Atypical hemolytic uremic syndrome led to AKI in a hemophagocytic lymphohistiocytosis patient

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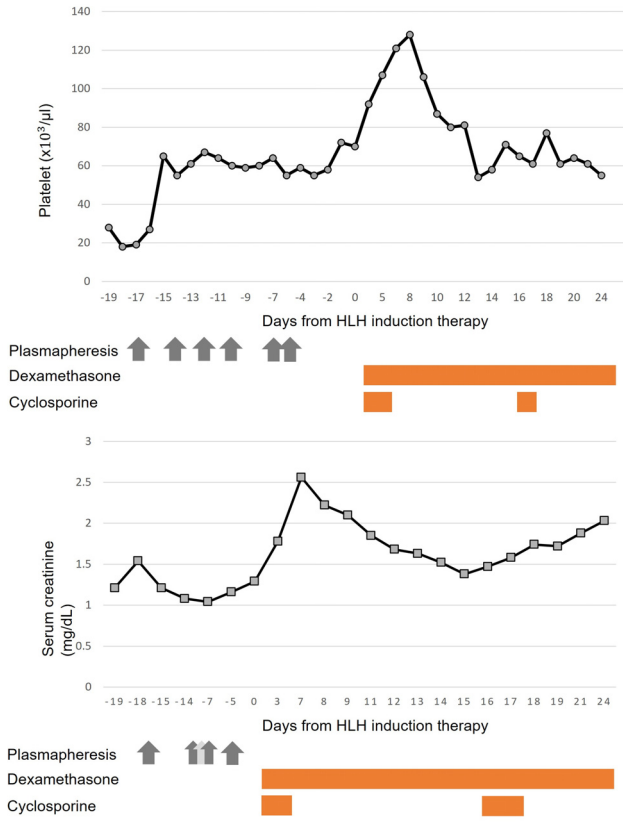
Hemophagocytic lymphohistiocytosis (HLH) is a fatal clinical syndrome frequently complicated by acute kidney injury (AKI). Atypical hemolytic uremic syndrome (aHUS) is also serious disease that manifests hemolytic anemia, thrombocytopenia, and AKI. There are only few reports on aHUS developing in patient with HLH. We reported a rare case of aHUS associated with HLH. A 53-year-old woman was admitted four times for hematemesis. She had no specific underlying disease and medication history. At the time of admission, the patient presented with AKI, anemia and thrombocytopenia. She had schistocytes on the peripheral blood smear, increased LDH, and normal ADAMTS 13 level. Based on these findings, aHUS was diagnosed. We initiated plasmapheresis using FFP. At 2nd and 3rd admission, renal function and thrombocytopenia improved after 3 plasmapheresis therapy. However, at 4th admission, thrombocytopenia persisted after 6 plasmapheresis. Serum ferritin, CRP and AST were increased. Additionally, the patient showed elevated LDH, elevated soluble IL-2 receptor, reduced NK cell activity, elevated TG and low fibrinogen. A blood smear was negative for schistocytes, and abdominal ultrasound revealed moderate splenomegaly. And the results of the autoimmune panel and virus serologic tests were negative. Finally, the patient was diagnosed with HLH based on splenomegaly, anemia and thrombocytopenia, hypofibrinogenemia, low NK cell activity, high ferritin and negative results on viral and autoimmune marker studies. Following HLH diagnosis, dexamethasone and cyclosporin were administered according to HLH-2004 protocol. On the 9st days after treatment, hemoglobin, platelet level and renal function were gradually improved. On the 22st days after treatment, the patient developed septic shock with respiratory failure, and was admitted to the ICU for mechanical ventilation, CRRT and VV ECMO support. Finally, the patient expired due to invasive aspergillosis at the 46 days after treatment. HLH is a severe condition and mortality can be reduced by early diagnosis and correct treatment. In patients with anemia, thrombocytopenia and AKI, the possibility of aHUS and HLH should be considered.



Complete blood count	
Hemoglobin (g/dL)	6.0
White blood cell (/μL)	4010
Platelet (/μL)	28000
C-Reactive protein (CRP) (mg/L)	6.26
Liver function test	
Aspartate transaminase (AST) (IU/L)	173
Alanine aminotransferase (ALT) (IU/L)	13
Total bilirubin (mg/dL)	2.90
Blood urea nitrogen (BUN) (mg/dL)	19.4
Creatinine (mg/dL)	1.21
Electrolytes (Na-K-Cl) (mmol/L)	141-41-96
Lactic acid (mmol/L)	14.1
Coombs' test (direct / indirect)	Negative / Negative
LDH (IU/L)	426
ADAMTS 13 activity (ELISA) (%)	42.2
Stool multiplex RT-PCR	Negative
NK cell activity (pg/mL)	<40
Soluble interleukin2 receptor (U/mL)	1045
Ferritin (ng/mL)	3419
Fibrinogen (mg/dL)	154