Osmotic Demyelination Syndrome: Reversible MRI Findings in Bilateral Cortical Lesions

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Abstract

A 70-year-old man who suffered from osmotic demyelination syndrome (ODS) is presented. Dyspnea, pseudobulbar palsy and motor weakness were seen. MRI in the acute stage revealed focal abnormal high-signal lesions in the pons, thalamus and bilateral cortical areas on T1-weighted and FLAIR images. With corticosteroid therapy he recovered from his dyspnea and severe pseudobulbar palsy, and the spastic quadriplegia gradually improved. One year later the brain lesions had disappeared on T1-, T2-weighted and FLAIR images. To detect the cortical or subcortical lesions in ODS, FLAIR imaging should be performed routinely.

(Key words: osmotic demyelination syndrome, extrapontine myelinolysis, cortical laminar lesion, MRI, FLAIR)

Introduction

Osmotic demyelination syndrome (ODS) or central pontine myelinolysis (CPM) and extrapontine myelinolysis (EPM) are well-known demyelinating disorders. ODS is mainly seen following rapid correction of the serum sodium level in hyponatremic patients, especially in patients with underlying chronic alcoholism, malnutrition and chronic liver disease. The initial symptoms of ODS are mutism and pseudobulbar palsy (1) and the disorder is accompanied by well-defined MRI findings: acute demyelinating lesions are symmetric and hypointense on T1-weighted images, and become hyperintense on T2-weighted images in the subacute stage (2). However, cortical and/or subcortical MRI signal abnormalities have rarely been described in the literature (3, 4).

We report a patient with ODS who showed symmetrical cortical lesions with hyperintense signals on T1-weighted and fluid-attenuated inversion recovery (FLAIR) images, including serial MRI image changes in symmetric central pontine and extrapontine cortical lesions.

Case Report

A 70-year-old man experienced headache in December 1999. He underwent brain MRI and was discovered to have a pituitary adenoma at a neurosurgical hospital. He was treated by transnasal and sphenoidal sinus hypophysectomy at the same hospital on January 18, 2000. On the 7th postoperative day he became unsteady and his serum sodium (Na) concentration was 110 mmol/l (normal 136-145 mmol/l), potassium (K) was 4.9 mmol/l (normal 3.4-4.5 mmol/l), and chloride (Cl) was 75 mmol/l (normal 100-108 mmol/l). The post-operative thyroid stimulating hormone (TSH) concentration was 1.1 μIU/ml (normal; 0.4-3.7) and there was a low response to TRH-test. The adrenocorticotropin hormone (ACTH) concentration was 8.1 pg/ml (normal; 9.0-52.0) and serum concentration of cortisol was 8.0 μg/dl (5.0-15.0). His fasting blood glucose level was 70 mg/dl.

Physiological saline, 1,500 to 2,000 ml per day, was given intravenously for five days, and the serum sodium concentration had increased to 138 mmol/l on the 13th postoperative day. At this time he began to suffer from dysarthria and progressive weakness of the bilateral upper limbs. On the 18th postoperative day he was unable to raise his hands and he had dysarthria, accompanied by the onset of dysphagia. He was admitted to our hospital on February 21. He had a 2-year history of hypertension and prostatomegaly, but there was no history of liver disease or chronic alcoholism. There was no family history of neuromuscular disorders.

On admission he was alert and fully oriented. He was 160 cm in height and weighed 54 kg. Blood pressure was 136/90 mmHg, and pulse rate was 80 beats per minute. Physical findings were normal. Neurological examination revealed...
right nasal quadrantanomia due to the hypophysectomy, severe dysarthria, and dysphagia with diminished soft palate reflex. Motor weakness was more severe in the upper extremities than in the lower extremities, and he was unable to walk. There was no involuntary movement including tremor or choreic movement. The muscle tone of his upper extremities was severely spastic. All deep tendon reflexes were hyperactive, showing bilateral positive Babinski signs. There was no sensory disturbance. Cerebellar and autonomic functions were intact. A complete blood cell count and routine blood chemistry, including electrolytes and glucose, were normal (Na 141 mmol/l, K 4.1 mmol/l, Cl 104 mmol/l, glucose 90 mg/dl).

In the MRI findings on February 23, T1-weighted images demonstrated a linear high-signal lesion in the cortex of the precentral gyrus bilaterally (Fig. 1C, D). T2-weighted images showed high-signal lesions in the pons and bilateral thalamic areas (Fig. 1E, F). FLAIR images demonstrated clear high-signal lesions bilaterally in the thalamus and the precentral gyrus (Fig. 1J, K, L). Diffusion-weighted images showed high intensities only in the thalamic lesions. However, on T1-weighted post-gadolinium imaging no enhancement was seen in any of these lesions (not shown).

Since the patient was dyspneic and the pseudobulbar palsy was worsening, corticosteroid therapy was started; steroid pulse therapy consisted of an initial dose of methylprednisolone 1,000 mg/day three times followed by prednisolone tapering from 30 mg/day to 0 mg during the following 50 days. With these therapies, he recovered from his dyspnea and severe pseudobulbar palsy, and the spastic quadriplegia gradually improved. He was eventually able to raise his hands and walk without assistance. The pseudobulbar palsy greatly improved one month after admission. FLAIR images on March 15 demonstrated high-signal lesions in the central pons, which coincided with longitudinal pontine faciculi, in addition to bilateral thalamic and cortical

![Figure 1. Brain magnetic resonance imaging (MRI). 1.5 Tesla axial imaging. T1-weighted (A, B, C, D; TR/TE=590/15 ms), T2-weighted (E, F, G, H; TR/TE=3,200/96 ms) and FLAIR imaging (I, J, K, L; TR/TE=9,000/110 ms) T1-weighted images demonstrate a linear high-signal lesion in the cortex of the precentral gyrus bilaterally. T2-weighted images show high-signal lesions in the pons and bilateral thalamic areas. FLAIR images demonstrate faint high-signal lesions in the pons and clear high-signal lesions bilaterally in the thalamus and the precentral gyrus.](image)
Figure 2. FLAIR imaging on March 15, 2000 (A, B, C; TR/TE=10,002/144 ms) and May 17, 2000 (D, E, F; TR/TE=9,000/105 ms). The images on March 15 demonstrate high-signal lesions in the central pons, which coincide with longitudinal pontine fasciculi, in addition to bilateral thalamic and cortical linear lesions. All of these lesions did not appear on the MRI examination on May 17.

linear lesions (Fig. 2A, B, C). However, all of these lesions had disappeared and were not visible on the MRI examination in May (Fig. 2D, E, F). One year later he was readmitted to our hospital, but no spasticity of the extremities or pseudobulbar palsy was seen.

Discussion

In the present case, hyponatremia was caused by hypopituitarism after hypophysectomy, and the rapid correction of the hyponatremia is assumed to have led to ODS. Many causes of hyponatremia have been described: SIADH, hepatic diseases, malnutrition, and adrenocortical insufficiency, among others. Chronic alcoholism, rapid correction of hyponatremia, and liver transplantation are particular underlying and concomitant factors in patients with ODS (5).

The clinical picture of ODS is very variable (2, 5) but the initial symptoms consist of mutism, dysarthria and dysphagia. The present case showed dysarthria and dysphagia without mutism. Spastic quadriplegia is also seen in patients with ODS. The clinical picture in our patient was unusual in that motor weakness was dominant in the upper extremities. FLAIR images demonstrated clear high-signal lesions along the bilateral cortical areas of the precentral gyrus, areas which are anatomically responsible mainly for the control of the upper extremities: the neuroradiological finding of involvement of the primary motor cortex seemed to be consistent with the clinical feature of upperlimb weakness.

MRI is useful for detecting the area of demyelinating lesions (6). Acute demyelinating lesions are usually symmetric and hypointense on T1-weighted images, and become hyperintense on T2-weighted images during the subacute stage (5, 7, 8). Hyperintensity on diffusion-weighted images was reportedly detectable during 1 week after the onset of ODS symptoms (9). Calakos et al reported a rare patient who had T1-weighted images showing hyperintensity in cortical/subcortical regions, and gadolinium enhancement was seen (4). The cortical lesions in the present patient showed hyperintensity on T1-weighted images, but no gadolinium enhancement was seen in the acute stage. The lesions were not visualized by diffusion-weighted imaging. The histopathological findings of ODS-related cortical lesions were reported to include laminar necrosis, laminar demyelination with gliosis, or both (4). In our patient, MRI findings had completely resolved three months after onset and neurological disability was much improved. Both strongly suggest that most lesions including those with linear cortical distribution consist of a reversible demyelinating process.

FLAIR is an MRI technique that can clearly reveal lesions in the proximity to cerebrospinal fluid, such as cerebral cortical lesions (10). In the present patient this kind of MRI image was the most valuable for demonstrating the cortical
lesions ascribable to ODS. The presence of cortical/subcortical lesions has not received much attention in patients with ODS, possibly due to unsatisfactory MRI images. To detect the cortical or subcortical lesions in ODS, FLAIR imaging needs to be performed routinely.

References