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ELEVENTH ANNUAL MEETING

HEPATOSPLENIC T-CELL LYMPHOMA: A RARE AND COMPLEX NEOPLASM IN AYA

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INTRODUCTION

Hepatosplenic T-Cell Lymphoma **(HSTCL)** is a **rare** sub-type of peripheral **T-Cell neoplasm**, that can affect adolescents and young adults (**AYA**). Diagnosis requires high clinical suspicion and histopathology confirmation; ideally under **multidisciplinary** collaboration. We present an AYA with HSTCL who required external consultation for correct diagnosis and prompt initiation of effective therapy.

CASE REPORT

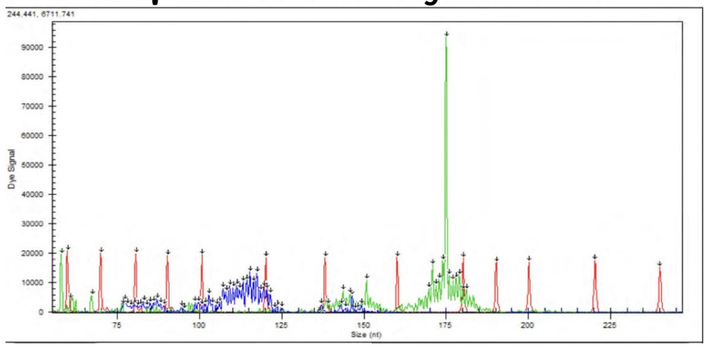
15-year-old male presented with weakness, fever and had massive splenomegaly. He was severely pancytopenic (Hb: 4.3 g/dl, WBC: 1900/mm3, platelets: 15.000/mm3). Initial assessment ruled out neoplasia and infectious etiologies. His splenomegaly progressed, requiring multiple transfusions. His splenomegaly progressed, requiring multiple transfusions.

External consultation allowed for correct diagnosis of HSTCL, by bone marrow morphology, IHC, blood immunophenotype, and PCR. Chemoterapy was given with good response and he remains in remission.

RESULTS

Chemotherapy started with ICE (ifosfamide 5g/m2 d2, carboplatin 5 AUC d2, etoposide 100mg/m2 d1-3) support, with good response and spleen shrinkage. On day 21, ESHAP (etoposide 40mg/m2 d1-4, cytarabine 2g/m2 d5, cisplatin 25mg/m2 d1-4, methylprednisolone 500mg d1-5) was completed, uneventfully. There was clinical improvement, but the patient refused to continue therapy. Six months later, he apparently remains in remission.

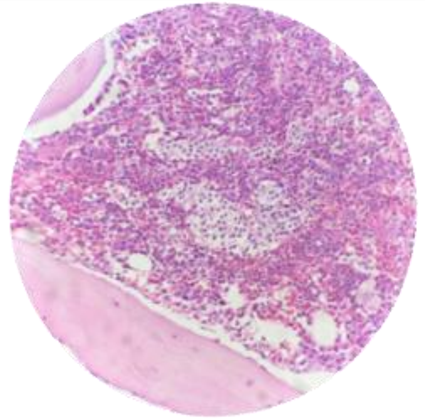
TCR-γ Chain Gene Rearrangement Studies



TCR-γ chain PCR using Vγ 9-Jγ (green) and Vγ 11-Jγ (blue) primer sets (DNA ladder shown in red)



CT Scan of t at diagnosis, showing massive splenomegaly.



H&E Bone Marrow intrasinusoidal growth pattern of medium-sized lymphocytes with moderately abundant cytoplasm, imparting a clear-cell appearance.

CONCLUSIONS

HSTCL is life-threatening. There is urgent need for research and collaborations for this rare neoplasm.

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CONTACT INFORMATION

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