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Atypical hemolytic uremic syndrome triggered by acute pancreatitis

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Dear Editor,

The patient was a 72-year-old male with a history of untreated gallstones. He came to the hospital due to abdominal pain lasting for one day. An abdominal CT scan revealed acute pancreatitis and gallstones (Fig.1A). Preliminary diagnosis indicated mild acute gallstone-pancreatitis[1], After 12 hours of routine treatment, the patient developed hemoglobinuria and hematochezia. Further examination revealed acute kidney injury, hemolytic anemia (with elevated total and indirect bilirubin levels), thrombocytopenia, and severe coagulation dysfunction. Initiate plasma exchange (PE) using fresh frozen plasma immediately (3000ml/session, once daily). Furthermore, the peripheral blood smears showed fragmented erythrocytes, elevated LDH, decreased C3 levels, normal C4 levels, Stool culture and Coombs test results were negative, and ADAMTS13 activity was 60.5%. The final diagnosis was atypical hemolytic uremic syndrome (aHUS) caused by acute pancreatitis. After three

PE sessions, his condition gradually improved, and coagulation normalized. However, due to financial constraints, he discontinued PE and eculizumab, opting for hemodialysis instead, eventually, the patient progressed to chronic kidney failure. Figure 1B illustrates the biochemical and hematological changes during treatment, and the timing of PE.

Thrombotic microangiopathy (TMA) is a group of acute clinical syndromes characterized by hemolytic anemia, consumptive thrombocytopenia, and microvascular thrombosis, and the kidney is the most commonly affected organ. AHUS is one of the TMAs, with triggers for activation of the complement alternative pathway—such as infection and pregnancy—present in 70-80% of patients. It should be distinguished from typical hemolytic uremic syndrome (HUS), thrombotic thrombocytopenic purpura (TTP), and disseminated intravascular coagulation (DIC)[2]. The pancreas acts as both a target and effector in inflammation, with complement cascade interactions playing a key role. Complement activation triggers acinar cell damage in early pancreatitis, leading to autodigestion by pancreatic enzymes, which further cleave and activate complement factors[3]. Therefore, pancreatitis can cause complement system dysfunction, leading to aHUS. Although rare, similar cases have been reported, in this case, the patient developed hemoglobinuria about 36 hours after abdominal pain onset, aligning with prior findings [4]. Specific treatments for aHUS include eculizumab and PE. Eculizumab inhibits the complement pathway, thereby reducing endothelial damage, thrombosis, and kidney injury. PE removes defective mutant complement proteins and autoantibodies, and it also avoids volume overload in patients with kidney injury. Before eculizumab became available for treating aHUS, PE was the preferred treatment[5].

In this case, the patient improved significantly after PE, but insufficient treatment led to serious sequelae. Thus, hemodialysis cannot replace PE in aHUS patients, and early, accurate, and adequate treatment is crucial for prognosis.

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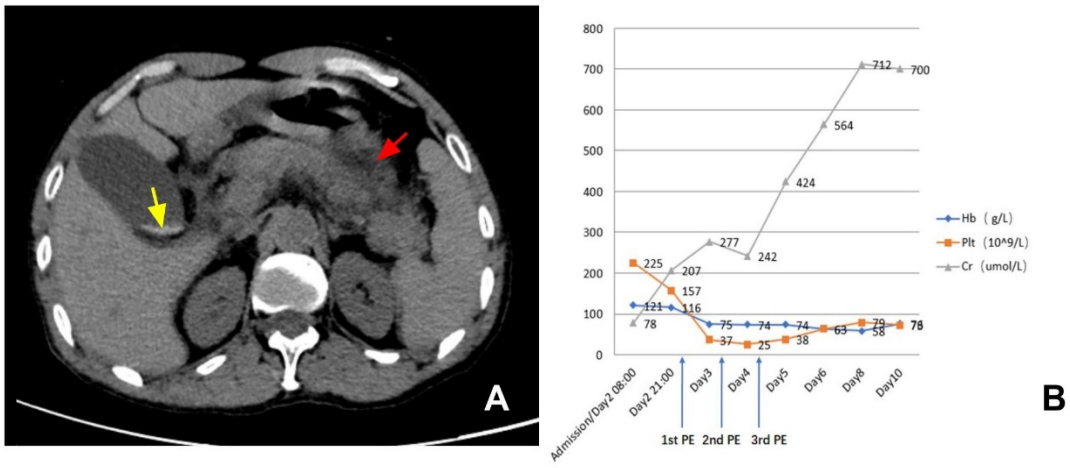

Figure 1. A. CT scan of the patient's abdomen. Gallbladder stones shown by yellow arrows and peripancreatic effusion shown by red arrows. B. Biochemical and haematological progression over the course of admission, and the timing of PE.

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