Case Report

Hepatosplenic T-cell lymphoma: A clinical diagnostic challenge

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Abstract

Hepatosplenic T-cell lymphoma (HSTCL) is a rare variant of T-cell lymphoma and comprises less than 5% of all peripheral T-cell lymphomas. These lymphomas have an aggressive course and dismal prognosis. We report a case of a 28-year-old male who presented with repeated episodes of fever and abdominal pain. Ultrasonography revealed hepatosplenomegaly. Liver biopsy performed showed sinusoidal lymphoid cell infiltration of the T-cell immunophenotype with aberrant antigen loss. The case was finally diagnosed as HSTCL.

Keywords: Hepatosplenic T-cell lymphoma, non-Hodgkin's lymphoma, T-cell lymphoma

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INTRODUCTION

Hepatosplenic T-cell lymphoma (HSTCL) is very rare type of T-cell non-Hodgkin's lymphoma. These lymphomas are characterized by primarily involving the liver and spleen, with relative sparing of lymph nodes. Bone marrow involvement is usually seen. Most of these cases are seen in young and adolescent males, usually presenting with fever and abdominal symptoms. Liver or spleen biopsy helps in establishing the diagnosis. We report a case of HSTCL in a 28-year-old male.

CASE REPORT

We report a case of a 28-year-old male software engineer by profession who presented with complaints of fever, abdominal pain, and blood in urine for the last 6 days and black-colored stools. His history revealed a high-grade fever 2 months back which was managed by supportive therapy. On examination, blood pressure was 130/80 mmHg; there was no history of pallor, icterus, cyanosis, clubbing, pedal edema, or lymphadenopathy. Laboratory investigations revealed hemoglobin of 10.1, white blood cell 4.2, eryhtrocyte sedimentation rate 06, platelet 130,000, urea 13 (13–43 mg/dl), creatinine 0.7 (0.72–1.18 mg/dl), bilirubin 0.47 (0.0–0.2 mg/dl), serum glutamic-oxaloacetic transaminase 77 (1–35 IU/l), serum glutamic pyruvic transaminase 217 (1–45 IU/l), serum alkaline phosphatase 191 (41–137 IU/l), total protein 5.6 (6.4–8.3 g/dl), albumin 3.3 (3.5–5.2 g/dl), Albumin ratio 1.4 (1.5–2.5), thyroid stimulating hormone 1.49, prothrombin time 18.4, ferritin 228.5 (20–250 ng/ml), iron 180, and total iron binding capacity 215 (250–450 μg/dl). Liver elastography showed increased liver stiffness of 16.3 kPa and increased median transmission of wave of 2.33 m/s (normal <1.4 m/s), indicative of fibrosis. Ultrasound abdomen showed right vesicoureteric junction

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calculus (4.5 mm), massive splenomegaly (18 cm), and mild hepatomegaly (15.6 cm). The patient was managed with antibiotics, intravenous fluid, antacids, antiemetics, and multivitamin. In view of hepatosplenomegaly and increased liver stiffness, liver biopsy was performed.

Liver biopsy tissue revealed diffuse sinusoidal infiltration by monotonous round cells, having round hyperchromatic nuclei, inconspicuous nucleoli, and pale cytoplasm. The infiltrate was predominantly sinusoidal with relative sparing of the portal tracts [Figure 1a and b]. Immunohistochemical (IHC) performed showed that the cells within the sinusoids are diffusely positive for CD45 and CD3 and negative for CD20, CD5, CD4, CD8, CD7, CD 56, EBV, tdt, and CD34 [Figure 2a and b]. Based on histology and IHC studies, the final diagnosis of HTCL was thus rendered.

DISCUSSION

Peripheral T-cell lymphomas are a heterogeneous group of postthymic, mature lymphoid malignancies, accounting for approximately 10%–15% of all non-Hodgkin's lymphomas.[1]

HSTCLs are rare subtypes of peripheral of T-cell lymphoma comprising less than 5% of all peripheral T-cell and natural killer cell lymphomas.[2]

HSTCL was first identified as a distinct category of T-cell lymphoma by Farcet *et al.* in 1990.[3] Since then, less than 150 cases have been reported so far.

In normal circumstances, γδ T-cells represent only 1%–3% of the lymphocytes in the peripheral blood and in liver comprise 3%–5% of all intrahepatic lymphocytes. These cells develop from CD4/CD8 thymocytes in the bone marrow.

It is believed that HSTCL arises usually from the peripheral γδ T-cells (or less commonly αβ) cytotoxic memory T-cells of the innate immune system.[4,5] HSTCL occurs more frequently in immunocompromised patients, especially in those receiving long-term immunosuppressive therapy. Immunomodulation thus may play a role in the activation of these cells. However, our patient was immunocompetent.

HSTCL occurs predominantly in adolescents and young adults, with a median age of 35 years (range, 15–65 years) at initial presentation. The male-to-female ratio is about 9:1. They are characterized by predominantly extranodal disease, with preferential involvement of the liver and spleen. Anemia and thrombocytopenia in patients with HSTL have largely been attributed to hypersplenism and to infiltration of the bone marrow by neoplastic cells.

Diagnosis is usually established by tissue biopsy. The histology typically shows sinusoidal infiltration by monotonous cells with medium-to-small round nuclei, inconspicuous nucleoli, and pale cytoplasm. Similar histological involvement was noted in our case. The most common immunophenotype in the patients with HSTCL is as follows: CD2+, CD3+, CD4−, CD5−, CD7+/−, CD8−, CD16+/−, CD 38+, and CD56+.[6]

Our patient had a common immunophenotypic profile of CD2+, CD3+, CD4−, CD5−, CD7−, CD8−, and CD56−.

Certain cytogenetic and molecular features have been found in patients with HSTCL, most notably, isochromosome 7q, and less commonly, trisomy 8.[7]

Despite these advances, HSTCL remains a very aggressive subset of T-cell lymphoma and confers a

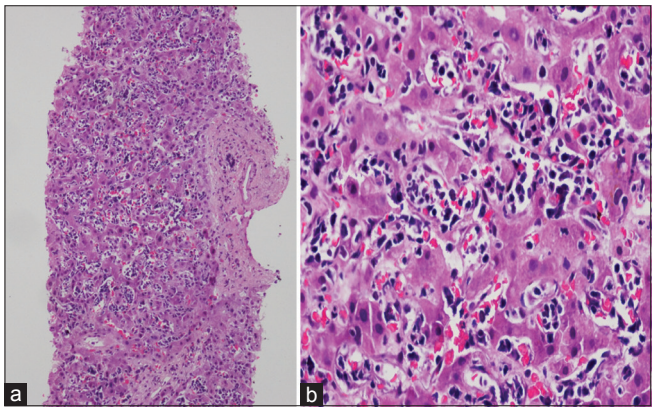

Figure 1: (a and b) Hematoxylin and eosin showing diffuse sinusoidal infiltration

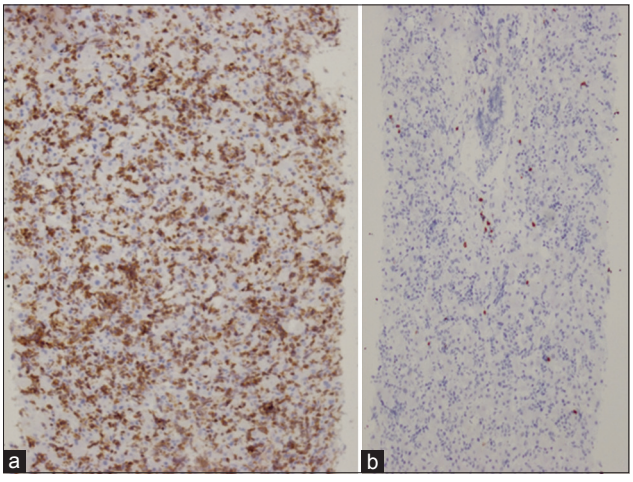

Figure 2: (a) Immunohistochemical showing diffuse CD3 positivity. (b) CD20 negative

poor prognosis, with a reported median survival of 6–11 months.[8,9]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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