DIAGNOSIS

In view of the clinico-radiological picture and the findings on flowcytometry, a diagnosis of γ-δ T cell lymphoma consistent with Hepatosplenic lymphoma was rendered.

DISCUSSION

Hepatosplenic T cell lymphoma (HSTL) is a rare subtype of peripheral T cell lymphomas characterised by proliferation of neoplastic T cells with predilection for sinusoids of liver, sinuses and red pulp of spleen and sinuses of bone marrow. It is a highly aggressive entity which is mostly derived from cytotoxic γ-δ T cells. A minority subset of patients exhibit the α-β immunophenotype. It typically involves young adults and has a predilection for males which constitute 80-90% of the cases. Upto 20 % cases arise in the background of chronic immunosuppression with solid organ transplants being the most common indication[1]. A few cases of HSTL have presented in the setting of prolonged antigenic stimulation as occurs in infection with P. falciparum and EBV. HSTL is characteristically known to occur in patients of Crohn's disease, especially children, treated with azathioprine and infliximab. The highest is observed in men younger than 35 years old who receive long term treatment (more than 2 years) with thiopurines[2]. Patients usually present with fever, abdominal pain and weakness, and have marked hepatosplenomegaly resulting in jaundice. Bone marrow involvement is seen in upto 80% cases. Lymphadenopathy is usually absent to minimal. Patients usually manifest with marked thrombocytopenia, which in many cases is a part of a broader pancytopenia. The cause of the cytopenias is multifactorial with marrow infiltration being the most common etiological factor. Other factors include the presence of splenic sequestration, haemolytic anaemia, immune-mediated thrombocytopaenia, or haemophagocytic syndrome. These can serve as a cause in isolation or combined with one another. Patients can also present in leukemic phase where in there is a spill of these abnormal cells in the peripheral blood. Lactate dehydrogenase levels are usually markedly raised [1,2,3].

Clinical differentials include hepatotropic virus acute liver infection, myelodysplastic syndrome, autoimmune thrombocytopenic purpura, in cases with isolated thrombocytopenia; lymphoblastic leukemia, and T-cell lymphoblastic lymphoma expressing γ-δ TCRs. The spleen