

Hidden in Plain Sight: A Rare Cause of Elevated Liver Enzymes



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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 $\gamma\delta$ variant and even less for the $\alpha\beta$ phenotype¹
- Features include constitutional B symptoms, cytopenias, elevated liver enzymes, hepatosplenomegaly, and lack of lymphadenopathy^{1,2}
- We present a rare case of likely HSTCL with $\alpha\beta$ variant in an elderly woman

Case Description

79 year old female with past medical history of Type 2 Diabetes mellitus and peptic ulcer disease status post gastric artery embolization presenting with a two day history of generalized weakness.

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

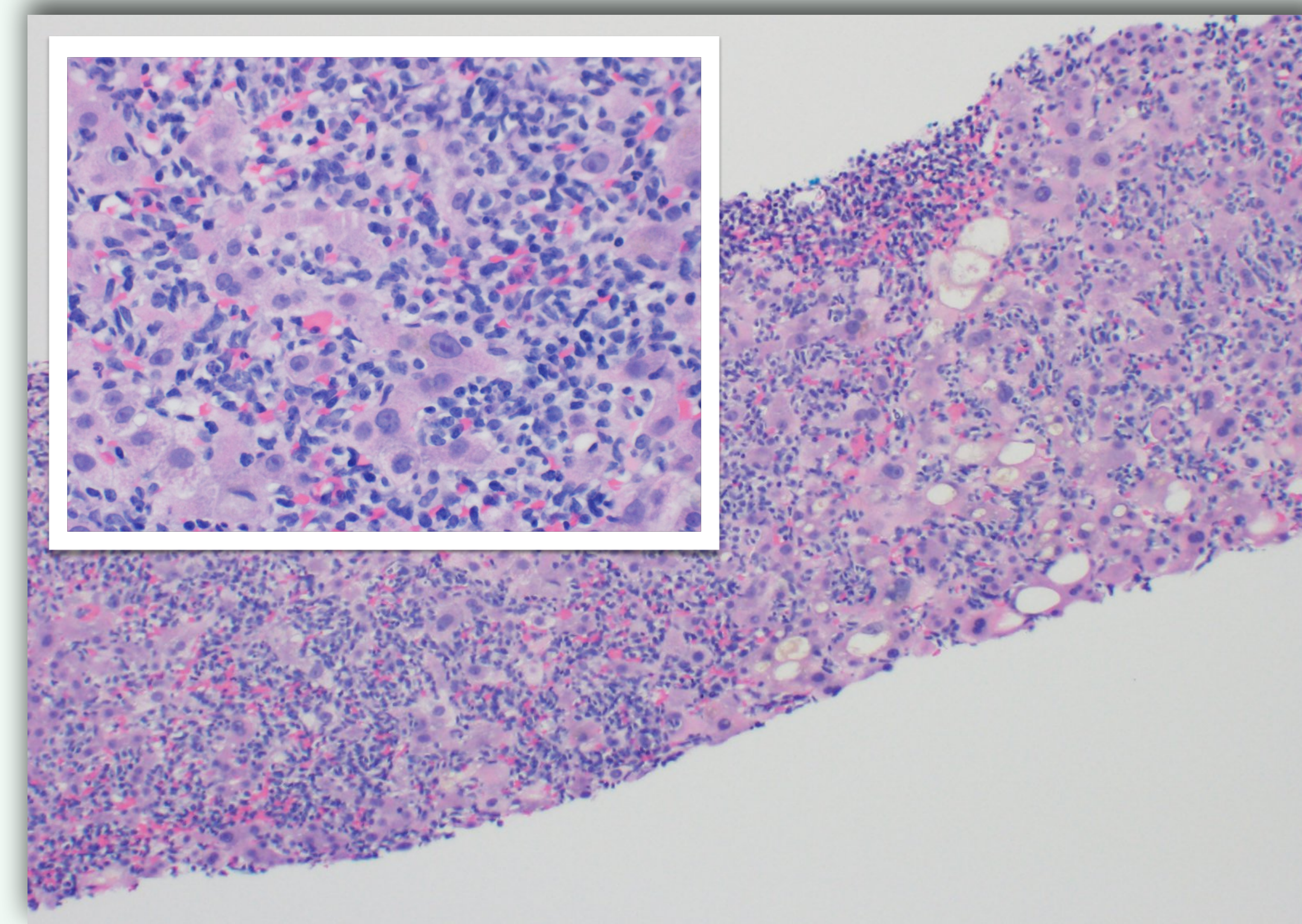


Figure 1. Liver biopsy showing infiltrate of atypical T-cells with high Ki67 index and $\alpha\beta$ positivity

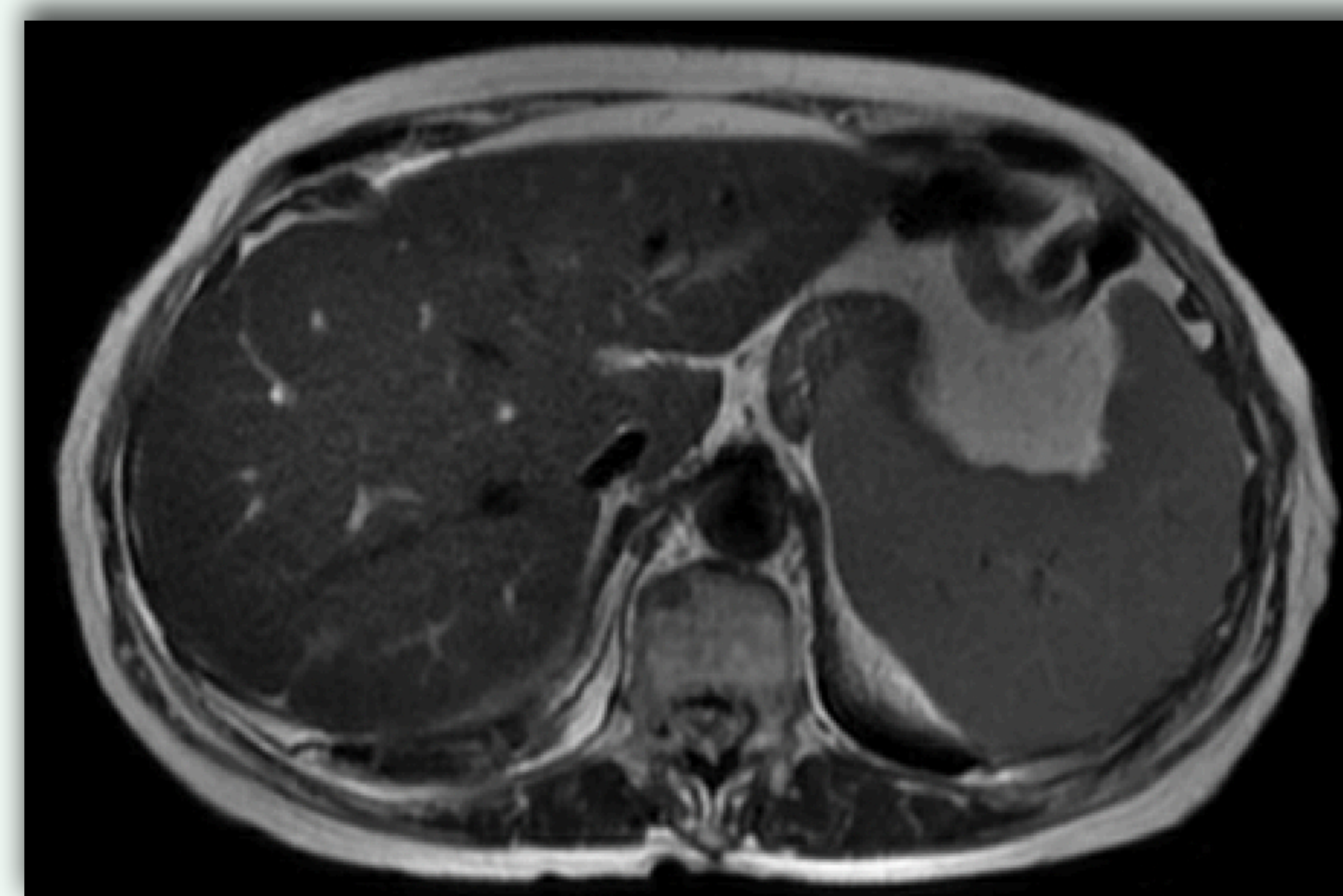


Figure 2. MRI Abdomen showing mild hepatosplenomegaly

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 $\gamma\delta$ cases¹
- Less than 40 reported cases of the $\alpha\beta$ phenotype²
- $\alpha\beta$ phenotype is seen more in women, in patients over 50 years of age, and has been associated with worse prognosis³
- 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 transplant¹
- Molecular testing is required for confirmation of diagnosis and is currently in process for our patient

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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3. Yabe M, Miranda RN, Medeiros LJ. Hepatosplenic T-cell Lymphoma: a review of clinicopathologic features, pathogenesis, and prognostic factors