



Posterior leukoencephalopathy syndrome due to hypertension

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Summary

Posterior leukoencephalopathy syndrome (PLES) which is associated with acute elevations in blood pressure is a rare syndrome characterised by headache, nausea, vomiting, confusion, seizures, and visual disturbances with specific signs on brain images. Here, we report three patients with PLES. Three patients who were on continuous ambulatory peritoneal dialysis because of chronic renal failure were admitted with seizure, headache and confusion. They were 6.5, 9 and 13 years old and had acute elevations in blood pressure. By means of clinical and neuroradiological signs they were diagnosed with PLES. While evaluating patients with chronic renal failure who have nausea, vomiting, confusion and visual disturbance accompanying hypertension, PLES should be considered in the differential diagnosis and brain MR images should be searched for the specific findings. (*Turk Arch Ped* 2013; 48: 156-159)

Key words: Chronic renal failure, hypertension, magnetic resonance imaging, posterior leukoencephalopathy

Introduction

Posterior leukoencephalopathy syndrome (PLES) is a rare disease characterized with headache, nausea, vomiting, confusion, visual disturbances and seizures caused by acute elevation of the blood pressure and specific findings on imaging. It was described by Hinchey et al. (1) in 1996 for the first time. The ethological factors include hypertensive encephalopathy, acute poststreptococcal glomerulonephritis, treatment with immunosuppressive drugs, eclampsia, fluid retention and blood transfusion. The neuroradiological findings of posterior leukoencephalopathy syndrome typically include high-signal changes at the cortical and subcortical levels in the parietal and occipital lobes in the posterior areas of the brain hemispheres. In addition, the frontal and temporal lobe, basal ganglia and cerebellum may also be involved (1,2). The clinical and neuroradiological findings of the diseases do not vary according to the etiology.

In this article, three patients who were diagnosed with PLES with clinical and neuroradiological imaging findings were presented.

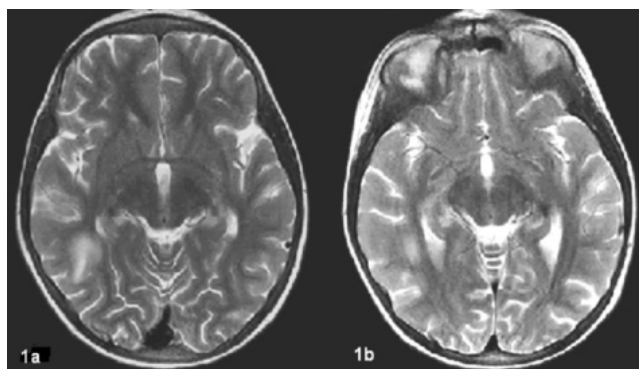
Case-1

A 9-year-old female patient who was being followed up in a continuous ambulatory peritoneal dialysis program (CAPD) for one year because of chronic renal failure (CRF) which developed secondary to rapidly progressing glomerulonephritis presented with complaints including headache, confusion and seizure. On physical examination, the consciousness was blurred and the blood pressure was found to be 170/100 mmHg. Deep tendon reflexes (DTR) were hyperactive and she had no pathological reflex. The laboratory findings were as follows: WBC 5880/mm³, hemotocrite 23.8%, hemoglobin 8.7 g/dL, platelets 132 000/mm³, glucose 102 mg/dL, BUN 96 mg/dL, creatinine 7.3 mg/dL, sodium (Na) 128 mEq/L, potassium (K) 3.1 mEq/L, calcium (Ca) 8.2 mg/dL, phosphorous (P) 7.6 mg/dL and alkaline phosphatase (ALP) 262 U/L. On brain magnetic resonance imaging (MRG), areas showing increased signal with irregular borders were found in the gray and white matter in the right temporooccipital region on T2A images (Picture-1a). The patient was hospitalized in the Pediatric Intensive Care Unit. The continuous ambulatory peritoneal

dialysis program was continued and sodium nitroprusside infusion was started for hypertension. The blood pressure was controlled in one hour. Sodium nitroprusside treatment was tapered and discontinued and supportive treatment for CRF was continued. On electroencephelogram (EEG), slow wave discharges arising from the parietooccipital region were observed in both hemispheres. The clinical findings of the patient improved in the follow-up and follow-up brain MRI findings were found to be normal one month later (Picture-1b). The case was considered as PLES because of presence of acute neurological findings in the patient who had chronic renal failure, specific imaging findings and improvement of the clinical and imaging findings in the follow-up.

Case-2

A 6,5 year-old female patient who was being followed up because of CRF which developed secondary to steroid-resistant nephrotic syndrome for four years and who was receiving CAPD presented with complaints including seizure, change in consciousness and dyspnea. The blood pressure was measured to be 200/110 mmHg. On neurological examination, the consciousness was blurred, DTR were normoactive and pathological reflexes were absent. On fundoscopic examination, the optic nerves were found to be edematous in the right and left side and streaks were found in the left macula. The laboratory findings were as follows: WBC 6 800/mm³, hemotocrite 21.1%, hemoglobin 6.8 g/dL, platelets 161 000/mm³, glucose 86 mg/dL, BUN 64 mg/dL, creatinine 6.2 mg/dL, Na 141 mEq/L, K 5.8 mEq/L, Ca 9.3 mg/dL, P 6.7 mg/dL and ALP 156 U/L. High-signal changes were observed in T2A series extending from the posterior part of the right temporal lobe to the right parietal lobe and in the subcortical white matter in the right posteroparietal region (Picture-2a). The patients

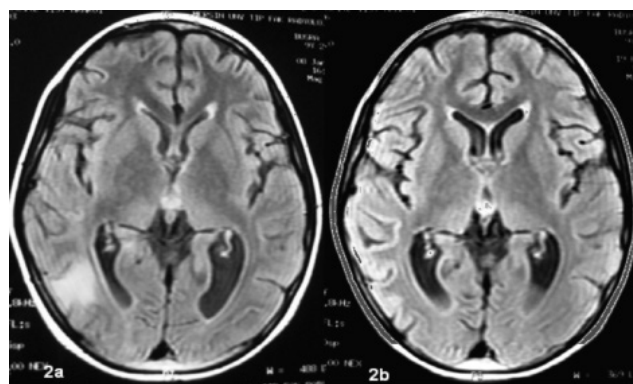


Picture 1a: Areas showing increased signal with irregular borders in the gray and white matter in the right temporooccipital region on T2A images. **1b:** Follow-up MRI: the lesion persists, though it is reduced in size.

was internalized in the Pediatric Intensive Care Unit and sodium nitroprusside infusion was started for hypertension. Her blood pressure was controlled. Sodium nitroprusside was tapered and discontinued in the first 8 hours. Dialysis and supportive treatments were continued for chronic renal failure. Follow-up brain MRI was found to be normal 15 days later (Picture-2b). The case was considered as PLES because of a history of CRF, presence of acute neurological findings including seizure, confusion and dyspnea and improvement of the clinical and imaging findings completely in the follow-up.

Case-3

A 13-year-old female patient who was being followed up because of CRF which developed secondary to vesicoureteral reflu for 5 years and who was receiving CAPD presented with complaints including seizure, change in consciousness and visual disturbance. The blood pressure was measured to be 190/110 mmHg. On neurological examination, the consciousness was blurred, DTR were active and the Babinsky reflex was positive. The laboratory findings were as follows: WBC 7 480/mm³, hemotocrite 28.5%, hemoglobin 9.8 g/dL, platelets 72 000/mm³, glucose 82 mg/dL, BUN 44 mg/dL, creatinine 4.4 mg/dL, Na 136 mEq/L, K 5.7 mEq/L, Ca 8.8 mg/dL, P 1.9 mg/dL and ALP 215 U/L. On brain MRI, high-signal changes were observed in the "FLAIR" series in the subcortical white matter in the posterior parts of bilateral occipital lobes (Picture-3a). The patients was internalized in the Pediatric Intensive Care Unit with a prediagnosis of hypertensive encephalopathy and sodium nitroprusside infusion was started. After the blood pressure was controlled in the first 24 hours, sodium nitroprusside infusion was tapered and discontinued considering the



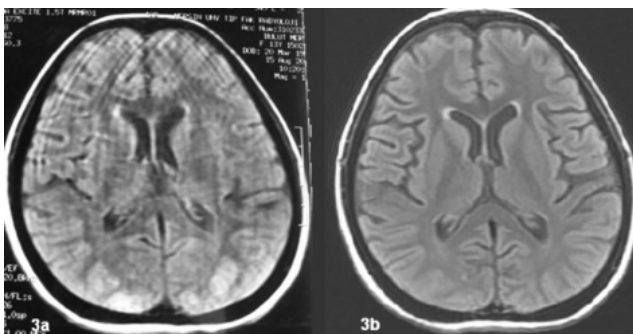
Picture 2a: High-signal changes in the FLAIR series extending from the posterior part of the right temporal lobe to the right parietal lobe and in the subcortical white matter in the right posteroparietal region. **2b:** Follow-up MRI was evaluated to be normal.

toxic effects. Dialysis and supportive treatments for chronic renal failure were continued. Follow-up brain MRI was evaluated to be normal 15 days later (Picture-3b). The case was considered as PLES because of a history of CRF, presence of acute neurological findings including seizure, confusion and visual disturbance and improvement of the clinical and imaging findings completely in the follow-up.

Discussion

Posterior leukoencephalopathy syndrome is a syndrome which occurs as a result of many different causes. Although it is observed rarely in the pediatric age group, the most common cause is hypertension which develops in relation with CRF. PLES has also been reported in acute post-streptococcal glomerulonephritis, hemolytic uremic syndrome, use of cytotoxic and immunosuppressive drugs, blood transfusion, collagen vascular diseases and acute intermittent porphyria which occur before advance disruption in renal functions (1,2,3,4). Each of our three patients was being followed up in the CAPD program because of end-stage renal disease and was diagnosed with PLES related with hypertension which developed suddenly.

The diagnosis of the disease is made with clinical and radiological findings. The most common clinical findings include headache, nausea, vomiting, change in consciousness, visual disturbance and seizures. Change in consciousness may range from lethargy to severe coma (3,4). Incecik et al. (5) evaluated 9 patients who were diagnosed with PLES retrospectively. They found that the most common clinical findings included seizures (6/9), headache (6/9) and change in consciousness (4/9). Nausea, vomiting and blurred vision were found more rarely. In our study, each of our three patients had complaints including change in consciousness and seizures. One patient presented with headache and another patient presented with visual disturbance.



Picture 3a: High-signal changes in the “FLAIR” series in the subcortical white matter in the posterior parts of bilateral occipital lobes being more prominent in the right side. **3b:** Follow-up MRI was evaluated to be normal.

Brain MRI findings in posterior leukoencephalopathy syndrome are specific for the disease. Generally, hyperdense signal changes are observed in bilateral, symmetrical T2A and “FLAIR” series in the posterior parts of the brain hemispheres especially in the parietooccipital white matter. These findings are considered pathognomonic for PLES (1,2,3,4). Hinchey et al. (1) found hyperdense signal changes in the T2A series in bilateral occipital lobes in 14 patients, in the posterior parietal lobes in 13 patients, in the posterior temporal lobes in 9 patients, in the pons in 2 patients and in the thalamus and cerebellum in one patient in their study which included 15 patients diagnosed with PLES. Incecik et al. (5) found hypointense lesions in the T1A series and hyperintense lesions in the T2A series in the parietal and occipital lobes in 6 patients, in the frontal lobes in 2 patients, in the cerebellum in one patient and in the basal ganglia in one patient in their study which included 9 patients. We found lesions in the temporooccipital region in two of our patients and only in the occipital region in one of our patients. In recent years, publications have reported that the brain stem and anterior supply areas are involved predominantly without substantial change in the posterior supply areas (6,7).

Two theories about the pathophysiology of the disease have been proposed. In the first theory, it is thought that ischemia and cytotoxic edema occur in the borders of the arterial supply areas due to vasospasm which develops as a result of acutely increased blood pressure. In the second theory, it is thought that the blood-brain barrier is affected as a result of endothelial dysfunction and loss of autoregulation of the arteries in the parietooccipital region due to acutely increased blood pressure. In this condition, plasma and red blood cells are transferred into the extracellular space and vasogenic edema develops. Due to lower sympathetic activity on the posterior brain arteries the lesions have been proposed to be observed more frequently in the parietooccipital region (8,9).

Early diagnosis and treatment of posterior leukoencephalopathy syndrome is substantially significant. It may lead to permanent brain damage, if not diagnosed and treated early. Treatment of the disease is evident. With elimination of the cause of the disease the clinical and radiological findings can improve completely. In our patients, it was observed that the clinical and radiological findings improved completely with control of hypertension. The blood pressure should be reduced in a restricted way in patients who develop PLES related with hypertension. To prevent development of multiple organ dysfunction and brain infarct related with acute reduction in the blood pressure the mean arterial blood pressure should be reduced by 20-25% in the first 1-2 hours (9,10).

Conclusively, PLES may develop in relation with many different causes. The diagnosis of the disease is made with clinical and specific radiological findings. The clinical and radiological findings can improve completely with early diagnosis and treatment. PLES should be considered in patients with CRF who present with headache, nausea, vomiting, change in consciousness, visual disturbance and seizures accompanying hypertension. MRI should be ordered in order to determine specific brain imaging findings.

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