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A Case of Mistaken Identity: Hemophagocytic lymphohistiocytosis presenting as acute liver failure

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Case Description

A 22-year-old female presented to Urgent Care clinic with symptoms of fever, abdominal pain, emesis, and "dark urine" with negative leukocyte esterase, nitrites, and bacteria on urinalysis. Her past medical history was significant for aggressive ulcerative colitis, obtaining partial remission on vedolizumab, mesalamine, and recently introduced mercaptopurine. Oral antibiotics were prescribed, but the patient presented to the local emergency department one week later with progressive fevers (>40°C) WBC count 18,000/mm^3 with lymphocyte predominance, abdominal tenderness, and conjugated hyperbilirubinemia. Blood cultures were obtained, and IV antibiotics started for presumed abdominal sepsis. Cultures resulted with no growth, and despite broadened antibiotic coverage, the patient developed worsening SIRS physiology with tachycardia, tachypnea, and persistent fevers. Altered mental status ensued with a blood ammonia level measured at 20µmol/L. Serum bilirubin continued to rise to 25mg/dL while serum transaminases remained within normal limits. The patient was diagnosed with fulminant liver failure of an unknown etiology, though toxicity from the recently started mercaptopurine was suspected. She was transferred to a liver transplant facility by air transport and admitted directly to the transplant surgery ICU service. Her clinical status continued to deteriorate with persistent fevers and new onset acute kidney injury, pancytopenia, and progressive coagulopathy with INR >3 and fibrinogen level 89mg/dL. The patient was subsequently listed status 1A for urgent liver transplantation while further diagnostics were performed. Abdominal ultrasound revealed hepatosplenomegaly, and liver biopsy was significant for periportal lymphocytic infiltration. A ferritin level was checked as part of her workup and found to be extremely elevated at 7,224ng/ml, with no evidence of iron deposition on liver biopsy to suggest hemochromatosis. A hematology consult was requested to address the profound pancytopenia and grossly elevated ferritin, with bone marrow biopsy demonstrating hemophagocytes characteristic of hemophagocytic lymph histiocytosis (HLH.) Aggressive management of HLH was begun, including IV-Ig, intravenous etoposide and dexamethasone, intrathecal methotrexate, and ultimately plasmapheresis based upon HLH-1994 guidelines [1]. She eventually made a full recovery over 25 days, and more recently has undergone total colectomy as curative therapy for her ulcerative colitis. The patient provided written consent for the publication of this case report.

Discussion

HLH, a syndrome of excessive inflammation and tissue destruction due to abnormal immune activation and excessive inflammation, is most commonly encountered in the primary form during infancy as a result of genetic mutations [2]. This case underscores the significant challenge and necessity of accurately and promptly identifying secondary, or acquired, HLH in the critically ill adult patient. Throughout her medical course, the disease mimicked urinary tract infection, gastroenteritis, abdominal sepsis, and cryptogenic versus medication-related hepatic failure, prompting multiple seemingly appropriate

interventions but without any evidence of improvement. It was fortunate that HLH was diagnosed prior to an orthotopic liver transplant being performed, as transplantation would not have treated the underlying disease and would have most likely resulted in disease recurrence in the transplanted organ if she were able to survive the operation in the first place. HLH is determined to be present when 5/8 of the following are present: fever >38.5°C; hepatosplenomegaly; cytopenia of 2/3 cell lines; hypertriglyceridemia and/or hypofibrinoginemia; hemophagocytes present in bone marrow, liver, spleen, or lymph nodes; low or absent natural killer (NK) cell activity; elevated ferritin >500mg/dL; elevated soluble CD-25 (an IL-2 receptor) [2] The patient in the present case report met 6/8 criteria at the time of recognition of HLH, and elevated soluble CD-25 and decreased NK cell activity were eventually detected as well giving her a total score of 8/8. This timeline emphasizes an important point in the management of HLH, and one most experts can agree upon: treatment for HLH must begin immediately, even when the diagnosis is not certain, to maximize the likelihood of a favorable patient outcome [3]. One unique element to this case was the effective use of plasmapheresis as salvage therapy when an immediate clinical response to HLH-specific therapies was not encountered. As previously described in case reports of severe HLH, plasmapheresis may act to temper the destructive processes of run-away immune activation by the clearance of cytokines and toxic plasma substances [4]. In the present case, profound clinical improvement was observed within four hours following the first course of plasmapheresis.

Conclusion

Part of the difficulty in diagnosing HLH is its ability to mimic a wide variety of disease states. To the author's knowledge, this is the first case of a patient nearly receiving a liver transplant for fulminant hepatic failure prior to the recognition of HLH as the underlying cause. Specifically, the grossly elevated ferritin level contributed to the recognition of HLH as a possible etiology, and the author recommends that a ferritin level be checked early in the diagnostic period of any patient with acute liver failure. Continuing to transplant in this circumstance would have been potentially disastrous, and the patient eventually made a full recovery with aggressive HLH-specific therapies.

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