A RARE CASE OF LYMPHOMA ASSOCIATED HEMOPHAGOCYTIC LYMPHOHISTIOCYTOSIS

Abstract

Back ground

Hemophagocytic Lymphohistiocytosis is a clinical condition associated with hyper inflammation. Uncontrolled activation of normal immune system results in hypercytokinemia. Primary hemophagocytosis due to genetic mutations is more common in pediatric population whereas Secondary hemophagocytosis due to infections, autoimmune disorders and malignancy is seen in adults. Among the secondary causes, Lymphoma Associated Hemophagocytic Syndrome is common and carries a therapeutic dilemma with it. Treatment modalities include combined chemotherapy regimens. Hematopoietic stem cell transplant appears to be beneficial.

Case Presentation

Here, we report a case of a seventy-five years old female with past history of Diabetes and Hypertension, who presented with a history of fever and impaired cognition and general wellbeing that progressed over the course of two weeks. She had constipation in addition. Her complexion was dark and she was both pale and icteric. There was an enlarged left axillary lymph node. She was febrile and was tachycardic. Significant hepatosplenomegaly was there with ascites. She had pancytopenia, hyperbilirubinemia, low serum fibrinogen level and markedly high serum ferritin level. Her bone marrow had the evidence of hemophagocytosis and the Contrast Enhanced Computed Tomography chest and abdomen had the typical features of a lymphoma with splenic involvement where the patient did not survive until a histological diagnosis.

Conclusion

Lymphoma associated syndrome of Hemophagocytic Lymphohistiocytosis(HLH) is rare and carries a poor prognosis. Especially in elderly patients when the

criteria for HLH is fulfilled the possible malignant causes and loci should be sought for. Aggressive chemotherapy followed by allogenic stem cell transplantation appears to be effective in some pediatric populations and the role is yet to be established in adults with Secondary HLH.

Key words: Hemophagocytic Lymphohistiocytosis, Lymphoma, Immune activation