SECONDARY HAEMOPHAGOCYTIC LYMPHOCYTOSIS PRESENTING AS PROLONGED FEVER AND CHOLESTATIC JAUNDICE - A DIAGNOSTIC DILEMMA

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Hemophagocytic Lymphohistiocytosis (HLH) is a rare but life-threatening hyperinflammatory syndrome caused by uncontrolled immune activation and excessive cytokine release. Secondary HLH may occur in association with infections, malignancies, or autoimmune disorders. Because its clinical manifestations overlap with severe infections and hepatobiliary diseases, HLH often presents as a diagnostic dilemma, particularly in tuberculosis-endemic regions. A 27-year-old male presented to Shaheed Suhrawardy Medical College Hospital with intermittent high-grade fever for seven months, progressive jaundice for three months, and significant weight loss. On examination he was febrile, deeply icteric, moderately anemic, and had hepatomegaly with ascites and pedal edema. Laboratory investigations revealed anemia, markedly elevated inflammatory markers, hyperbilirubinemia (total bilirubin 19.2 mg/dL), elevated liver transaminases, severe hypoalbuminemia, and markedly raised serum ferritin levels. Imaging studies including ultrasonography and contrast-enhanced CT scan demonstrated hepatomegaly, intra-abdominal lymphadenopathy, and ascites, while MRCP showed no evidence of biliary obstruction. Viral markers and autoimmune screening were negative. Bone marrow examination revealed areas of marrow necrosis with preserved hematopoiesis. The constellation of prolonged fever, hyperferritinemia, cytopenia, liver dysfunction, hepatomegaly, and bone marrow abnormalities fulfilled several HLH-2004 diagnostic criteria, raising strong suspicion of secondary HLH. Further evaluation suggested an underlying infectious trigger, possibly disseminated tuberculosis. The patient was managed with supportive therapy and evaluation for potential triggers. Recognition of the hyperinflammatory syndrome enabled timely consideration of HLH-directed management along with treatment of the underlying cause. Secondary HLH should be suspected in patients presenting with prolonged fever, hyperferritinemia, cytopenias, hepatomegaly, and liver dysfunction when routine investigations fail to reveal a clear diagnosis. Early recognition using established diagnostic criteria is crucial because delayed diagnosis significantly increases mortality.

Keywords: Hemophagocytic lymphohistiocytosis, hyperferritinemia, prolonged fever, cholestatic jaundice, hyperinflammatory syndrome.

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