Diffuse Brain Involvement in Hypertensive Encephalopathy Secondary to Renal Artery Stenosis: Case Report

Renal Arter Stenozuna Sekonder Gelişen Hipertansif Ensefalopatide Yaygın Beyin Tutulumu

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Geliş Tarihi/*Received:* 05.12.2008
Kabul Tarihi/*Accepted:* 24.02.2009

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ABSTRACT A nine-year-old male patient admitted to a pediatry clinic with severe headache, nausea, vomiting, high fever and visual dysfunction. The patient's blood pressure was 220/170 mmHg. He had optic disc edema on the right eye, hemorrhage in the left eye; high serum urea and creatinine levels with high plasma renin activity and low cretinine clerance values. In renal ultrasonography, left kidney dimensions was decreased and there was right renal compensatory hypertrophy. In renal magnetic resonance angiography (MRA), a high grade stenosis at the proximal segment of the left renal artery was diagnosed. In cranial conventional MR images, diffuse hyperintense lesion areas were seen in bilateral basal ganglions, mesencephalon, cerebellum, subcortical areas of brainstem and in deep areas of white matter. Diffusion weighted imaging (DWI) was normal and in apparent diffusion coefficient (ADC) map signal intensity was mildly increased. These clinical findings and MR imaging (MRI) abnormalities were consistent with those of hypertensive encephalopathy, and resolved with control of hypertension.

Key Words: Hypertensive encephalopathy, renovascular hypertension, magnetic resonance imaging

ÖZET Dokuz yaşında erkek hasta, çocuk hastalıkları kliniğine şiddetli baş ağrısı, bulantı, kusma, yüksek ateş ve görme bozukluğu ile başvurdu. Hastanın kan basıncı 220/170 mmHg idi. Sağ gözde optik disk ödemi, sol gözde hemoraji olan hastada yüksek serum üre ve kreatinin düzeyleri, yüksek plazma renin aktivitesi ve düşük kreatinin klerens testi mevcuttu. Renal ultrasonografide sol böbrek boyutları azalmıştı ve sağ böbrekte kompansatuar hipertrofi mevcuttu. Renal manyetik rezonans anjiyografi (MRA)'de sol renal arterin proksimal bölümünde ileri derecede darlık saptandı. Konvansiyonel manyetik rezonans imajlarda bilateral bazal ganglionlar, mezensefalon, serebellum, beyin tabanının subkortikal alanları ve derin beyaz cevher alanlarında yaygın hiperintens lezyon alanları görüldü. Difüzyon ağırlıklı görüntüleme (DWI) normaldi ve görünür difüzyon katsayısı (ADC) haritasında sinyal intensitesi hafif artmıştı. Sol laparoskopik nefrektomi ve antihipertansif tedaviden sonra, hastanın semptomları ve manyetik rezonans incelemedeki bulgular büyük oranda geriledi. Bu klinik bulgular ve manyetik rezonans görüntüleme (MRG) anormallikleri hipertansif ensefalopati ile uyumluydu ve hipertansiyonun kontrol altına alınmasıyla düzeldi.

Anahtar Kelimeler: Hipertansif ensefalopti, renovasküler hipertansiyon, manyetik rezonans görüntüleme

Turkiye Klinikleri J Cardiovasc Sci 2009;21(3):484-8

Posterior reversible encephalopathy (PRES) is also known as hypertensive encephalopathy, reversible posterior cerebral edema and posterior reversible leucoencephalopathy.[1] Clinically it is characterised with headache, seizures, visual dysfunctions, focal neurological signs and

changes in mental status. Morbidity and mortality are closely related with early diagnosis and appropriate antihypertensive treatment.[2-7] Typical MRI findings are vasogenic edema areas that are symmetrically scattered in posterior cerebral circulation areas. Generally it effects white matter, but rarely cortical lesions can also be seen. Hypertensive encephalopathy in renal artery stenosis effects whole brain very rarely. In this child case, we showed a diffuse brain involvement secondary to arterial hypertension because of a renal artery stenosis.

CASE REPORT

Our case is a nine-year-old male patient who had admitted to a pediatry clinic with severe headache, nausea, vomiting and fever. Visual dysfunction and headache were present for 1 year and in physical examination. Meningeal irritation findings were found and Kernig's/Brudzinski's tests were positive. Arterial blood pressure was 220/170 mmHg. In ophtalmoscopic examination, papillary edema and diffuse exudations around optic disc of the right eye and intravitreal hemorrhage in the left eye were diagnosed. Left ventricular hypertrophy was seen in echocardiographic evaluation. Blood urea nitrogen (BUN) was 72 mg/dL (<50 mg normal). Na, K and Cl values were 126 mEq/l, 2.6 mEq/l, 88 mEq/l respectively (normal values are 136-145 mEq/l, 3.5-5.1 mEq/l, 98-107 mEq/l respectively). Blood creatinine level was in normal range (0.70 mg/dL). Plasma renin activity was calculated as 31 ng/mL/sec (normal range is 0.50-5.90 ng/mL/sec). Urine Na value in 24 hour urine collection was calculated as 270 mg/day (normally >200 mg/day). Creatinine clearance was calculated as 50 mL/min/1.73 m² (normal range is 83-92 mL/min./1.73 m²).

Cranial MRI evaluation was performed with 1.5 Tesla (Gyroscan NT Interna, Philips Medical Systems, Holland) MR machine. In axial plane T2 and fluid-attenuated inversion-recovery (FLAIR) weighted sections; symmetrically placed, hyperintense, multiple patchy lesions were diagnosed in bilateral frontoparietooccipital regions, bilateral basal ganglions, periventricular and supraventricular areas, mesencephalon, cortical and subcortical white matter, brainstem and bilateral cerebellum (Figure 1: A-C). After contrast infusion, there was no contrast uptake in lesion areas. In ADC map, mildly increased signal intensity was determined in aforementioned lesion areas (Figure 1: D). In DWI, no diffusion restricted areas were present (Figure 1: E). DWI was performed in axial plane with the values of b=1000 s/mm². In addition, in order to eleminate T2 shine-through effect, ADC map was obtained from diffusion weighted sections.

At the first glance, no diffusion restrictions in lesion areas and no contrast uptake in basal cisterns and parenchyma, ruled out a possible diagnosis of acute infarction and menengitis/metastasis respectively. Severe arterial hypertension, elevation of BUN value, decrease in blood electrolyte levels, increased plasma renin activity index and decrease in creatinine clerance test were accepted as diagnostic clues of encephalopathy secondary to hypertension of renal origin. In renal ultrasonography (Toshiba Aplio, Japan), left kidney was atrophic (60*35 mm) and right kidney dimensions were increased (116*46 mm) consistent with compensatory hypertrophy. In renal MRI evaluation; left kidney was detected as atrophic (Figure 2). In this radiologic evaluation, balance turbo field echo (B-TFE) sequence was used and scanning parameters were: TR/TE values= 3.8/1.9 msec; cross-sectional thickness=8 mm; section interval= 0 mm; scanning area (FOV)=300 cm; matrix dimensions= 256 x 256; flip= 60; accuisition time= 25 sec.

At 12th day of antihypertensive treatment, diffusion and T2 weighted MRI evaluations were repeated and complete resolution of aforementioned lesions were seen (Figure 3: A-C). In addition, laparoscopic left nephrectomy was performed at 24th day of treatment. After the operation, evident regression in blood pressure values and evident improvements in biochemical blood parameters were seen.

DISCUSSION

PRES is a clinical entity developing after hypertension acutely or subacutly and characterized

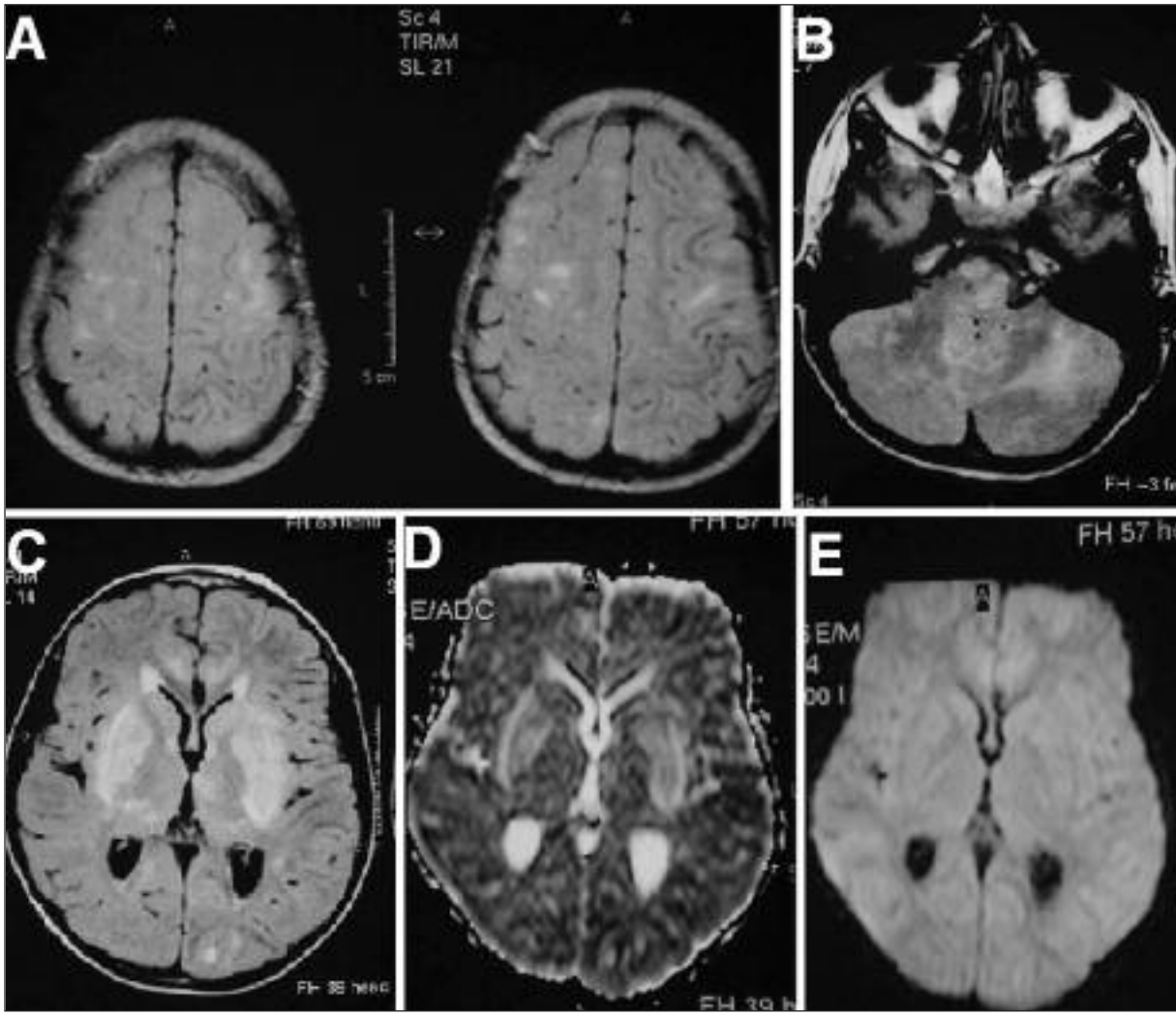

FIGURE 1: Initial MR images: A-C, Axial FLAIR MR images show symmetric abnormal signal intensity in supraventriculer cortico-subcortical regions, pons and cerebellum, basal ganglions (respectively). D, In ADC map, increased signal intensity areas in basal ganglions. E, Normal findings in DWI.

with reversible radiologic and clinical signs. An increase in the vascular permeability and dysfunction in cerebral autoregulation in hypertensive states and cerebral vasogenic edema, arising as a result of these abnormalities were thought to be responsible for neuroparenchymal lesions seen in PRES.[3,6,8,9] It is suggested that, characteristic radiological views of lesions, in the posterior cerebral circulation are derived from the regional differences of intracranial adrenergic receptor distribution.[9] Since, sympathetic innervation of the posterior cerebral vasculature is scarce, compensatory protective response to the sudden increases of arterial blood pressure is not efficient.

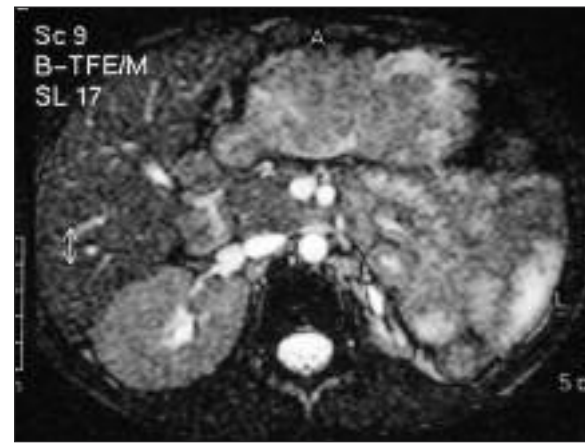

FIGURE 2: In renal MRI evaluation, atrophy of the left kidney and compensatory hypertrophy of the right kidney.

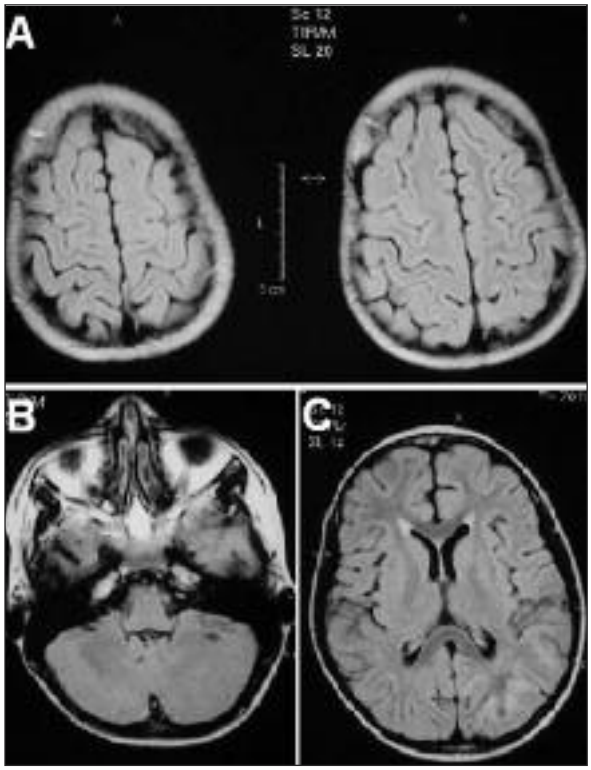

FIGURE 3: Follow-up MR images 12 days after initial MRI. A-C, FLAIR images show complete resolution of lesions.

Lesion localisations seen in PRES are correlated with the prognosis of the patient. In cases of mild hypertension, the findings are usually supratentorial; in cases of more severe hypertension, similar changes are noted in the basal ganglia, cerebellar hemispheres, and brainstem.[3,8,9] In hypertensive encephalopathy, lesions in the posterior cerebral circulation areas are typical. Although periventricular region can be effected, cortical and subcortical involvement is rare. In addition to involvement of posterior cerebral circulation areas in PRES, involvement of anterior cerebral circulation areas, pons, cerebellum and brain stem were reported infrequent.[1] A few cases of hypertensive encephalopathy with dominant brainstem involvement were reported in the literature.[10-12]

Fujiwara et al reported a case of 38-year-old male patient as a rare form of hypertensive encephalopathy characterized with diffuse involvement of brainstem and nearly total preservation of supratentorial region.[13]

In hypertensive encephalopathy, with the use of DWI method, vasogenic edema can be reliably discriminated from cytotoxic edema seen in cerebral ischemia, with no restriction in diffusion in vasogenic edema. In complicated PRES cases with irreversible lesions, diffusion restriction is seen.[14]

Brainstem involvement is only seen in sudden arterial blood pressure elevations and is related with bad prognosis.[12] If lesions are restricted to the base of the brain, glioma, acute infarction, osmotic myelinolisis, encephalitis and vasculitis such as neurobehcet should be thought in differential diagnosis. In differential diagnosis, determination of acute infarction in acute cases has clinical importance. In addition to its great capability to reach a definitive diagnosis, quantitative diffusion evaluation can also be helpful to start an appropriate and early treatment.

In pediatric population, PRES is rare and generally occurs after renal diseases as acute glomerulonephritis, renal vascular hypertension or chronic renal failure.[15-17] Estepa et al reported 6 cases of hypertensive encephalopathy in a group of 21 patients aged between 5-15 years with renovascular hypertension after renal artery stenosis.[18] Bıçak et al reported two cases of PRES in a pediatric population after poststreptococcic glomerulonephritis, with bilateral symmetrical hyperintense lesions in parietooccipital region and after appropriate treatment they showed that, all lesions were subsided in control MRI scannings.[19]

In our case, we have seen pathological involvement of bilateral basal ganglions, bilateral frontoparietooccipital regions, periventricular and supraventricular areas, mesencephalon, brainstem, bilateral cerebellum and cortical and subcortical white matter secondary to hypertension as a result of unilateral renal artery stenosis. After antihypertensive treatment, complete resolution of the lesions were shown.

In cases of delayed treatment, progression to ischemia, massive infarction and rarely mortality can be seen. Clinical symptoms and radiological findings in PRES are completely reversible after treatment. As a result, in hypertensive encephalopathy, radiological evaluation is important for early diagnosis and early initiation of treatment.

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