Acute Chorea Characterized by Bilateral Basal Ganglia Lesions in a Patient with Diabetic Nephropathy

Diyabetik Üremik Hastada Bilateral Bazal Ganglion Lezyonu ile Karakterize Akut Korea

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ABSTRACT

The syndrome of acute bilateral basal ganglia lesions associated with uremia presents with parkinsonism, altered mental status, and chorea in association with specific imaging findings in the basal ganglia. It is an uncommon syndrome seen generally in patients with diabetes mellitus and renal failure. We report a male patient with diabetes mellitus who received hemodialysis treatment 3 days a week for 5 years and suffered from choreic movements developed suddenly and associated with bilateral basal ganglia lesions. In the brain magnetic resonance (MR) imaging, isointense was detected in sequence T1 in the bilateral basal ganglions and hyperintense lesion was determined in T2 and FLAIR sequences. The patient was administered daily hemodialysis and neuroleptic treatment. After intensified hemodialysis, his symptoms and follow-up brain MR imaging showed marked improvement. The underlying mechanism of such lesions may be associated with metabolic, as well as vascular factors. Acute choreic movements may be seen in patients with diabetic nephropathy and intensification of hemodialysis treatment along with blood glucose regulation may provide improvement in this syndrome.

KEY WORDS: Basal ganglia, Chorea, Diabetes mellitus, Uremia

ÖZ

Üremi ile ilişkili akut bilateral bazal ganglion lezyonları ile seyreden sendrom, parkinsonizm, mental durum değişikliği ve korea kliniği ile ortaya çıkan, bazal ganglionlarda spesifik görüntüleme bulguları saptanan bir durumdur. Bu nadir sendrom çoğunlukla diyabetik üremili hastalarda saptanmaktadır. Burada 5 yıldır haftada 3 gün düzenli hemodiyaliz tedavisi alan, ani gelişen koreik hareket bozukluğu olan ve bilateral bazal ganglion lezyonları ile ilişkilendirdiğimiz bir erkek hastayı sunmaya çalıştık. Beyin manyetik rezonans (MR) incelemesinde bazal ganglion seviyesinde T1 sekansında izointens T2 ve FLAİR sekanslarında hiperintens görüntüler saptandı. Hastaya nöroleptik tedavi ile birlikte günlük hemodiyaliz tedavisi uygulandı. Yoğun hemodiyaliz tedavisi sonrası semptomlarında ve kontrol MR bulgularında belirgin gerileme saptandı. Bu lezyonların temel mekanizması metabolik ve/veya vasküler nedenlerle ilişkili olabilir. Diyabetik nefropatili hastalarda görülen akut koreik hareket bozukluğunun yoğun hemodiyaliz tedavisi ve kan glukoz regülasyonu ile düzeltilebileceği kanısındayız.

ANAHTAR SÖZCÜKLER: Bazal ganglion, Korea, Diabetes mellitus, Üremi

INTRODUCTION

Uremia is a clinical metabolic condition that develops as a result of renal dysfunction. Uremic encephalopathy occurs due to involvement of the brain cortex in case of acute or chronic renal failure. A movement disorder associated with reversible bilateral basal ganglion lesions was first defined by Wang et al. in 1998 (1) and it is considered to be a separate entity from uremic encephalopathy. This rare condition is observed increasingly in patients with diabetic nephropathy. Most of the patients diagnosed are Asians. Certain symptoms of parkinsonism such as bradykinesia, rigidity, tremor and dysarthria are frequently observed in these patients. Magnetic Resonance imaging (MRI) is frequently

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used in the diagnosis and typically shows transient edema in the basal ganglia. The definite cause of this transient condition is uncertain. We report a diabetic patient who was admitted to our hospital for generalized acute chorea while receiving hemodialysis treatment.

CASE PRESENTATION

A 52-year-old male patient had type 2 diabetes mellitus and hypertension for 15 years. He had been in the hemodialysis program 3 times a week for about 5 years. Generalized, involuntary, irregular, continuous, choreoathetotic movements had begun 2 days ago. These involuntary movements were particularly profound at the distal end of the extremities and in the peroral region. The involuntary movements disappeared during sleep, tended to decrease during rest and were aggravated during activity. He had no history of hypotension or hypoglycemia. No abnormality was detected in his neurological examination except for a decrease in deep tendon reflexes and chorea. Vital findings were normal and the following values were found in the laboratory examination: BUN 66.5 mg/dl, creatinine 12.5 mg/dl, glucose 174 mg/dl, Na 140 mmol/L, K 4.49 mmol/L, Ca 8.6 mg/dl, P 7.4 mg/dl, Cl 100 mmol/L, albumin 4.4 g/dl, HbA1c 8.9%, C-reactive protein 339.8 mg/L (N<5), ferritin 1541 ng/ml, and iPTH 54.76 pg/ml. Blood gas analysis revealed pH: 7.326, HCO_3: 18.4 mmol/L, lactate:1.9 mmol/L. Thyroid function tests were normal. Treatment with 1 mg/day of haloperidol was started and titrated to 2 mg/day gradually. The brain MRI taken 48 hours after beginning of symptoms of the patient (Figure 1) showed isointense lesions in T1 sequence and hyperintense lesions in T2 and FLAIR sequences in bilateral basal ganglia. There was bilateral signal intensity increase in the globus pallidus and putamen. Diffusion sequence showed ill-defined hyperintense areas in the putamen and thalamus. There was no other abnormality except for senile atrophy. The patient underwent daily hemodialysis. The choreic movements of the patient were significantly decreased within 72 hours after daily hemodialysis and haloperidol treatment, and completely disappeared at the end of the first week. The following values were found in the tests performed at week 1 following hospitalization: glucose 137 mg/dl, BUN 48 mg/dl, creatinine 8.3 mg/dl, Na 136 mmol/L, and K 4.6 mmol/L. His neurological findings were completely normal on day 10. A cranial MRI was obtained on day 90 for follow-up purposes and showed nearly complete regression of the basal ganglia lesions (Figure 1).

DISCUSSION

We presented a male patient with diabetes mellitus who received hemodialysis treatment 3 days a week for 5 years and suffered from choreic movements that developed suddenly and were associated with bilateral basal ganglia lesions. Uremic encephalopathy is an organic brain syndrome that is frequently associated with visual impairment, changes in consciousness with various severities from mild confusion to coma, tremor, asterixis, multifocal clonus, chorea and seizures (2). Short-term fluctuations may be observed in the findings and symptoms. Typically, the cerebral cortex is affected and basal ganglion

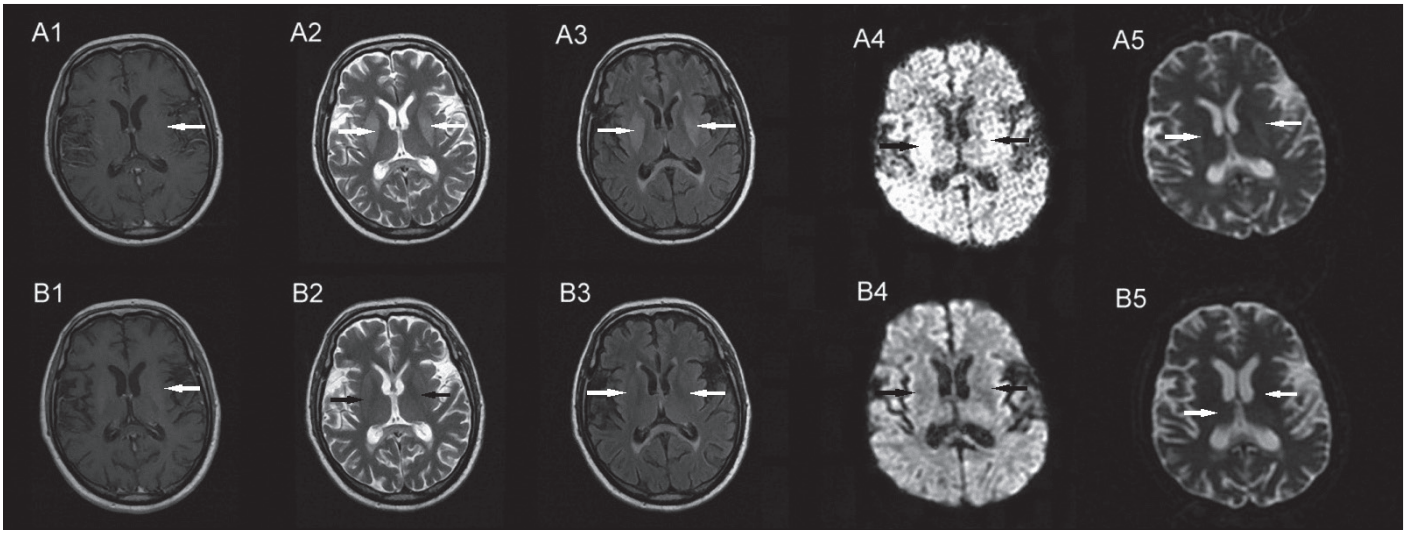

Figure 1: *1. T1-weighted image, 2. T2-weighted image, 3. Fluid-attenuated inversion recovery (FLAIR) image, 4. Diffusion-weighted image, 5. Apparent diffusion coefficient (ADC) map. Initial brain MR imaging demonstrating bilateral basal ganglia lesions (A1-5). Intense edema is seen in the basal ganglia indicated by arrows. These areas show hyperintensities on T2- and diffusion-weighted, and FLAIR images, while the same lesions show isointensity on the T1-weighted image. The corresponding areas have slightly increased signal intensities on the ADC map. No significant changes were observed in the other areas of the brain except minimal senile atrophy. Follow-up MR imaging (B 1-5) performed 3 months after onset shows decreased signal change and size of the focal edema in the basal ganglia compared with the previous study. Discrete symmetrical signal changes over bilateral globus pallidus structures are noted (arrows; hyperintensities on T2- weighted image and ADC map, and isointensities on T1-weighted, and FLAIR images).*

involvement is not frequent (3-5). Acute movement disorder accompanied by bilateral basal ganglia involvement in patients with diabetic uremia is a separate clinical syndrome (3). It was reported that this syndrome is most commonly observed in Asian patients. Patients are often admitted with symptoms of acute parkinsonism (bradykinesia, rigidity, postural instability, and gait disorder). Our patient did not show any parkinsonism symptom. Bilateral basal ganglion lesions are also observed in cases of carbon-monoxide intoxication, hypoxia, toxins, small vein vasculitis, and infections, but such clinical cases do not regress spontaneously (6,7). Our patient did not have hypotension, vasculitis or an active infection. Bilateral basal ganglia involvement may also occur in nonketotic hyperglycemia and it is observed as hyperdense lesions in cranial computed tomography (5). There was no significant hyperglycemia at the hospitalization of our patient and blood glucose levels were regulated during hospitalization with adjustment of insulin doses. Similar manifestation may be observed in myelinolysis which occurs as a result of rapid correction of hyponatremia, but our patient did not have a history of correction of hyponatremia. A similar manifestation may be caused by hypoxia, but our patient did not have hypoxemia. Methylguanidine, parathyroid hormone, aluminum, thiamine deficiency, and thromboembolic conditions are also held responsible for the occurrence of this picture (8).

It has been observed that most of the patients with uremic basal ganglia involvement are diabetics. Nearly 90% of these patients also have metabolic acidosis (9). Association of diabetes and uremia may exacerbate the negative effects of uremic toxins on basal ganglion cells based on diabetic microangiopathic damage or the compromised glucose metabolism (10). In patients with diabetic nephropathy, bilateral basal ganglia lesions show low signal intensity at the T1 sequence and increased signal intensity at the T2 sequence in MRI. Findings of cytotoxic edema surrounded by vasogenic edema are detected at the affected regions (11). Spontaneous improvement in baseline clinical symptoms and significant regression in imaging finding are typical. An F-18 fluorodeoxyglucose positron emission tomography (FDG-PET) study performed in 2 cases by Wang et al. demonstrated grossly decreased glucose metabolism in basal ganglia, especially in the putamen. They postulated that these patients already have compromised cellular function due to long-term diabetes through microangiopathic changes or energy utilization failure (3).

Our patient showed complete clinical recovery from the neurological disorder within 2 weeks after increasing the frequency of hemodialysis. Reports from the published literature show that the clinical recovery may be complete in 20% of the cases, partial with residual deficits in 50% of the cases, or absent in 30% of the cases. Radiological recovery is seen in 90% of the cases (5).

In conclusion, bilateral basal ganglion lesions develop as a result of the combination of deposition of uremic toxins and microvascular dysfunction in patients with diabetic uremia. Dysregulation of blood glucose due to receiving improper anti-diabetic treatment, and high creatinine levels as found in our patient suggest that the metabolic disorder and insufficient dialysis may play a role in occurrence of this syndrome. Hence, the complaints of our patient rapidly disappeared when he received daily dialysis and the blood urea nitrogen and creatinine levels regressed, and blood glucose levels were better regulated.

CONCLUSIONS

Choreic movement disorders may be seen especially in patients with diabetic nephropathy. We suggest that sufficient dialysis treatment should be performed and blood glucose should be better regulated in such patients. The patients should be carefully assessed about dialysis sufficiency and acid-base and electrolyte abnormalities.

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