al. : A reversible posterior leukoencephalopathy syndrome. *N Engl J Med* 334 : 494-500, 1996
7. Ijima T, Kubota Y, Kuroiwa T, Sankawa H : Blood-brain barrier opening following transient reflex sympathetic hypertension. *Acta Neurochir Suppl (Wien)* 60 : 142-144, 1994
8. Legriel S, Pico F, Azoulay E : Understanding Posterior Reversible Encephalopathy Syndrome. *Ann Upd Intens Care Emerg Med* 1 : 631-653, 2011
9. Medici E, Arabi S, Sebastianelli M, Pirani O, Giaccaglioni E : A case of hypertensive encephalopathy with prominent brainstem presentation. *Neurol Sci* 30 : 133-135, 2009
10. Shintani S, Hino T, Ishihara S, Mizutani S, Shiigai T : Reversible brainstem hypertensive encephalopathy (RBHE) : clinicoradiologic dissociation. *Clin Neurol Neurosurg* 110 : 1047-1053, 2008