

Posterior Reversible Encephalopathy Syndrome Mimicking Brainstem Infarction: A Dilemma

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Abstract-

Purpose: We had recently reported one case at American Journal of Emergency Medicine about central-variant posterior reversible encephalopathy syndrome (PRES) in an 84-year-old woman with an initial misdiagnosis as central pontine myelinolysis (CPM). Here, we introduce another case of central-variant PRES in a 49-year-old man mimicking as acute brainstem infarction in the cranial computed tomography (CT) findings.

Case Report: A 49-year-old man was admitted to the emergency department with a 5-day history of vertigo, cognitive decline, and difficulty in walking. Neurologic examination revealed drowsiness with a Glasgow Coma Scale score of 12 (eye opening: 3, best verbal response: 4, and best motor response: 5), slow movement in pursuit and saccades, and gait instability with a Medical Research Council scale of grade 4-5. Non-contrast cranial CT showed hypodense lesions in the pons, and antiplatelet agent was initiated for presumed pons infarction. However, the brain magnetic resonance imaging (MRI) demonstrated vasogenic edema in the corresponding area, consistent with the diagnosis of central-variant PRES.

Conclusion: This case report raises the awareness that when hypodense brainstem lesions on brain CT in patients with progressive neurological dysfunction, the rare condition of central-variant PRES should be considered in the differential diagnosis to avoid inadequate management. Cranial magnetic resonance imaging (MRI) may help in diagnosis and dealing with of these patients with similar radiological and clinical abnormalities.

Key Words: posterior reversible encephalopathy syndrome, central-variant posterior reversible encephalopathy syndrome, hypertensive encephalopathy, pontine infarction, central pontine myelinolysis.

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INTRODUCTION

Central-variant posterior reversible encephalopathy

syndrome (PRES) is an uncommon form of PRES, which characterized by failure of autoregulation of the cerebral blood flow contributing to vasogenic edema of

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brainstem, is a neurological emergency. Central variant-PRES may have overlapping clinical presentations with other etiologies affecting the brainstem such as central pontine myelinolysis (CPM) or pontine infarction. However, misunderstanding by the physicians results in an inadequate treatment and may lead to irreversible neurologic deficits or even fatality. In our clinical experience, we encountered only 2 cases of central variant-PRES in the past 10 years. We had recently reported one case at American Journal of Emergency Medicine about central-variant PRES in an 84-year-old woman with an initial misdiagnosis as CPM. Here, we introduce another case of central variant-PRES in a 49-year-old man mimicking as acute brainstem infarction in the cranial computed tomography (CT) findings.

CASE REPORT

A 49-year-old man was admitted to the emergency

department with a 5-day history of vertigo, cognitive decline, and difficulty in walking. He had medical history of chronic renal insufficiency without medications. He denied recent head trauma or drug abuse.

On admission, his vital signs were temperature of 36.2°C, pulse rate of 99 per minute, respiratory rate of 20 per minute, and blood pressure of 202/138 mmHg. His conjunctivae were not icteric, breathing sounds was clear, and cardiac sounds were regular without a murmur acoustically. Neurologic examination revealed drowsiness with a Glasgow Coma Scale score of 12 (eye opening: 3, best verbal response: 4, and best motor response: 5), slow movement in pursuit and saccades without fundoscopic papilledema, and gait instability with a Medical Research Council scale of grade 4-5. No neck stiffness was detected. Routine blood and biochemical analysis were unremarkable except for an elevated serum creatinine level at 3.3 mg/dL (reference value: 0.7-1.2 mg/dL). Non-contrast computed tomography (CT) of the brain showed

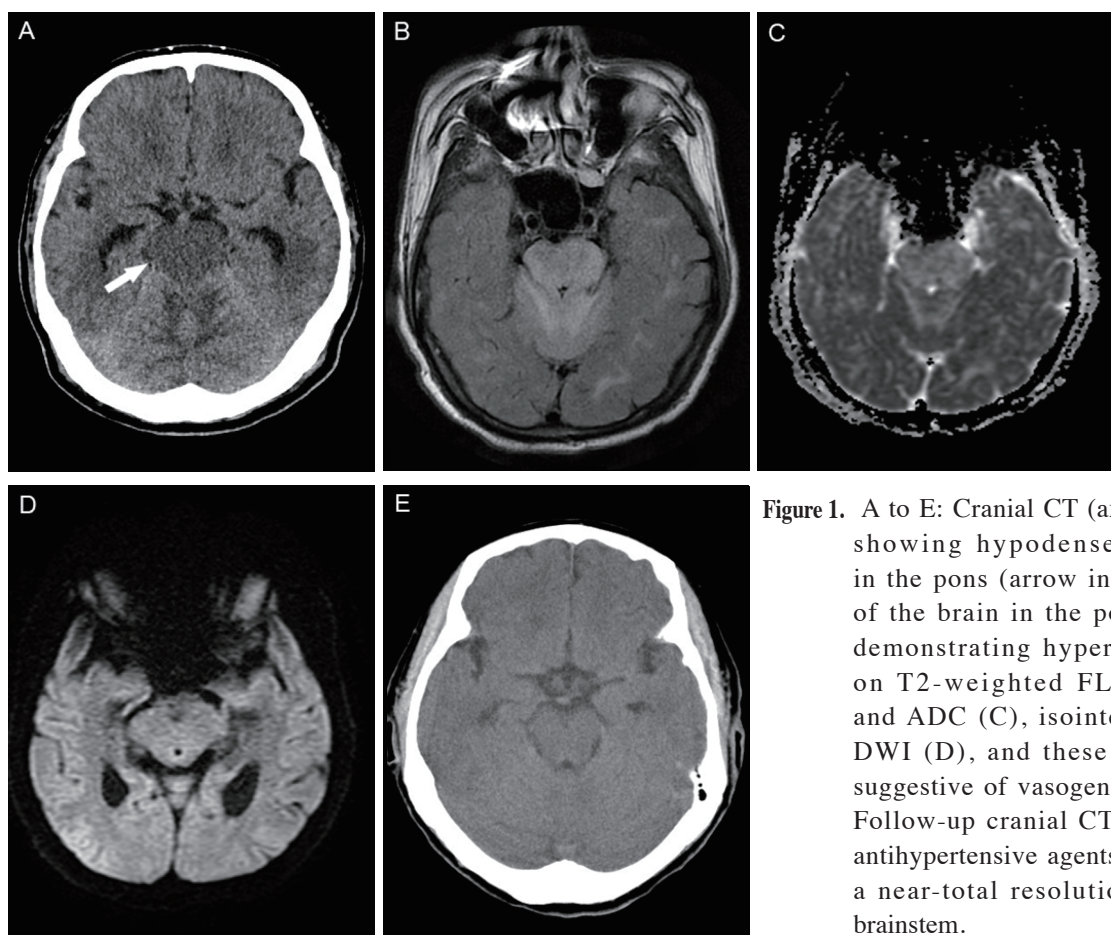


Figure 1. A to E: Cranial CT (axial view) showing hypodense lesions in the pons (arrow in A), MRI of the brain in the pons areas demonstrating hyperintensity on T2-weighted FLAIR (B) and ADC (C), isointensity on DWI (D), and these findings suggestive of vasogenic edema. Follow-up cranial CT (E) post antihypertensive agents depicted a near-total resolution of the brainstem.

hypodense lesions in the pons without obvious mass effect (Figure A). Immediate antiplatelet therapy for presumed pons infarction with oral aspirin 300mg was initiated.

To identify the possible mechanism, additional magnetic resonance imaging (MRI) was ordered, which demonstrated high signal on T2-weighted fluid attenuation inversion recovery image (FLAIR) (Figure B) and apparent diffusion coefficient (ADC) (Figure C) without restricted water diffusion on diffusion-weighted image (DWI) (Figure D) in the corresponding areas, findings suggestive of a process of vasogenic edema and exclusion of cytotoxic edema such as acute ischemic infarcts and CPM. The patient regained his consciousness dramatically 12 hours after administering antihypertensive agents with continuous intravenous nicardipine (1 mg/kg/day). On the next day, he was discharged in stable condition.

From the aforementioned clinical evidences, we confirmed the diagnosis of central-variant PRES. 10 days later, a follow-up cranial CT obtained in outpatient department showed a near-total resolution of the brainstem (Figure E). At a 2-month follow-up, he had no clinical relapses and appeared to have made a full recovery.

DISCUSSION

PRES, also known as hypertensive encephalopathy, results from uncontrolled malignant hypertension progressing to failure of autoregulation of the cerebral blood flow leading to cerebral hyperperfusion, by which can progress to breakdown of the blood-brain barrier, these in turn can result in vasogenic brain edema⁽¹⁻³⁾. Uncontrolled hypertension, uremia, immunosuppressant therapy, and systemic lupus erythematosus have been reported as higher prevalence of PRES⁽¹⁾. In our case, the patient was in stage 4 chronic kidney disease with an estimated glomerular filtration rate of 21.2 mL/min/1.73 m², whom was in the populations at high risk for PRES.

PRES typically involves symmetrically over bilateral parieto-occipital and posterior frontal lobes⁽¹⁾. Unexpectedly, PRES rarely affects only in the brainstem, termed central-variant PRES⁽⁴⁾, which may resemble other etiologies involving the same anatomical areas such as CPM or pontine infarcts in the clinical features and difficulty in differential by the CT of brain obtained in the emergency department⁽³⁾.

The clinical manifestations of these different etiologies involving the brainstem could share overlapping clinical features, are diverse and may exhibit dizziness, cranial nerve palsies, limb or trunk ataxia, seizures, consciousness decline, and ultimately fatality in untreated patients, which is a lethal condition^(1,3). In our case, the similar hypodense pontine lesions in the brain CT associated with altered consciousness may be difficult to differentiate brain stem infarction from central-variant PRES, thus direct physicians to a dilemma: whether or not introduce antihypertensive agents for possible PRES, whereas normalization of blood pressure is a contraindication during the acute phase of ischemic stroke. However, misdiagnosis by the physicians results in an inadequate management and may lead to irreversible neurologic deficits or even death, and possible medico-legal problems. Clues for the diagnosis of central-variant PRES are established based on the use of antihypertensive agents followed by dramatic relief from neurological dysfunction in combination with characteristic homogeneous pontine hypodense lesions in the brain CT⁽²⁾.

MRI may help in differentiating, from which the hyperintensity on T2-weighted FLAIR and ADC series in combination with isointensity to hypointensity on DWI map might indicate a process of vasogenic edema as in our case, whereas these signals will demonstrate contrary results in the cytotoxic edema such as CPM and ischemic stroke⁽¹⁻⁵⁾. Therefore, we suggest that an immediate MRI should be obtained when encountered such dilemma of choosing antihypertensive agents or therapeutic methods (such as antiplatelet agents, intra-arterial/venous thrombolysis, recanalization for basilar artery occlusion) for presumed ischemia brainstem infarct, which further guided the treatment of this patient. Under the condition of lacking MRI facility in emergency department, an additional CT angiogram⁽⁶⁾ for survey of the posterior circulation could also be helpful in assisting decision making.

In conclusion, this case report aims to raise the awareness that when hypodense brainstem lesions on brain CT in patients with progressive neurological dysfunction, the unusual condition of central-variant PRES should be considered in the differential diagnosis to avoid inadequate management. Cranial MRI may help in diagnosis and dealing with of these patients with overlapping

radiological and clinical abnormalities.

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