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A Case of Autoimmune Thyroid Disease Presenting Posterior Reversible Encephalopathy Syndrome

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Abstract

A 40-year-old woman was admitted to our hospital with disturbance of consciousness and seizure. We diagnosed encephalopathy associated with autoimmune thyroid disease (EAATD). Fluid-attenuated inversion recovery and diffusion-weighted MRI demonstrated hyperintense lesions in the left occipitotemporal lobe on admission, but these findings disappeared on day 11 without neurological deficits, compatible with posterior reversible encephalopathy syndrome (PRES). We report here this case of autoimmune thyroid disease presenting as PRES.

Introduction

Non-specific neurological manifestations, such as disturbance of consciousness and seizures, are occasionally observed in patients with autoimmune thyroid diseases. These clinical conditions have recently been reported as encephalopathy associated with autoimmune thyroid disease (EAATD). There are a limited number of MRI reports on EAATD, which describe reversible or irreversible focal or diffuse hyperintense lesions in white matter, cerebellum, or brainstem on T2-weighted images.
When hyperintense lesions on T2-weighted MRI are observed in the posterior white matter in a patient with encephalopathy, posterior reversible encephalopathy syndrome (PRES) should be considered. Diffusion-weighted MRI (DWI) in patients with PRES often reveals isointensity with increased apparent diffusion coefficient (ADC) values.

Because our patient had lesions in the cerebral posterior region, we diagnosed PRES associated with EAATD. However, increased intensity of DWI and decreased ADC values were observed. Few reports have described the neuroradiological findings of PRES with acute exacerbation of EAATD. We report here a rare case of EAATD presenting with MRI findings of PRES.

Case report

A 40-year-old woman had a history of partial resection of the thyroid gland ten years previously for Graves' disease. Since then, she had on her own judgment not visited any hospital, and had thus not undergone periodic medical care. Two months before hospitalization, she had intermittently suffered periods of disturbance of sensation in her right arm and face, lasting 2 - 3 minutes. On the day of admission, she had noted speech impairment and memory deficits
from midday, and then experienced generalized seizure for the first time in her life. On examination, she exhibited incomplete right hemiparesis, disturbance of consciousness, and restlessness. She was intubated using diazepam, and then transferred to our hospital. On admission, her blood pressure was 180 / 70 mmHg, with a pulse of 110 per minute and a body temperature of 36.8 °C. On physical examination, bilateral exophthalmos and a surgical scar on the neck were noted. Neurological examination revealed acute confusional state with restlessness and roving eyes, but the pupils were normal in size and reactive to light. There was no facial asymmetry. The reaction to painful stimuli of her right upper and lower extremities was weaker than that of the left extremities. Muscle tone was normal. Deep tendon reflexes were not exaggerated. Plantar extensor responses were present bilaterally. Neck stiffness was not observed. Laboratory evaluation revealed hyperthyroidism with decreased thyroid stimulating hormone (TSH) of < 0.01 mIU/mL (normal range, 0.4 - 6.0), and increased free T4 of 1.83 ng/dL (0.8 - 1.6) and free T3 of 4.17 pg/dL (1.5 - 3.5). On extensive evaluation of the thyroid, anti-thyroperoxidase (TPO) antibody, anti-thyroglobulin antibody, TSH receptor antibody, and thyroid stimulating antibody levels were 4.3 IU/mL (< 0.3), < 0.3 IU/mL (< 0.3), 5.0 IU/L (< 1.0), and 168 % (< 180), respectively.
Markers of autoimmune diseases were negative, including normal antinuclear antibody, rheumatoid factor level, anti-double stranded DNA antibody, anti-SS-A antibody, anti-SS-B antibody, anticardiolipin antibody, myeloperoxidase anti-neutrophil cytoplasmic antibody, and proteinase-3 anti-neutrophil cytoplasmic antibody. No clotting abnormalities were detected. Cerebrospinal fluid (CSF) examination revealed 0.3 cells per microliter, glucose 60 mg/dL (50 - 75), protein 21 mg/dL (10 - 40), normal CSF IgG index, negative bacterial and fungal cultures, and an elevated lactic acid of 18.0 mg/dL (8.7 - 13.5) and pyruvic acid of 1.18 mg/dL (0.37 - 0.75). Electroencephalography (EEG) revealed sporadic diffuse and slow activity in the background. Neuroradiological studies were performed on admission. Brain CT demonstrated no abnormal structures or density. However, DWI findings revealed abnormalities including patchy hyperintense lesions in the left occipitotemporal grey and white matter (Figure 1, panel A). ADC values were lower in the hyperintense lesions on DWI than in the corresponding regions on the contralateral side ($0.48 \times 10^{-3}$ mm$^2$/s in left cortex vs $0.83 \times 10^{-3}$ mm$^2$/s in right cortex). On fluid-attenuated inversion recovery (FLAIR) images, the lesion exhibited slight high signal intensity (Figure 1, panel
B). No abnormal signal was detected in other portions of the brain. Cerebral angiography was performed on admission and revealed no stenotic lesions.

On hospital day 2, her blood pressure spontaneously decreased to 110 / 65 mmHg without antihypertensive therapy, and disturbance of consciousness and right hemiparesis disappeared. On hospital day 4, contrast-enhanced MRI revealed no abnormal enhancement. The hyperintense lesions on DWI and FLAIR and decreases in ADC values completely disappeared on day 11 (Figure 2, panels A and B). This resolution of neuroradiological findings appeared to represent PRES, since the abnormalities in MRI findings in occipitotemporal white and grey matter disappeared in parallel with improvement of symptoms.

Although we performed muscle biopsy to rule out mitochondrial myopathy, encephalopathy, lactic acidosis, and stroke-like episodes (MELAS), we found only mild infiltration by lymphocytes around the venules and muscle fibers in the specimens. Ragged red fibers, a finding specific for MELAS, were not detected. Finally, we diagnosed EAATD, since there were no other causes of encephalopathy and anti-TPO antibody was positive. Because her symptoms had disappeared, corticosteroid treatment was not performed. She was treated
with mercaptomethylimidazole, and the sensory disturbance in her right arm gradually diminished. She was discharged without neurological deficit on day 30.

**Discussion**

This patient with Graves’ disease exhibited subacute encephalopathy and positive anti-TPO antibody. FLAIR imaging demonstrated transient hyperintense lesions in the left occipitotemporal hemisphere. This finding is similar to those noted in previous reports of EAATD. The cerebellar hemisphere, temporal lobe, brainstem, and hippocampus were also involved in some cases. In a limited number of the cases of EAATD, cortical involvement was observed, as in our case. It has been suggested that these signal abnormalities reflect either edema or inflammation.

Both the neurological manifestations and hyperintense lesions on FLAIR imaging in our patient resolved during hospitalization. Our findings for the most part concur with those noted in Kato’s report of an EAATD case accompanied by the neuroradiological features of PRES. It is clear that PRES is a leukoencephalopathy clinically characterized by headache, altered mental status, visual loss, and seizures. In typical cases of PRES, neuroimaging
demonstrates bilateral subcortical and cortical edema with a predominantly posterior distribution on MRI and CT. PRES includes a large number of diseases and conditions with various mechanisms, such as hypertension, use of immunosuppressant drug, and hypercalcemia, though its exact pathophysiology remains unclear.

Kato et al. noted that the initial hyperintense lesions on DWI disappeared on follow-up MRI study and that elevated ADC values normalized in a patient with Hashimoto’s disease. Most patients with PRES, such as the one in Kato’s report, have lesions with unrestricted diffusion and increased ADC values indicating vasogenic edema. However, our patient had decreased ADC values in affected DWI lesions, as in previous cases of PRES. These findings raise the possibility that brain tissue injury due to autoimmune disorder in Graves’ disease may contribute to various changes in ADC values noted on initial MRI studies.

In conclusion, we have reported a case of EAATD presenting with PRES. This report provides new and important information on neuroimaging findings of EAATD.
Reference


Figure legends

Figure 1.

(A) Diffusion-weighted MR image and (B) fluid-attenuated inversion recovery image reveal cortical and subcortical hyperintense lesions in the left occipitotemporal lobe.
Figure 2.

(A) Diffusion-weighted MR image and (B) fluid-attenuated inversion recovery image demonstrate no lesions of abnormal intensity 11 days after onset.