Possible Acute Hemorrhagic Leukoencephalitis Manifesting as Intracerebral Hemorrhage on Computed Tomography

—Case Report—

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Abstract

A 15-year-old girl presented with meningeal irritation and bilateral cerebral signs after contracting influenza. A lumbar puncture revealed bloody cerebrospinal fluid and polymorphonuclear predominant pleocytosis with an elevated protein level and normal glucose level. Computed tomography showed a hematoma in the right basal ganglia and lateral ventricles. Symmetrical low density areas were also noted in the bilateral white matter. The preliminary diagnosis was hemorrhagic cerebrovascular disease of unknown cause. However, her neurological condition deteriorated. Magnetic resonance (MR) imaging showed diffuse high intensity signals in the bilateral white matter and small spotty lesions, indicating hemorrhages in various stages. The final diagnosis was acute hemorrhagic leukoencephalitis (AHL). However, high-dose steroid administration and plasmapheresis failed to improve her condition. Hypothermia could not control her intracranial pressure and she died 12 days after admission. The neuroimaging findings indicated the histological characteristics of AHL, but the hematoma formation is rare. AHL is a fulminant form of brain demyelination and can be fatal, so early diagnosis and aggressive treatment are important for successful recovery. Therefore, early investigation by MR imaging is necessary.

Key words: acute hemorrhagic leukoencephalitis, intracerebral hemorrhage, computed tomography, magnetic resonance imaging

Introduction

Acute hemorrhagic leukoencephalitis (AHL) is a fulminant, particularly severe form of acute demyelinating encephalomyelitis and can cause death within a week after onset. Therefore, early diagnosis and aggressive treatment are extremely important. AHL is often preceded by an upper respiratory infection or several viral diseases and autoimmune mechanisms may be involved in the pathogenesis. Here, we describe a case of AHL manifesting as massive hemorrhage, but atypical radiological findings caused confusion in the early diagnosis, resulting in a delay in aggressive treatment and an unfavorable outcome.

Case Report

A 15-year-old girl was admitted to the previous hospital because of fever and headache. The patient had been well until 1 week earlier, when symptoms of influenza and low-grade fever had developed. On admission, she showed signs of meningeal irritation. The preliminary diagnosis was meningitis. A lumbar puncture performed on the same day revealed bloody cerebrospinal fluid (CSF), and an opening pressure of 140 mmH2O, white blood cell count of 232/mm3 (71% segmented forms), glucose level of 31 mg/dl, protein level of 75 mg/dl, and negative bacterial culture. Computed tomography (CT) on the next day showed a hematoma in the right basal ganglia and lateral ventricles. Symmetrical low density areas were also noted in the bilateral white matter (Fig. 1). Based on the CT findings, she was treated under a diagnosis of hemorrhagic cerebrovascular disease of unknown cause. However, her neurological condition deteriorated and she developed right hemiparesis and involuntary movement in the right upper extremity. Therefore, she was transferred to our department for further
Fig. 1 Computed tomography scans showing a hematoma in the right basal ganglia and lateral ventricles. Symmetrical low density areas in the bilateral white matter can be seen.

Fig. 2 T2-weighted magnetic resonance images showing symmetrical diffuse high intensity signals in the bilateral white matter and patchy low or high intensity lesions, as well as the hematoma in the right caudate nucleus.

Fig. 3 Computed tomography scans taken 5 days after admission showing diffuse low density areas in the white matter, including the entire cerebrum and cerebellum. The hematoma in the right caudate nucleus has become obscure.

On admission, she was alert and complained of headache. She had a body temperature of 37.5°C and showed signs of meningeal irritation. Neurological examination revealed right hemiparesis with occasional athetotic involuntary movement in the right upper extremity, but the pathological reflexes were positive on both sides. Peripheral blood analysis revealed elevated white blood cell count (13300/mm³ with predominant neutrophils), but otherwise no abnormalities, including the viral titer for herpes simplex. Cerebral angiography revealed no abnormalities. Magnetic resonance (MR) imaging showed the previously observed hematoma, diffuse high intensity signals in the bilateral white matter, and small spotty lesions, indicating hemorrhages in various stages (Fig. 2). Administration of gadolinium-diethylenetriaminepenta-acetic acid caused no enhancement.

Our diagnosis was AHL based on the clinical course, CSF data, and MR imaging findings. However, high-dose steroid administration and plasmapheresis failed to improve her condition. After admission, she became drowsy and developed decorticate posture 2 days later. Five days after admission, she became comatose. CT revealed an obscured hematoma and diffuse low density areas in the white matter, including the entire cerebrum and cerebellum (Fig. 3). Hypothermia could not control the intracranial pressure and brain death occurred 12 days after admission. No autopsy was permitted.

Discussion

The present patient showed several clinical and laboratory features supporting the diagnosis of AHL, including preceding influenza, fever with acute encephalopathy with bilateral cerebral signs, CSF polymorphonuclear predominant pleocytosis with an elevated protein level and normal glucose level, and polymorphonuclear predominant peripheral leukocytosis. The clinical course was also compatible with AHL. However, no histological diagnosis could be obtained.

The histological characteristics of AHL are con-
AHL Manifesting as Hematoma

considered to be necrotizing angitis of the venules and capillaries of the white matter, causing ball and ring hemorrhages around the affected vessels with fibrinoid necrosis and perivascular leukocytic exudates.\textsuperscript{1,2,7,10} Our MR imaging findings indicated these histological characteristics as diffuse non-enhanced high intensity signals in the white matter and multiple patchy lesions.\textsuperscript{3,7,8} AHL might also cause macroscopic hemorrhage, although the exact mechanism of hematoma formation remains uncertain.\textsuperscript{1} However, such hematoma formation is rare and there are no reports of radiological findings with similar temporal profiles to the present case. In our case, the hematoma rapidly became obscure on CT. This rapid hematoma absorption is different from that of an ordinary intracerebral hemorrhage and might be characteristic of AHL.

The first diagnosis was cerebrovascular disease based on the initial CT findings. However, the symmetrical low density areas in the bilateral white matter, notable on CT, might indicate AHL, excluding the hematoma formation. Neuroimaging findings of AHL can be detected within 3 days after clinical onset,\textsuperscript{7} so early investigation by MR imaging would avoid any delay in aggressive treatment.

References

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