Isolated Cerebellar Reversible Leukoencephalopathy Syndrome in a Patient with End Stage Renal Disease

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Abstract

We report an adult end-stage renal disease patient with only cerebellum involvement of reversible posterior leukoencephalopathy syndrome (RPLS). A 37-year-old woman, diagnosed as chronic glomerulonephritis, was admitted to our hospital with progressive visual disturbance and severe headache. MRI revealed hyperintense signal intensity changes restricted in the cortex and subcortical white matter of the cerebellum. With appropriate control of blood pressure and intracranial pressure, her symptoms were improved and complete resolution of previous hyperintense lesion was shown on MRI after the 19th day of admission. This is first case of isolated cerebellar involvement of RPLS without other cerebral involvement in an adult.

Key words: leukoencephalopathy, hypertension, end-stage renal disease, cerebellum

(Introduction)

Reversible posterior leukoencephalopathy syndrome (RPLS) is a clinico-radiological syndrome that includes symptoms such as headache, confusion, seizures, and visual disturbances and radiological findings of edema involving the white matter in the posterior regions of the cerebral hemispheres, and in particular bilaterally in the parieto-occipital regions (1). Involvement of additional areas of the brain in RPLS patients, such as the brain stem, cerebellum, basal ganglia and frontal lobes, has also been reported (1, 2). Involvement of RPLS in only this brain area has rarely been reported; there have been reports on an adult case with only brain stem involvement (3) and a child case with only cerebellum involvement (4). Here, we report case an adult patient with end-stage renal disease (ESRD) with only cerebellum RPLS involvement.

Case Report

A 37-year-old woman visited our emergency room because of severe headache and progressive visual disturbance. She had been diagnosed with chronic glomerulonephritis and hypertension about 4 years earlier at a nearby university hospital and renal biopsy had shown IgA nephropathy at that time. Thereafter she received losartan 100 mg and amiodipine 5 mg. She had decided to discontinue the treatment about 5 months previously. Visual deterioration and generalized edema developed 2 months previously and her headache started 10 days previously. The headache revealed persistent squeezing at both frontal areas. Her headache worsened even with all analgesics that were given.

On physical examination, her blood pressure was 210/150 mmHg, the pulse rate was 120 beats per minute, the breath rate was 20 times per minute and the body temperature was 36.4°C. There was no anemia at her conjunctivae and sclera. The chest examination revealed no abnormal findings. There was no hepatomegaly or splenomegaly on the abdominal examination. There was no evidence of vasculitis. Pretibial pitting edema was shown on both legs. The neurological examination revealed no specific findings. However, papilledema and hemorrhage were found on the funduscopic examination.

Her initial serum BUN and creatinine level was 85 mg/dl and 14.4 mg/dl, respectively. The peripheral WBC count
Figure 1. Cranial MRI images. The T2-weighted and fluid-attenuated inversion recovery (FLAIR) images (A, B) revealed no signal intensity changes in cerebral areas and (C) bilateral hyperintense signal intensity changes in the cortex and subcortical white matter of the cerebellum (arrow).

was 10,130/mm³ (neutrophils: 83.9%, lymphoid cells: 11.3%, monocytes: 2.1%). The hemoglobin level was 11.4 g/dL and the platelet level was 170,000/mm³. The total protein level was 5.6 g/dL and the albumin level was 3.1 g/dL. The serum electrolyte showed that the sodium level was 134.5 mmol/L, the potassium level was 4.4 mmol/L and the chloride level was 102.1 mmol/L. Her serum osmolality was 307 mosm/kg. The echocardiography revealed left ventricular hypertrophy. The kidney ultrasonography study showed bilateral shrunken kidneys and increased parenchymal echo. The T-2 intensified MRI image revealed hyperintense signal intensity changes that were restricted to the cortex and the subcortical white matter of the cerebellum and no abnormal density in the other areas (Fig. 1). Blood pressure control and intracranial pressure control was conducted immediately with IV labetalol, dexamethasone and mannitol under the suspicion of reversible posterior leukoencephalopathy syndrome. Total urine output was 400 ml per day and she showed volume overload finding on chest x-ray. We preferred continuous veno-venous hemodiafiltration to control volume overload and increased intracranial pressure. Three days after the hospitalization, the blood pressure was controlled at 150/90 mmHg and her headache improved. The MRI images conducted at the 19th day after admission showed complete disappearance of the high signal intensity at both cerebellum areas (Fig. 2). The previous hemorrhage and papilledema were also improved on the funduscopic examination. The patient is currently undergoing continuous ambulatory peritoneal dialysis and she is without any neurological sequelae.

Discussion

We report here on an adult case of end-stage renal disease with only cerebellum involvement of RPLS who was completely improved by controlling the blood pressure and intracranial pressure. This is the first case of isolated cerebellum involvement of RPLS in an adult ESRD patient.

The causes of RPLS are diverse, but the common precipitants are acute elevation of blood pressure, renal decompensation, fluid retention and treatment with immunosuppressive drugs (1). Hypertension from a renal origin has been reported to be a significant cause of RPLS, accounting for over 25% of cases in one study of both pediatric and adult patients (1). Yet there were no ESRD patients who have required renal replacement therapy in the above-mentioned report (two patients had lupus nephritis, one had acute glomerulonephritis and one had hepatorenal syndrome that was caused by acetaminophen intoxication). Only a few cases of RPLS associated with acute poststreptococcal glomerulonephritis (APSGN) have been reported in children (5).

The pathophysiology of RPLS is multifactorial. Most investigators favor the theory of hyperperfusion for RPLS.
There is an upper limit to cerebral autoregulation. But sudden elevations in blood pressure can disrupt the autoregulatory capacity of the brain vasculature. Focal transudation of fluid and petechial hemorrhage can occur due to the breakdown of the blood brain barrier (1). The relative paucity of sympathetic innervation in the posterior brain contributes to the susceptibility to hyperperfusion and vasogenic edema during acute blood pressure elevation (6).

RPLS is clinically characterized by the acute or subacute onset of lethargy, confusion, headache, visual disturbance and seizures in the setting of severe hypertension. It is important to differentiate RPLS from uremic encephalopathy, disequilibrium syndrome and other intracranial vascular infarctions if these signs and symptoms occur in ESRD patients. The radiologic findings of uremic encephalopathy usually show symmetric reversible lesions in the basal ganglia, internal capsule and paraventricular white matter. Disequilibrium syndrome usually occurs at the start of acute dialysis. Patients with RPLS have lesions in both the cortex and the subcortical white matter in the parieto-occipital lobes. But the calcarine and paramedical occipital lobe structures are usually not spared during bilateral infarction of the posterior cerebral artery territory. Simultaneous bilateral infarction of the posterior cerebral artery territory occurs in patients with embolism at the rostral basilar artery (7). We could exclude disequilibrium syndrome, uremic encephalopathy and infarction by considering that our patient has not started hemodialysis and there were no other uremic signs and symptoms except for headache and visual disturbances, and complete resolution on the neuro-imaging study was seen after 2 weeks, without any specific lesion.

The most common abnormality on neuroimaging in RPLS patients was edema involving the white matter in the posterior portions of the cerebral hemispheres, especially in the bilateral parieto-occipital regions (1). Involvement of additional areas of the brain in patients with RPLS, such as the brain stem, cerebellum, basal ganglia, and frontal lobes, has also been reported (1, 2). Because the lesions that are located in the brain stem and/or cerebellum may accompany the supratentorial parieto-occipital lesions, isolated involvement of RPLS in this area has rarely been reported, yet there has been an adult case with only brain stem involvement (3) and a child case with only cerebellum involvement (4). Okamoto et al reported that RPLS is one of the diseases involving the middle cerebellar peduncles, but the neuroimaging features included the midbrain, basal ganglia and thalamus in addition to the cerebellar peduncles in their RPLS patients (8).

We could not precisely explain why RPLS on neuroimaging study in the present patient was restricted to the cerebellum. The previous reports of isolated involvement into the brain stem or cerebellum also could not determine the cause of this finding (3, 4). Several reports in RPLS showed additional cerebellar involvement aside from other brain lesions (1, 2, 8). Although we could not explain the pathophysiology of isolated cerebellar involvement of RPLS, the present patient is the first case of isolated cerebellar RPLS without involvement of other cerebral regions in an adult ESRD patient.

In conclusion, RPLS in ESRD patients might have various radiological findings. As the radiological images of RPLS are quite diverse, the atypical lesions seen using MRI could rule out the diagnosis of RPLS in the presence of inappropriate clinical findings. It is important to consider the possibility of this rare disease in ESRD patients because there is rapid resolution of the clinical signs and symptoms and the imaging abnormalities of RPLS when the blood pressure is lowered without other management.

References