Histologic Evidence of Reversible Posterior Leukoencephalopathy Syndrome

To the Editor: In 1996 Hinchey et al (1) described 15 patients who had subcortical edema without cerebral infarction. The lesion was found mainly in the posterior cerebral hemisphere, and both the clinical and radiological manifestation were reversible. Hinchey and others (2) discussed the pathophysiologic aspects of this condition, and noted its similarity to hypertensive encephalopathy, cyclosporine encephalopathy and human immunodeficiency virus (HIV) encephalopathy. However, because this condition is reversible, autopsy has not been reported. Here we report an autopsy case.

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The patient was a 49-year-old woman with a diagnosis of progressive systemic sclerosis who died of an unrelated cause 10 months after she presented with this syndrome. She developed stiffness of fingers on both hands, Raynaud’s phenomenon, nonproductive cough and weight loss of 14 kg in 5 months. A positive test for anti-nuclear antibody, the results of skin biopsy and chest X-ray confirmed the diagnosis of progressive systemic sclerosis-associated pulmonary fibrosis. The pulmonary lesion worsened, and she was admitted to our hospital in May 1991. On August 22, during her first hospitalization, she had an abrupt episode of delirium and a visual field defect. A computed tomography (CT) scan of the brain at that time was negative, and 4 days later it showed low-density areas in the occipital and temporal regions bilaterally (Fig. 1A). A single photon emission CT with 123I-IMP showed perfusion defect in those areas. However, a brain CT on September 18 returned to normal (Fig. 1B). The clinical manifestation also disappeared. In the light of the report by Hinchey et al, we believe that this was a case of reversible posterior leukoencephalopathy syndrome. The patient was admitted to the hospital again on June 30, 1992, because of severe abdominal pain. A roentgenogram showed free air in abdominal cavity. Perforation of the colon was suspected and a laparotomy was done. The patient’s condition worsened postoperatively, and she died of respiratory failure on July 14, 1992.

Autopsy was done. The findings confirmed pneumatosis cystoides intestinalis, progressive systemic sclerosis, and interstitial pneumonia associated with honeycomb change. Macroscopic examination of the brain showed no remarkable changes. There was no evidence of cerebral infarction. Microscopical examination showed mild astrocytosis in both parietal cortices. Astrocytosis is nonspecific; it can occur in Creutzfeldt-Jakob disease, acquired immunodeficiency syndrome (AIDS) encephalopathy, multiple-system atrophy, motor neuron disease and metabolic encephalopathy. Although more cases need to be examined, this provides histologic evidence of the reversibility of the posterior leukoencephalopathy syndrome.

Sadayoshi Ohbu, MD, Toshimasa Uekusa, MD*, Fumihiko Watanabe, MD** and Naohiko Chohnabayashi, MD

The Departments of Internal Medicine, Pathology*, and Radiology**, St. Luke’s International Hospital, 9-1 Akashi-cho, Chuo-ku, Tokyo 104-8560
Reprint requests should be addressed to Dr. Sadayoshi Ohbu, Director of Clinic, Mitsui Bussan Clinic, Mitsui & Co., Ltd., 2-1, Ohtemachi 1-chome, Chiyoda-ku, Tokyo 100-0004

References


Figure 1. (A, B) CT scan of the brain.