



Hepatosplenic T Cell Lymphoma: A Rare Disease

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Case Report

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Abstract

Hepatosplenic T cell lymphoma (HSTCL) is an uncommon neoplasm comprising 5% of peripheral T-cell lymphomas. We report an uncommon case of Peripheral T-cell lymphoma that is characterized by primary extranodal disease with malignant T cell proliferation in spleen, liver and bone marrow. 19 year old male patient presented with fever, weakness and pain abdomen for 2 months. On clinical examination he was pale and had massive hepatosplenomegaly. The diagnosis was quite challenging as thorough clinical, hematological and immunophenotypic correlation was required.

Keywords: Hepatosplenic; Gamma-delta receptor; Bone marrow; Immunophenotypic; Biopsy

Abbreviations: HSTCL: Hepatosplenic T Cell Lymphoma; REAL: Revised European-American Lymphoma.

Introduction

HSTCL is an uncommon neoplasm which in majority of cases express gamma-delta receptor [1]. Typical clinical features include predominance in young males with an aggressive clinical course, massive hepatosplenomegaly with sinusoidal infiltration of liver, spleen and bone marrow. This case highlights the importance of considering diagnosis of HSTCL in patients presenting with massive hepatosplenomegaly.

Case Report

A 19 year male patient presented with a history of lethargy, weight loss, fever and distended pain abdomen for 2 months. On physical examination he had pallor and massive hepatosplenomegaly with no lymphadenopathy. His CBC revealed hemoglobin of 12.1 g/dl. WBC count of 3900cells/cumm and platelet count of 50000/cumm. Differential counts revealed: polymorphs-42%, lymphocytes-41%. Monocytes- 5%. Atypical cells -12%. Peripheral blood smear showed predominantly normocytic normochromic RBCs with atypical lymphocytes.

Bone marrow examination revealed aparticleate yet cellular smears. Nucleated cell: erythroid ratio 3.16:1. Erythroid cells showed normoblastic maturation with features of mild dyserythropoiesis. Myelogram: atypical cells: 39%, myelocytes: 08%, Metamyelocytes: 04%, Polymorphs and band forms: 16%, Eosinophils: 02%, Lymphocytes: 07%, and erythroid cells: 24%. Atypical cells were large with scant basophilic cytoplasm, round to oval nuclei having open chromatin with 1-4 prominent nucleoli (Figure 1).

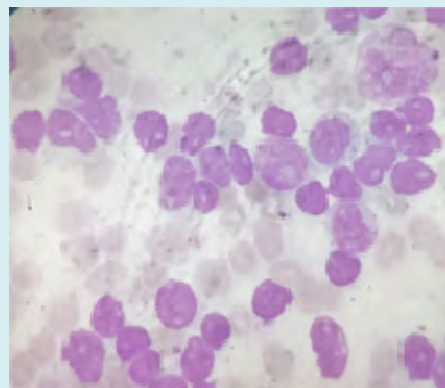


Figure 1: Showing infiltration by medium to large sized atypical lymphoid cells having fine chromatin and prominent 1-4 nucleoli (Leishman stain; 1000X).

Clinical findings	Peripheral blood smear findings	Bone marrow examination findings	Bone marrow biopsy findings	Immunophenotypic Findings
Pallor with massive hepatosplenomegaly	DLC: polymorphs-42%, lymphocytes-41%. Monocytes -5%. Atypical cells -12% . PBS:normocytic normochromic RBCs with atypical lymphocytes.	Nucleated cell: erythroid ratio -3.16:1 . Erythroid cells showed normoblastic maturation with features of mild dyserythropoiesis. Myelogram : atypical cells:39%, myelocytes:08%, Metamyelocytes: 04%, Polymorphs and band forms: 16%, Eosinophils :02%, Lymphocytes : 07%, and erythroid cells : 24%.	Sinusoidal and interstitial infiltrarion by medium to large sized atypical lymphoid cells.	CD3 ,CD7 , CD45 , TCR gamma delta positive

Table 1: Showing clinical, PBS, Bone marrow aspiration, Biopsy and immunophenotypic findings.

Flow cytometry was done on bone marrow aspirate that revealed cells with low SSC and moderate CD45 expression. The gated atypical T lymphoid cells showed positivity for CD3, CD7, CD45 and TCRgamma-delta. So a diagnosis of HSTCL was made. Bone marrow biopsy showed hypercellular bone marrow with overall cellularity of 90-95%. Sinusoidal and interstitial infiltration by medium to large sized atypical lymphoid cells was noted (Table 1).

Discussion

Hepatosplenic T-cell lymphoma is a rare entity of peripheral T-cell lymphomas that is characterized by hepatosplenomegaly without significant peripheral lymphadenopathy. Initially, the term gamma-delta hepatosplenic T-cell lymphoma was proposed as a provisional entity in the Revised European-American Lymphoma (REAL) classification [2]. However, identification of patients who demonstrated an alpha-beta phenotype with the clinicopathologic features of HSTCL, the term 'hepatosplenic T-cell lymphoma' was adopted for use in the current World Health Organization classification [3].

The incidence of HSTCL is unknown. However, since its initial description in 1990 by Farcet, et al. more than 150 cases have been reported in the literature, approximately 20% of these cases occurred in young men with history of immunosuppression including solid organ transplantation and leukemia [3,4]. In addition, patients with inflammatory bowel diseases receiving immunosuppressant and anti-tumor necrosis factor- α agent are also at risk for developing HSTCL. Clinically, patients with HSTCL present with B-symptoms,

jaundice and hepatosplenomegaly. Lymphadenopathy has been reported only in a few patients during the course of the disease [5]. The predominant laboratory findings include pancytopenia and deranged liver chemistry. The diagnosis of HSTCL is not always straightforward because of the rarity of the disease and occasionally the misleading symptoms. Therefore, in most cases patients will require liver biopsy and/or splenectomy to establish the diagnosis [6].

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