

Abstract

Hemophagocytic lymphohistiocytosis (HLH) is a rare but potentially fatal disorder that can result in multiple organ damage associated with significant morbidity and mortality. It is characterized by the uncontrolled proliferation of lymphocytes and histiocytes leading to substantial cytokine storm. Furthermore, it carries a broad spectrum of clinical presentation with numerous underlying causative factors such as infective causes, autoimmune conditions, and neoplasms that means vary depending on the individual. This is a rare case scenario regarding a patient who presented with a protracted history of fever reflecting pyrexia of unknown origin whose initial clinical picture did not suggest any successive clues directing into a probable differential diagnosis, thereby enhancing the complexity of the case profile. However, thanks to the high degree of clinical suspicion ultimate diagnosis of HLH could be made in the background of long-term fever, pancytopenia, and bone marrow evidence of Hemophagocytosis. Subsequently, further efforts were taken to identify the possible etiological factors attributed to HLH, as it is important in the proper management of the patient. Eventually, the final diagnosis of HLH secondary to probable lymphoplasmacytic lymphoma was made following a trephine biopsy. Unfortunately, at the climax of this case scenario patient's demise occurred depicting the diverse spectrum of variable clinical presentation and rapidly deteriorating nature as well as the poor outlook of the illness if met with undue delaying in early commencement of HLH treatment. Interestingly, this is an extremely rare case scenario in which there is co-existence of a HLH and plasma cell disorder. Moreover, this case report emphasizes the importance of the initial commencement of HLH therapy at its first encounter without waiting for further confirmatory studies or fulfillment of diagnostic criteria for a better outcome of the patient.