

성인 용혈성 요독 증후군 환자에서 발생한 후두부 가역적 뇌병증 증후군

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Posterior Reversible Encephalopathy Syndrome in an Adult Patient with Hemolytic Uremic Syndrome

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Introduction: Posterior reversible encephalopathy syndrome (PRES) is a clinical condition characterized by an acute or subacute onset of neurological symptoms, such as visual disturbance and seizures in association with imaging of the brain. Although rare causes of PRES include thrombotic thrombocytopenic purpura, it has not yet been described in adult patients with hemolytic uremic syndrome (HUS).

Case: A 45-year-old woman without remarkable past medical history presented with a 1-week history of abdominal pain and watery diarrhea. 1 day before onset of her symptoms, she had consumed cooked pork and rice. Her laboratory results on admission showed features of acute renal failure due to HUS with normal level of ADAMTS-13 activity. In addition to plasmapheresis, she required continuous renal replacement therapy due to anuria and progressive azotemia. On hospital day 7, she developed the high blood pressure 155/95 mmHg, followed by a generalized tonic-clonic seizure and decreased mentality. The magnetic resonance imaging (MRI) of the brain showed that the patient had T2 high signal intensities and swelling in the areas of gyri at both the superior frontal lobes and the parieto-occipital lobes, which findings were consistent with PRES. Over the next 7 days, an improvement of confusion and attentional difficulties was observed. Within 1 month, renal replacement therapy was discontinued as her urine output continued to rise. The follow-up MRI five weeks after initial presentation showed the corresponding lesions with abnormal signal intensities were resolved.

Conclusion: This case report shows the uncommon development of PRES in an adult HUS patient, which was confirmed by serial MR studies. Although the pathogenesis responsible for PRES remains unclear and controversial, HUS-related endothelial dysfunction might provide a clue to the pathophysiology.

Key Words: 용혈성요독증후군, 후두부 가역적 뇌병증 증후군, 급성신부전
HUS, Posterior reversible encephalopathy syndrome, ARF