DISCUSSION

Hepatosplenic T-cell lymphoma (HSGDTCL) is a rare entity, characterized by primary extra nodal disease with typical sinusoidal infiltration of the liver, spleen and bone marrow by T cells expressing T-cell receptor (TCR)γδ chain [1]. It has an aggressive course with hepatosplenomegaly without lymphadenopathy and a predominance of young male adults [2,3]. Bone marrow is involved in two-third cases. As many as 20% of HSGDTCLs arise in the setting of chronic immune suppression, most commonly during long term immunosuppressive therapy for solid organ transplantation or prolonged antigenic stimulation. Patients typically presents with hepatosplenomegaly and systemic symptoms. Patients usually manifest marked thrombocytopenia, often with anemia and leucopenia. Given the almost constant bone marrow involvement, virtually all patients have Ann Arbor stage IV disease [5]. Flowcytometry enables the diagnosis on bone marrow hence avoiding the need for liver or spleen biopsy. A phenotype which has been accepted to be common in Hepatosplenicγδ T cell lymphoma is CD2+, CD3+, CD4-, CD5-, CD7+, CD8-, CD56 +/- and TCR-γδ +[4].

CONCLUSION

HSGDTCL is a unique subtype of T cell lymphoma affecting only liver, spleen and bone marrow. It is clinically challenging to diagnose and seeking help from flowcytometry/IHC required.Though no standard guideline available to treat this aggressive type of tumour, combination of chemotherapy (CHOP like) can prolong duration of survival.

REFERENCES

1. Khan WA, Yu L, Eisenbrey AB, Crisan D, al Saadi A, Davis BH, Hankin RC, Mattson JC.Hepatosplenic gamma/delta T-cell lymphoma in immunocompromised patients. Report of two cases and review of literature.Am J ClinPathol. 2001 Jul; 116(1):41-50.

2. Salhany KE, Feldman M, Kahn MJ, Peritt D, Schretzenmair RD, Wilson DM. Hepatosplenicgammadelta T-cell lymphoma: ultrastructural, immunophenotypic, and functional evidence for cytotoxic T lymphocyte differentiation.Hum Pathol. 1997 Jun; 28(6):674-85.

3. Weidmann E. Hepatosplenic T-cell lymphoma: a review on 45 cases since the first report describing the disease as a distinct lymphoma entity in 1990. Leukemia. 2000;14(6):991–7.