Abstract

Haemophagocytic lymphohistiocytosis (HLH), is a hyper-inflammatory state which can lead to multi-organ failure and even death. It is characterized by fever with cytopaenias, splenomegaly and haemophagoctyes in bone marrow. Characteristically there are laboratory changes such as hypertriglyceridaemia, hypofibrinogenaemia, hyperferritinaemia, low or absent natural-killer cell activity and raised soluble CD25 levels. We came across a 19-year-old school girl, who was evaluated for pyrexia of unknown origin with polyarthralgia. She developed bicytopaenia with splenomegaly. There were multiple, small cervical and inguinal lymph nodes. Investigations revealed the possibility of sepsis due to the raised procalcitonin level (PCT). However with the administration of several antibiotics from different classes, there was no clinical response. She continued to have high-spiking fever. Therefore the possibility of HLH was considered due to the presence of hypertriglyceridaemia and hyperferritinaemia. Even with the absence of haemophagocytes on bone marrow, she fulfilled the diagnostic criteria for HLH. The fever disappeared with the initiation of intravenous dexamethasone. Further investigations revealed her anti-nuclear antibody titre to be 1:80 (positive) with a rheumatoid factor of 664 U/ml (<10). Antibodies against cytomegalovirus also turned out be positive (IgM). She made a remarkable recovery with dexamethasone and hydroxychloroquine. This case illustrates that sepsis is an important trigger for the development of HLH, especially among patients who are predisposed for macrophage activation. It is also important to consider HLH as a possible diagnosis in cases of prolonged fever with cytopaenias.