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Reversible MR Findings of Hemolytic Uremic Syndrome with Mild Encephalopathy

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Summary: We report the reversible MR findings in a 7-year-old girl with hemolytic uremic syndrome and mild encephalopathy. The splenium of the corpus callosum showed isointense to low signal intensity on T1-weighted images and high signal intensity on T2-weighted images, representing local edema. These findings returned to near normal on MR images obtained 1 week later. The patient recovered without CNS impairment.

CNS complications have been reported as a significant mortality risk factor in patients with hemolytic uremic syndrome (1). While evaluation of cerebral organic changes by imaging is necessary to clarify the pathophysiology of encephalopathy related to this disorder, few reports have mentioned the associated MR findings (2–4). We present a case of hemolytic uremic syndrome with mild encephalopathy in which local reversible changes in the splenium of the corpus callosum were shown on MR images.

Case Report

A 7-year-old girl with hemolytic uremic syndrome caused by enteritis stemming from infestation by O-157 Escherichia coli was admitted from a local hospital on the second day after the onset of illness. On admission, blood and urine data showed severe hemolytic anemia (hemoglobin, 7.2 g/dL; hematocrit, 20.0%; lactate dehydrogenase, 1896 U/L in blood), thrombocytopenia (platelets, 10,000/μL), and acute oligoanuric renal failure (creatinine, 0.9 mg/dL; BUN, 29 mg/dL; free hemoglobin in urine, 74.8 mg/dL; albumin in urine, 3540 mg/L). Hypertension (144/86 mm Hg) was also noted. Neurologic examination showed normal results except for mild encephalopathy manifested by disorientation and hallucinations for a period of 7 days after admission. On the third day, MR studies revealed remarkable changes in the splenium of the corpus callosum with isointense to low signal intensity on T1-weighted images and high signal intensity on T2-weighted images (Fig 1A and B). These findings were not detected on cranial CT scans obtained the same day. There was no evidence of infarction, hemorrhage, or global cerebral swelling. The patient received supportive therapy with haptoglobin, gamma globulin, transfusions, a diuretic, and a calcium antagonist. The severe intravascular hemolysis gradually improved beginning the sixth day after admission, and the CNS symptoms completely disappeared by the seventh day. A follow-up MR examination on the 10th day showed a faint increase in signal on T2-weighted images in the splenium of the corpus callosum (Fig 1C). On the 17th day, MR findings returned to normal. The patient was discharged from the hospital on the 40th day without CNS impairment.

Discussion

Hemolytic uremic syndrome caused by O-157 E. coli enteritis is a general microangiopathic disorder, which potentially causes fatal CNS complications in infants and children. Clinical CNS involvement is represented in the form of disattention, disorientation, hallucinations, alterations of consciousness (such as coma or stupor), focal and generalized seizures, cortical blindness, hemiplegia, and decerebrated posturing. In a study by Hahn et al (5), 16 of 78 children with identified hemolytic uremic syndrome had neurologic manifestations, and five of these died of CNS complications. Autopsies revealed evidence of focal ischemia with large nonhemorrhagic or hemorrhagic infarcts.

Cranial CT findings of hemolytic uremic syndrome include cortical infarction with hemorrhage, extensive blood-brain barrier disturbance at the gray-white matter interface, and diffuse cerebral edema (5, 6). Few reports have mentioned the MR findings of associated encephalopathy (2–4). DiMario et al (2) found increased signal intensity on T1- and T2-weighted images in the caudate, putamen, and globus pallidus, which they ascribed to subacute hemorrhagic infarction. Sherwood et al (3) reported the MR findings of bilateral lesions in the basal ganglia that had increased signal intensity on T1- and T2-weighted images in the caudate, putamen, and globus pallidus, which they ascribed to subacute hemorrhagic infarction. Sherwood et al (3) reported the MR findings of bilateral lesions in the basal ganglia that had increased signal intensity on T1- and T2-weighted images in the caudate, putamen, and globus pallidus, which they ascribed to subacute hemorrhagic infarction. Jeong et al (4) found abnormally high signal intensity on T1- and T2-weighted MR images in the lentiform nucleus, the posterior limb of the internal capsule, the external capsule bilaterally, and the left extreme capsule, which indicated infarct with focal hemorrhage.

In our patient, MR findings in the splenium of the...
corpus callosum were reversible, which was thought to represent transient local edema formation with no evidence of hemorrhage or infarction. These changes were not detected on CT scans. The reason for these local cerebral changes is not clear. It is unlikely that the changes were related to any therapy given to the patient or that hypertension, as a common symptom of hemolytic uremic syndrome, contributed to the findings in the corpus callosum. One possible explanation is that verotoxin from O-157 E. coli binds to a specific area of the cerebral vascular endothelium, inducing localized microvascular angioathect and formation of perivascular edema (7–9).

The distribution of verotoxin and verotoxin receptors in the brain has not been thoroughly clarified. In a mouse model administered verotoxin-2, immunoreactivity of the toxin was reported to be localized in damaged myelin sheaths of neuron fibers accompanied by edematous axons in the brain cortex (7). In addition, in a rabbit model challenged with verotoxin-1, CNS lesions were reported to include massive and diffuse pericellular and perivascular edema, associated with endothelial cell swelling and mild thickening of capillary walls in the brain and brain stem (8). Specific binding of verotoxin to the vascular endothelium has also been reported to cause direct toxin-mediated injury of the tissue (9). These reports may support the hypothesis that verotoxin binding caused the diffuse axonal damage associated with edema formation in the splenium of the corpus callosum in our patient.

Summary

Reversible MR findings in a patient with hemolytic uremic syndrome and mild encephalopathy included isointense to low signal intensity on T1-weighted images and high signal intensity on T2-weighted images in the splenium of the corpus callosum. These changes may have represented local edema formation, which reflected the binding of verotoxin to a specific area of the brain.

References