ABSTRACT

Background: Langerhans cell histiocytosis (LCH) is a rare histiocytic disease that causes organ infiltration by histiocytes along with other hemopoietic cells, macrophages and eosinophils. Disease restricted to the lymph nodes and spleen is even more rare.

Case: This is a case of a 63 years old man with diabetes mellitus presenting with fever for 2 months. Evaluation for infectious and inflammatory etiologies were negative and eventually a mediastinal lymph node biopsy revealed the diagnosis of Langerhans cell histiocytosis.

Conclusion: Langerhans cell histiocytosis (LCH) is a rare cause of pyrexia of unknown origin. Involvement of the spleen which is a "risk organ" has a poor prognosis.