SOHO ANNUAL MEETING 2024

HEMOPHAGOCYTIC LYMPHOHISTIOCYTOSIS CO-PRESENTING WITH NON-CIRRHOTIC PORTAL HYPERTENSIVE GASTROPATHY: A CASE REPORT

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CONTEXT

Hemophagocytic lymphohistiocytosis (HLH) is a rare but life-threatening hyperinflammatory syndrome characterized by excessive immune activation. This report discusses a case of HLH co-presenting with non-cirrhotic portal hypertensive gastropathy (NCPHG), a combination that poses significant diagnostic and therapeutic challenges.

CASE PRESENTATION

A 54-year-old male patient with a history of H. pylori gastritis presented to the emergency department with black tarry stools and hematemesis. Initial endoscopy revealed severe exudative esophagitis, Grade 2 esophageal varices with red wale sign, and findings consistent with NCPHG. Endoscopic variceal band ligation was performed, and the patient was discharged on proton pump inhibitors (PPI)The patient was readmitted with high-grade fever and epistaxis. Blood tests revealed anemia, thrombocytopenia, and borderline leukopenia. Despite negative blood cultures, empirical antibiotics were initiated. As hematological parameters continued to decline, suspicion for HLH increased. Diagnostic tests confirmed HLH with elevated triglycerides (220 mg/dL), markedly raised serum ferritin (12,833.6 ng/dL), elevated fibrinogen (231 mg/dL), and increased serum LDH (1179 U/L). Ultrasound showed fatty liver changes, and peripheral blood smear indicated pancytopenia with a low reticulocyte count. Liver function tests revealed raised bilirubin (2.6 mg/dL) and GGT (239 U/L). Bone marrow biopsy confirmed HLH with 15%-20% macrophage infiltration. The patient developed oral bleeding and abdominal distension, with 2 liters of hemorrhagic fluid drained from ascites. Despite treatment with Filgrastim, nystatin mouthwash, PPI, and antibiotics, his condition deteriorated. He was started on the HLH-94 protocol with etoposide. Three days later, he experienced respiratory distress and was intubated. Despite intensive care, the patient remained unresponsive and was designated DNR due to the high risk of bleeding and family concerns. The patient subsequently died..

DISCUSSION

This case highlights the complexity and severity of HLH, especially when co-presenting with NCPHG. The overlapping symptoms and rapid progression emphasize the need for prompt recognition and aggressive management of HLH.

CONCLUSION

HLH is a critical condition that can co-present with other severe pathologies such as NCPHG. Early diagnosis and comprehensive treatment protocols are vital for improving patient outcomes in such complex cases.

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