Abstract

Haemophagocytic lymphohistiocytosis (HLH) is an aggressive systemic inflammatory disorder characterized by the presence of abundant haemophagocytes in the body. Haemophagocytes are the macrophages that engulfed red blood cells, white blood cells and platelets. There are several infective, inflammatory and malignant etiologies for HLH. HLH secondary to anaplastic lymphoma kinase (ALK) positive anaplastic large cell lymphoma (ALCL) is found to be very rare. It carries a significant mortality despite of available treatments. HLH should be considered in an ill patient with rapid clinical deterioration. They usually present with high fever, organomegaly, lymphadenopathy, pancytopenia and multiorgan failure. Elevated serum ferritin, triglycerides and lactate dehydrogenase and hypofibrinogenemia are striking laboratory findings.

We present a rare case of HLH as the presenting manifestation of an underlying anaplastic lymphoma kinase (ALK) positive ALCL in a young female. She presented with fever, lymphadenopathy, hepatosplenomegaly, ascites and bilateral pleural effusions. She ended up in multi-organ failure and death within few days of hospitalization. Her bone marrow and lymph node histology reports revealed numerous haemophagocytes. Immunohistochemistry of lymph node biopsy revealed ALCL. Clinical features along with biochemical data reinforced the diagnosis of HLH.

Diagnosis and management of HLH in combination with ALCL is challenging due to its nonspecific clinical features and relentless progression. It carries a very high overall fatality. However, timely diagnosis will generate better outcome.