Abstract:

Hemophagocytic lymphohistiocytosis (HLH) is a rare , but an aggressive , life threatening clinical syndrome due to excessive immune activation. HLH most frequently affects the age group from birth to 18 months, but can rarely occur in adults of any age. HLH generally has a poor prognosis with mortality rates exceeding 50% and the greatest barrier to better outcome is misdiagnosis or delay in the diagnosis of HLH itself and its trigger which defers treatment resulting in worse outcomes due to multi organ failure. HLH can present with prolonged fever of unknown origin with a trigger such as infection, malignancy, autoimmune condition or without any such trigger.

Here we present a case of a previously well young male patient who presented with prolonged fever with no identifiable source of infection, found to have HLH with a possible trigger from bacterial sepsis. Further evaluation revealed bone marrow infiltration by a tumour of Ewing sarcoma family, which would have been the underlying etiology for his presentation with HLH and bacterial Sepsis.

This case report reiterates the importance of considering the possibility of HLH in cases of prolonged fever of unknown origin and this emphasizes the need to diagnose HLH and its triggers early to avoid disastrous outcomes as delay in HLH specific therapy and treatment of the underlying etiology (ie : malignancy , infection etc) would result in inevitable mortality.