DIAGNOSIS OF THE CASE:

Hepatosplenic T-cell lymphoma in young male

DISCUSSION:

HSTCL is a very rare type of the peripheral T-cell lymphomas, which accounts for less than 1% of non-Hodgkin lymphomas and about 3% of all T-cell lymphomas/leukaemias.[1]

HSTCL is more commonly seen in young adulthood, more common in males. History of immunosuppression is found in 10-20% of patients either through treatment for inflammatory conditions (inflammatory bowel disease) or malignancy (such as acute myeloid leukemia or non-Hodgkin lymphoma).[2,3]

Cytogenetic studies of patients of hepatosplenic T-cell lymphoma showed isochromosome of chromosome 7 (i(7)(q10)), a recurrent genetic abnormality. It is seen either in isolation or along with a loss of a sex chromosome and trisomy 8[4-7]

Hepatosplenic T-cell lymphoma is very rare and intially the diagnosis may be overlooked. Differential diagnosis of hepatosplenic lymphoma includes T-cell large granular lymphocytic leukaemia, splenic marginal zone lymphoma, NK-cell leukaemia, enteropathy-associated T-cell, and primary cutaneous γ-δ T-cell lymphoma, myelodysplasic syndrome, tuberculosis, leishmaniasis and infectious mononucleosis.[8-13]

Hepatospenic T-cell lymphoma can be differentiated from T-Large granular lymphoma with the help of immunohistochemistry. Atypical lymphoid cells in T-LGL express CD57, granzyme B, and perforin. In addition, T-LGL is more common in elderly individuals, has indolent course, and sometimes associated with an underlying autoimmune disorder.

This lymphoma is clinically aggressive and has poor prognosis and median overall survival of 11 months although more recent studies suggest improved survival with intensive induction chemotherapy followed by early high-dose therapy and hematopoietic stem cell transplantation.[14]