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Isolated Brainstem Involvement in Posterior Reversible Encephalopathy Syndrome

İzole Beyin Sapı Tutulumu Yapan Posterior Reversibl Ensefalopati Sendromu

Tarkan Ergün, Hatice Lakadamyalı
Başkent University Faculty of Medicine, Department of Radiology, Antalya, Turkey

Summary

Posterior reversible encephalopathy syndrome (PRES) is a clinical and radiologic entity characterized by headache, variable mental status, epileptic seizures, visual disturbances, and typical transient changes in the posterior cerebral perfusion. Parieto-occipital region is the most commonly involved site in this syndrome. Less commonly, brainstem, basal ganglia, and cerebellum are involved besides the supratentorial white matter areas. However, isolated brainstem involvement is very rare. We here present a case of isolated brainstem involvement in posterior reversible encephalopathy syndrome diagnosed by diffusion-weighted magnetic resonance imaging. *(Turkish Journal of Neurology 2013; 19:44-111-3)*

Key Words: Brain stem, posterior reversible encephalopaty, hypertensive encephalopathy

Özet

Posterior reversibl ensefalopati sendromu (PRES), baş ağrısı, metal durum değişiklikleri, epileptik nöbetler, görme bozuklukları ve beynin posterior dolaşım alanında yaptığı tipik geçici değişiklikler ile karakterize klinik ve radyolojik bir antitedir. Bu sendromda en sık parietooksipital alan tutulmaktadır. Daha az sıklıkta ise supratentorial beyaz cevherdeki lezyonlara ek olarak beyin sapı, bazal ganglia ve serebellumda da tutulum izlenebilir. Ancak izole beyin sapı tutulumu oldukça nadirdir. Burada difüzyon ağırlıklı manyetik mezonans görüntüleri ile tanısı konan ve izole beyin sapı tutulumu ile giden bir PRES olgusu sunulmaktadır. (Türk Nöroloji Dergisi 2013; 19:111-3)

Anahtar Kelimeler: Beyin sapı, posterior reversibl ensefalopati, hipertansif ensefalopati

Introduction

Posterior reversible encephalopathy syndrome (PRES) is a clinical and radiological condition characterized by headache, mental state changes, epileptic seizures, visual deficits and the temporary changes in the posterior circulatory area of the brain (1). The most common finding in the imaging is the white matter involvement in the posterior parts of the cerebral hemisphere, especially the parietooccipital lobes on both sides. Less frequently, supratentorial white matter lesions and basal ganglia and cerebellar involvement are also seen. Isolated brainstem involvement, on the other hand, is extremely rare.

Case

The 56-year-old male patient came to our emergency service with headache and disequilibrium which had been going on for 2 days. His medical history included a history of myocardial infarction and insufficiently managed hypertension. His neurological examination showed only mild ataxia. His consciousness was clear and the cranial nerves were functional. His motor and sensory examination was normal. His deep tendon reflexes were bilateral normoactive, and plantar reflex was bilateral flexor. The initial measurement of the blood pressure at the emergency service was 23-/140 mmHg which prompted an aggressive antihypertensive treatment.

Address for Correspondence / Yazışma Adresi: Tarkan Ergün MD, Başkent University Faculty of Medicine, Department of Radiology, Antalya, Turkey
Phone: +90 242 511 25 11 E-mail: tarkanergun@yahoo.com
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In the laboratory examination, leucocyte count was 14600/mm3, hemoglobin concentration was 16.5 g/dL, hematocrit was 49.1%, C-Reactive protein was 0.28 mg/L, blood glucose concentration was 105 mg/dL, BUN 11 mg/dL, serum creatinine was 0.89 mg/dL. Tests for liver function (aspartate aminotransferase: 25 U/L, alanine aminotransferase: 17 U/L) were normal. Serum electrolyte concentrations for calcium, potassium and sodium were 9.6 mmol/L, 4.10 mmol/L and 146 mmol/L, respectively.

Multiple areas with low attenuation were found in the pons in the computerized tomography (Figure 1). In his brain magnetic resonance (MR) imaging, FLAIR and T2-weighted sequences showed diffused hyperintensities in brainstem. There were no other pathologies beside the lacunar infarction located in right supraventricular white matter of the supratentorial area, and the non-specific ischemic gliotic lesions located in both periventricular white matter and centrum semiovale. The diffusion weighted MR volume was normal. In the apparent diffusion coefficient (ADC) map, the ADC value (lesioned region: 1.0^3x10^3 mm^2/s, normal region 0.88x10^3 mm^2/s) was found to be high (Figure 2). After the intense antihyperintensive drug treatment, the patient's complaints were reduced and he was discharged without any remaining symptoms at the end of the 3rd day. In the follow-up MRI after two weeks, the lesions disappeared completely.

Discussion

PRES involves primarily the parietooccipital lobe. If the main lesions are located outside of parietooccipital lobe, those conditions are classified as atypical PRES. In a recent MR study where McKinney et al. evaluated 76 PRES cases, the 98.7% of the cases had lesions in parietooccipital lobe, 78.9% in frontal lobe, 68.4% in the temporal lobe, 34.2% in cerebellum, 30.3% in thalamus, 18.4% in brainstem, and 11.8% in basal ganglia (2). Only 1 case showed high intensity lesions in the brainstem, thalamus and deep white matter in the absence of parietooccipital involvement. Bartynski et al. found similar quantities in their study involving 136 PRES cases and they detected primarily supratentorial region involvement in the cases where the brainstem had lesions (3). In the literature, even though there are numerous instances of lesions predominantly involving brainstem, isolated brainstem involvement is actually extremely rare, much like in our case study (4, 5, 6, 7, 8).

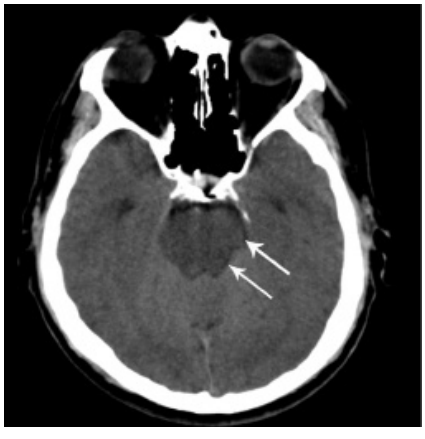

Figure 1: Computerized brain tomography without contrast agents show wide spread hypodense lesions in the pons

PRES diagnosis can be made on the basis of typical MR findings accompanying hypertension. However, the diagnosis is more challenging in the isolated brainstem involvements and requires ruling out brainstem infarction, glioma in pons, central pontine myelinolysis and infective encephalitis (4). In our case, the possibility of an infarction was not significant due to the low agreement between clinical findings and the severity of radiological findings, also the lack of diffusion restriction in the diffusion-weighted imaging. During the clinical course, no electrolyte disorder was present that could cause central pontine myelinolysis. The fact that reducing the blood pressure alleviated the neurological findings associated with severe hypertension suggested the diagnosis of PRES due to isolated involvement of the brainstem. The cerebrospinal fluid examination and MR spectroscopy required for eliminating encephalitis and glioma were not performed due to this reason.

In PRES cases, it is possible to see an increase in ADC level in the diffusion-weighted MR sequences, which can be reversible on the condition that proper treatment is followed. This knowledge can be valuable to differentiate between PRES and infarctions, central potine myelino myelinolysis, encephalitis, and the metabolic diseases with high cellularity mass and cytotoxic edema. However, it is also possible to see decreased diffusion indicating irreversible lesions, as a marker of unfavorable prognosis of a PRES case (9). In our report, there were no diffusion restrictions in the diffusion-weighted volumes and the lesions disappeared after 2 weeks.

The known reasons of PRES include hypertension, eclampsia, preeclampsia, immunosuppressive and cytotoxic drugs, kidney failure with hypertension, collagen vascular diseases, severe hypercalcemia, amyloid angiopathy, thrombotic thrombocytopenic purpura, HIV infection, systemic lupus erythematosus, acute

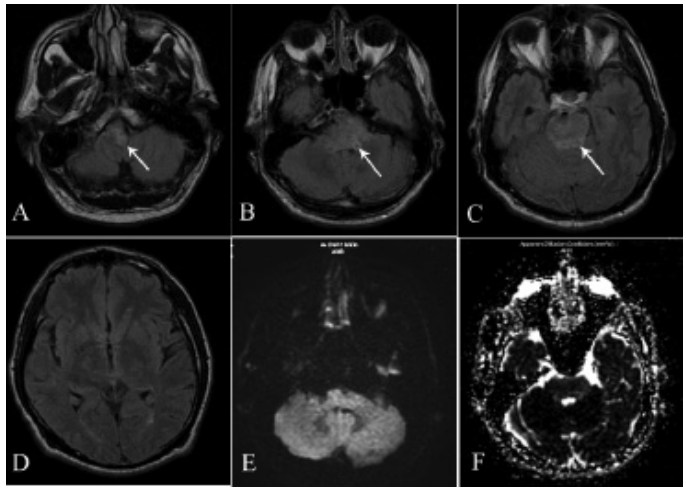

Figure 2: Axial FLAIR images (A, B, C, D) show a wide-spread increase in intensity in the brainstem. The parietooccipital lobe is seemingly preserved. The diffusion-weighted images (E) show a normal outlook while the increase in ADC (F) draws attention.

intermittent porphyria and organ transplantation (10). The cause for the central variant of PRES is hypertension, as seen in our case (4). In a literature study involving 20 cases, Shintani et al. found the blood pressure of the patients to be 213.2±29.7 and 127.6±25.6 mmHg at the time of initial consultation (11).

There have been 2 hypotheses regarding the pathophysiology of PRES. First one suggests the vasospasm caused by acute blood pressure and the second one pertains to the loss of autoregulation. In the first hypothesis, the possibility of ischemia in the arterial border zone and cytotoxicity due to vasospasm comes to mind (12). In the other hypothesis that was put forward recently and supported by diffusion imaging, it was argued that the loss of autoregulation causes dilation of cerebral arterioles. In this case, plasma and red blood cells cause vasogenic edemas by passing from intracellular area to the extracellular one (13). Due to the fact that the sympathetic innervation is lower in the posterior cerebral arterial circulation, the lesions are predominantly focused on the parietooccipital region. In the pathological investigation of the PRES brain, interstitial edema, petechial microhemorrhage, fibrinoid necrosis on the arteriole wall and little or no infarction finding are seen.

Early diagnosis and treatment is extremely important for PRES. Delays in the diagnosis and treatment may lead to ischemic and hemorrhagic lesions and subsequently permanent neurological damage or even death (14). However, Kitaguchi et al. (15) reported a more optimistic prognosis for brainstem variant of PRES as compared to the classical PRES (15). The regulation of blood pressure is essential in the treatment. Following the proper treatment of hypertension, the neurological involvement soon disappears.

To conclude, both the clinician and the radiologist should be aware that PRES can present with isolated brainstem involvement. The lack of association between the clinical findings and the diffused locations of the lesions, the co-occurrence of neurological findings with hypertension and the rapid response to treatment present important clues for the diagnosis.

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