

An atypical presentation of smoldering hemophagocytic lymphohistiocytosis (HLH)

Komisarof JA, McGann K, Evans AG, Morgan MA

Abstract:

Hemophagocytic lymphohistiocytosis (HLH) is a rare and life-threatening syndrome of immune system dysregulation characterized by the phagocytosis of various cells by histiocytes in the bone marrow. HLH can present in one of two ways: Primary HLH, which is caused by mutations in genes essential to T and NK-cell function, and secondary HLH, typically caused by Epstein-Barr virus (EBV) infection or malignancy. Because of the rapid progression and high mortality of this disease, prompt diagnosis is essential to good outcomes. Here, we report the 2-month clinical course of a patient who presented initially with altered mental status and recurrent fever of unknown origin. Initially he did not meet diagnostic criteria for HLH and had a negative bone marrow biopsy; however, he eventually progressed to full-blown HLH secondary to EBV+ Hodgkin lymphoma. This case is unusual for the slow and smoldering course of the patient's disease and highlights the importance of aggressively working up all diagnostic criteria in patients with suspected HLH to ensure the initiation of definitive therapy as soon as possible.