Acute Wernicke’s Encephalopathy: Comparison of Magnetic Resonance Images and Autopsy Findings

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This is the first report of acute Wernicke’s encephalopathy in which characteristic magnetic resonance (MR) findings have been verified by postmortem histopathological examination. The high-signal areas surrounding the third and the fourth ventricles and the aqueduct on the T2-weighted images reflected the spongy disintegration of the neuropil, which is the typical pathological finding in Wernicke’s encephalopathy. We confirmed that MR imaging is essential to the early diagnosis of Wernicke’s encephalopathy.

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Introduction

Wernicke’s encephalopathy (WE) is a neurologic disorder caused by a nutritional deficiency of thiamine. It mainly affects the mamillary bodies and periventricular regions around the third and fourth ventricles and aqueduct (1). Although magnetic resonance (MR) studies show characteristic findings, there have been no reports of precise comparison between MR images and postmortem histopathological findings in the acute phase of WE. We present a case of acute WE and correlate the MR findings with the postmortem findings.

Case Report

A 54-year-old man was admitted to our hospital in a comatose state. On admission he was undernourished; he had no history of alcohol abuse. Body temperature was 36.0°C, and blood pressure was 120/60 mmHg, with a regular pulse of 80 beats per minute. He could not move his legs except in response to painful stimuli. Neurological examination showed the pupils to be equally constricted, with sluggish reaction to light. Both eyes were in the mid-line position.

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Oculocephalic reflex was absent. Involuntary movements in the form of fine tremors were observed from the neck to the right arm. Deep tendon reflexes were absent in all extremities, and the plantar responses were flexor. There was no nuchal rigidity. Laboratory data revealed leukocytosis (WBC: 17,000/mm³) and an increase in C-reactive protein (9 mg/dl). Liver and kidney function was normal. Blood glucose and ammonia levels were normal. Blood gas analysis, a chest X-ray and electrocardiogram also yielded normal findings. The brain CT images revealed only cerebral atrophy, with no focal abnormal densities. Cerebrospinal fluid examination showed an increase in cell count (12/μl; neutrophils, 3/μl; lymphocytes, 9/μl) and protein concentration (8.9 mg/dl), but bacteriological and cytological examinations were negative. Since these findings suggested metabolic coma or encephalitis of unknown origin. Drip infusion of cefmetazole, acyclovir and amino acids was started. A 100 mg dose of thiamine chloride was also intravenously administered on the day of admission. Nevertheless, the patient’s pulse and respiration became irregular.

On the 3rd hospital day, brain MR imaging was performed. The MR images were produced with a 1.5T system (Siemens, Magnetom Vision, Erlangen, Germany). The brain imaging protocol included T1-weighed (spine echo 600/14) and T2-weighted (turbo spine echo 2,500/85) sequences in the transaxial plane. Transaxial images were obtained with a 7.0-mm section thickness with a 2.0-mm inter-section gap. T2-weighted turbo spin echo images showed bilaterally symmetrical high-signal areas surrounding the aqueduct and the third and fourth ventricles (Fig. 1). These findings were typical of WE; thiamine chloride (300 mg daily, i.v.) was added to the treatment regimen. His pulse and respiration improved temporarily, but multiple organ failure developed, and he died on the 5th hospital day. Subsequent laboratory data also supported the
diagnosis of WE: the serum level of vitamin B1 was 3.3 μg/dl (normal range: 4.4–11.2 μg/dl) and transketolase activity was 0.59 IU/gHb (normal range: 0.75–1.30 IU/gHb) after the injection of 100 mg of thiamine chloride on admission.

Postmortem examination was performed two hours after the patient's death. The brain weighed 1,080 g and showed mild cerebellar atrophy. Transaxial section of the brain revealed small brownish spots and edematous change surrounding the third and fourth ventricles and the aqueduct (Fig. 2), but these findings were not detected in the mamillary bodies. Microscopic examination revealed marked spongy disintegration of the neuropil and neuron sparing (Fig. 3). Swelling of capillary endothelial cells and extravasation of red blood cells were also observed. There was no evidence of gliosis. These findings were compatible with the diagnosis of acute WE.

**Discussion**

WE is a neurologic disorder with an acute onset, characterized by the triad of ocular abnormalities, ataxia, and a global confusional state (2, 3). Diagnosis of WE is difficult, especially when the patient is in a comatose state, and is non-alcoholic. Lindboe and Løberg (4) reported 18 autopsy cases of acute WE. Seven of the cases (39%) were non-alcoholic, and none of these
seven had been diagnosed as WE clinically.

Some reports have described low-density areas in thalamic or brain stem lesions on CT (5–7), but CT is usually of little help in the diagnosis of WE, as was true in the present case.

The MR findings of WE have been reported by several investigators (8–11). High-intensity areas surrounding the third and fourth ventricles and the aqueduct on T2-weighted images are typical and useful in correctly diagnosing acute WE, and we were able to diagnose WE antemortem based on these MR findings in the present patient.

Gallucci et al (9) reported that the high signals on T2-weighted images represent pathologically non-specific findings, such as edema, demyelination and gliosis. However, their symmetrical distribution allows differential diagnosis of WE from other diseases.

No previous reports have confirmed these MR findings of acute WE by pathological examinations. This is the first report verifying MR findings by autopsy. In the present case, MR imaging was performed 48 hours before the postmortem examination. Comparison of the MR images with the pathological findings showed that the high signals seen on the T2-weighted images corresponded to edematous change induced by spongy disintegration of the neuropil. Torvik (12) described the histological characteristics of WE as consisting of progressive destruction of the neuropil with severe capillary endothelial swelling and definite sparing of the neurons in the mamillary bodies and the subependymal structures along the third and fourth ventricles and the aqueduct. The pathological findings in our case were consistent with his description. These edematous changes caused elongation of T2 relaxation time, resulting in high signals on the T2-weighted images.

Although the mild changes in the mamillary bodies were unusual, no abnormal signals were detected in the mamillary bodies on the MR images. This consistency demonstrated that MR images correctly reflect the pathological changes in the brain parenchyma.

In some reports, the mamillary bodies have been enhanced on MR images in acute WE (13, 14). If our patient had lived longer, such findings in the mamillary bodies might have become apparent.

Our comparisons with the autopsy findings led us to conclude that the MR findings of acute WE reflected edematous change induced by the spongy disintegration of the neuropil. The definite findings compatible with acute WE on MR images make it possible to establish the diagnosis early and to start treatment promptly, even in cases which are confusing clinically. MR imaging should be performed immediately whenever a diagnosis of WE is suspected.

References

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