Hidden in Plain Sight: A Rare Cause of Hepatosplenic T-Cell Lymphoma

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Introduction

- Hepatosplenic T-Cell Lymphoma (HSTCL) is an aggressive subgroup of non-hodgkin's lymphoma
- Clonal expansion and infiltration of cytotoxic T-cells[1,2]
- Worldwide incidence is approximately 1.4% for the γδ variant and even less for the αβ phenotype[1]
- Features include constitutional B symptoms, cytopenias, elevated liver enzymes, hepatosplenomegaly, and lack of lymphadenopathy[1,2]
- We present a rare case of αβ variant of HSTCL in an elderly woman

Case Description

79 year old female with past medical history of Type 2 Diabetes mellitus and hyperlipidemia presenting with generalized weakness and weight loss.

Review of Systems: Loss of appetite, weight loss

Medications: Atorvastatin, Metformin

Physical Exam: No stigmata of chronic liver disease

Labs:
- Viral Hepatitis A, B, C: negative
- Immunoglobulin G: negative
- Epstein Barr virus, Herpes simplex virus, Cytomegalovirus: negative
- Strongyloides antibody: negative
- Smooth muscle antibody, Anti-Liver kidney microsome antibody, Antimitochondrial antibody, Antinuclear antibody: negative
- INR 1.1

	Day 1	Day 3	Day 27
AST (U/L)	860	1419	222
ALT (U/L)	938	1048	97
ALP (U/L)	416	336	188
Total Bilirubin (mg/dL)	5.8	7.7	24.1
Platelets (10*3/mcl)	129	115	26
Albumin (g/dL)	4.1	3.7	2.1

Table 1. Liver function tests and additional laboratory data
AST: Aspartate aminotransaminase; ALT: Alanine aminotransaminase; ALP: Alkaline phosphatase

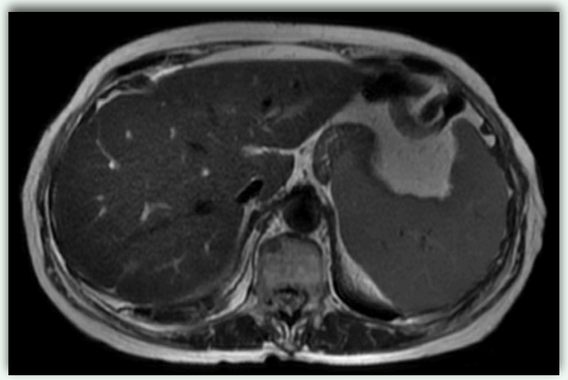

Figure 1. MRI Abdomen showing mild hepatosplenomegaly

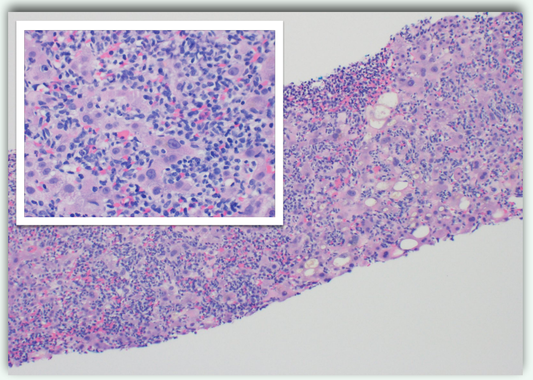

Figure 2. Liver biopsy showing infiltrate of atypical T-cells with high Ki67 index and αβ positivity

Molecular testing confirmed the presence of monoclonal T-cell receptor (TCR) beta and gamma populations highly suggestive of hepatosplenic T-cell lymphoma with αβ phenotype.

Discussion

- HSTCL is a rare disease entity with ambiguous presentations
- The median age of diagnosis is 34 and elevated liver enzymes can be seen in around 38-43% of γδ cases[1]
- Less than 40 reported cases of the αβ phenotype[2]
- αβ phenotype is seen more in women over 50 years of age and has been associated with worse prognosis[3]
- Most cases occur de novo, affecting the liver, spleen, and bone marrow and approximately 20% of cases arise in a setting of immunosuppression, often associated with autoimmune disorders and inflammatory bowel disease
- Outcomes are poor with a 5 year survival of < 10% without bone marrow transplant[1]

Teaching Points

- Albeit rare and difficult to diagnose, HSTCL should be considered on the differential for elevated liver enzymes.
- Early diagnosis is crucial for immediate induction chemotherapy

References

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