Case of the month

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Chief complaints

- 19 years old male presented with complaints of generalized weakness, significant weight loss and intermittent high grade fever since last 8-10 months
- There was no lymphadenopathy

Examination

- General physical examination : Pallor++
- Systemic examination : Splenomegaly present
  : Lymphadenopathy absent

Radiological investigation

- PET-CT – Hepatosplenomegaly with FDG uptake involving bone marrow distribution of axial and appendicular skeleton with mild FDG avid subcutaneous stranding and thickening with mild ascites (? Malignant lymphoma)

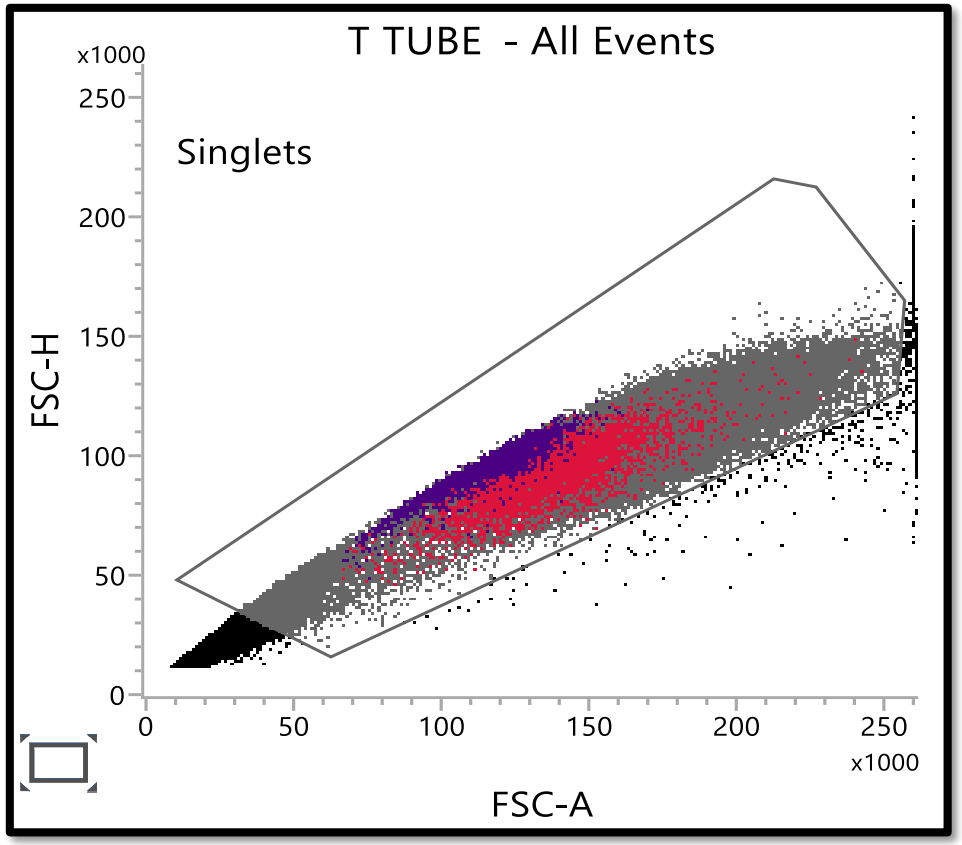
Hematological investigations

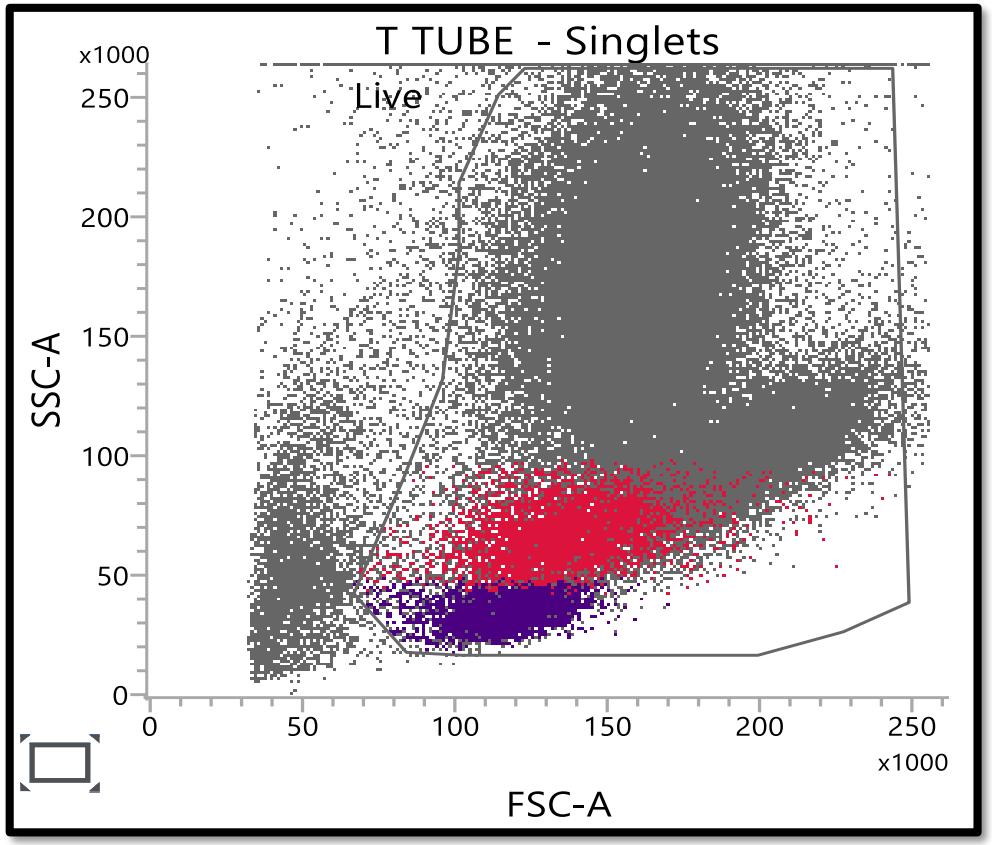
- **CBC and peripheral smear-** Hb :5.4 g/dl, Platelet count: 1,72,000 / cu mm on smear, TLC: 4580 / cu mm with mild shift to left and occasional blastoid cells (2%)
- **Bone marrow aspiration**: 16% large atypical lymphoid cells with round to oval nucleus, inconspicuous nucleoli and moderate amount of basophilic cytoplasm.
- **Bone marrow biopsy:** Focal presence of ? immature cells having vesicular chromatin and central prominent nucleoli. No definitive marrow infiltration was seen

IN VIEW OF SEVERE PERIPHERAL BLOOD ANEMIA AND LARGE ATYPICAL LYMPHOID CELLS IN BONE MARROW ASPIRATION AND BIOSPY, FLOW CYTOMETRY WAS ADVISED

Flow cytometry

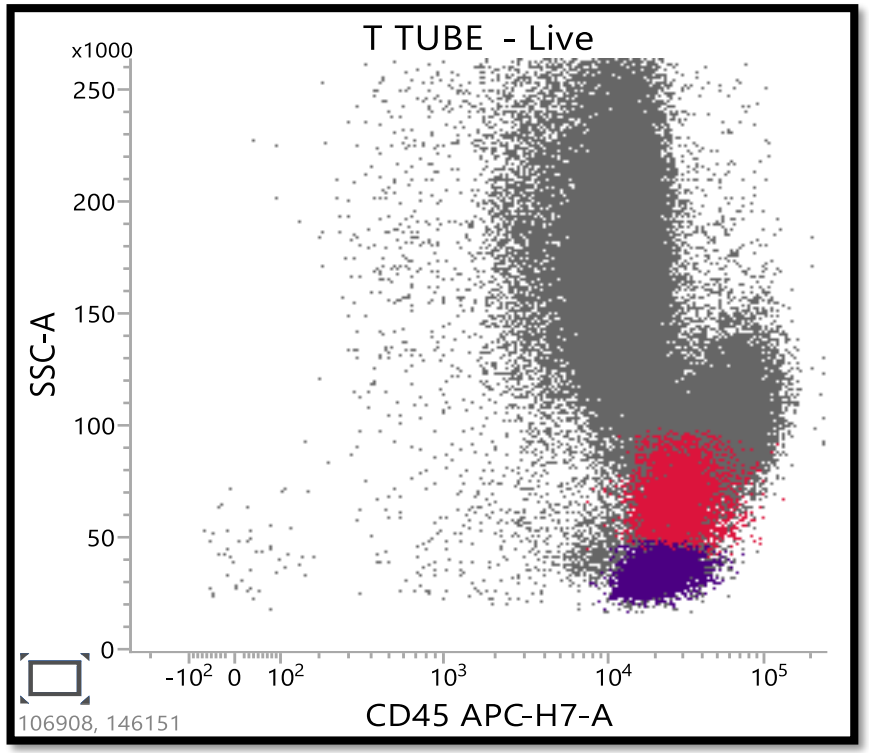
Flow cytometry





Flow cytometry

CD45 Vs SSC

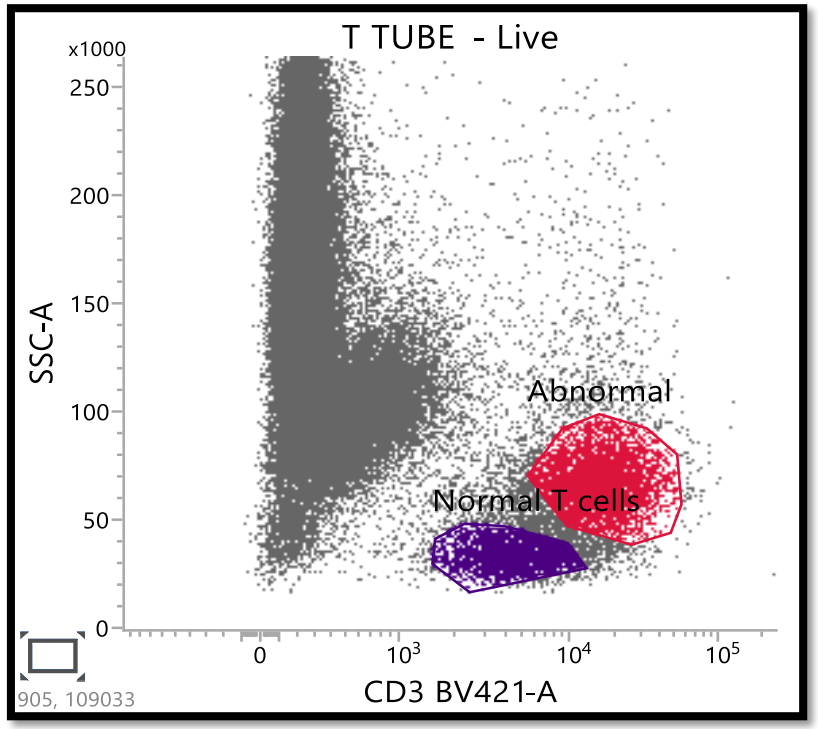


Interpretation

- An abnormal population is seen in bright positive CD45 region(red in colour)
- This population has CD45 expression equivalent to that of normal lymphocytes (purple in colour)
- This population also has higher side scatter i.e increased granularity and nuclear chromatin complexity

Flow cytometry

SSC Vs CD3

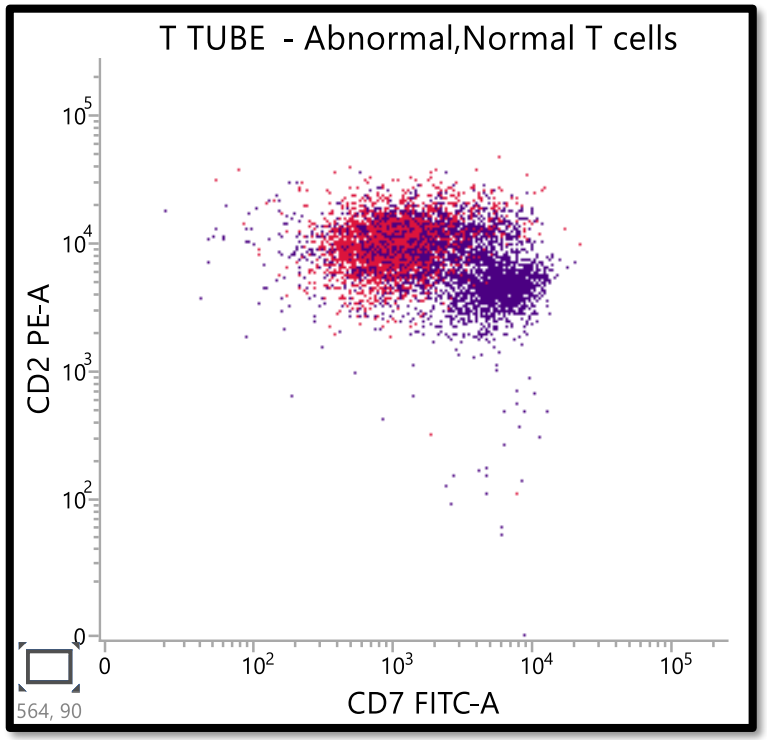


Interpretation

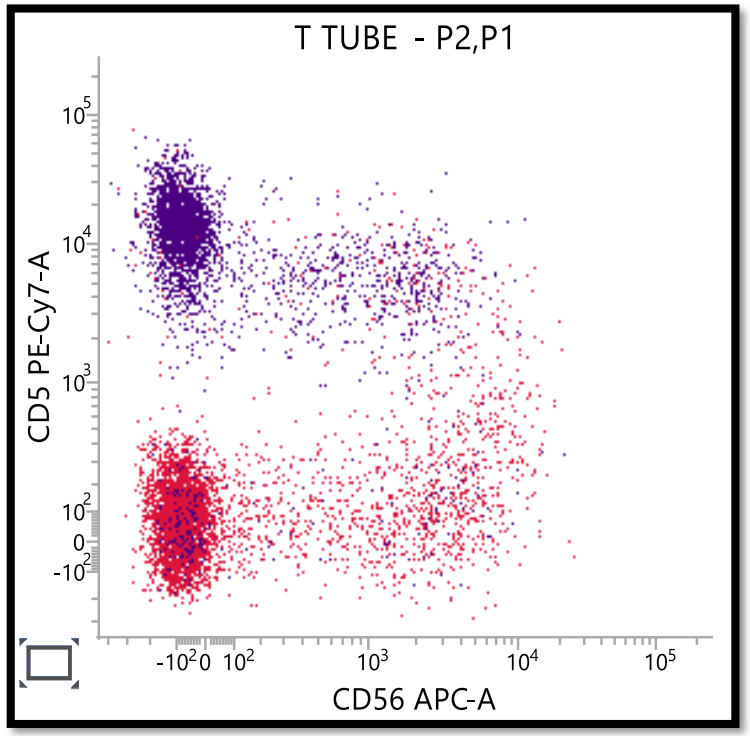
- Abnormal population shows bright CD3 expression
- This expression is brighter than the normal T lymphocytes

Flow cytometry

CD 2 + CD 7+

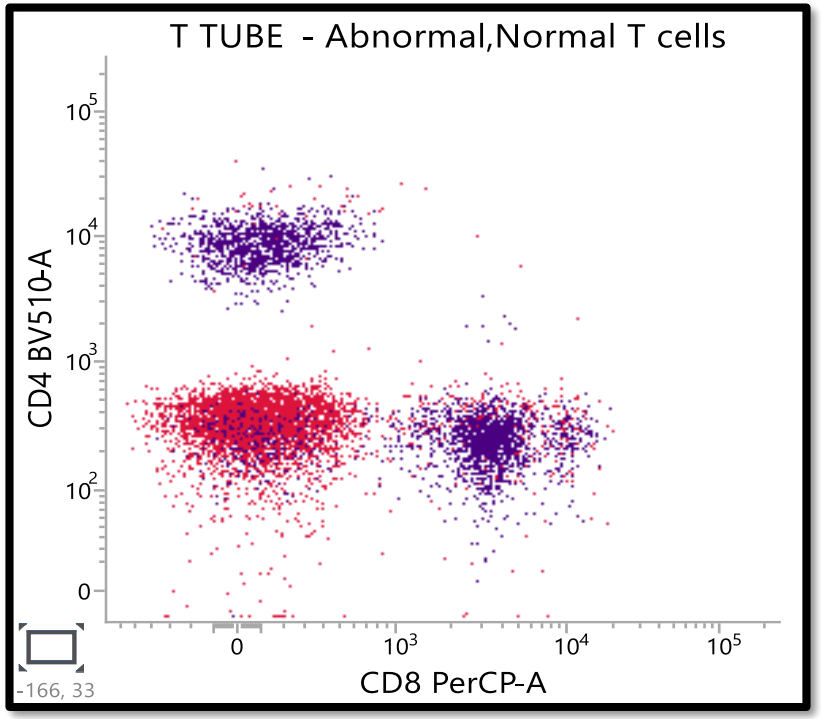


CD 5 – CD56 -



Flow cytometry

CD 4 -, CD8-



Interpretation

- 6 % cells were gated in the bright CD 3 and higher SSC region than normal lymphocytes (monocytic region on SSC VS CD 45)
- POSITIVE MARKERS- CD3 BRIGHT,CD45 BRIGHT,CD2 BRIGHT
- NEGATIVE MARKERS- CD5,CD4,CD8,CD56

Final Diagnosis: Hepatosplenic T cell lymphoma

(INVIEW OF SPLENOMEGALY AND ABSENT LYMPHADENOPATHY

Hepatosplenic T cell lymphoma

❑Definition

- Hepalosplenic T cell lymphoma (HSTL) is an extranodal and systemic neoplasm derived from cytotoxic T cells usually of yδ T-cell receptor type. These neoplasms were earlier called gamma delta T cell lymphomas but it was found that they can sometimes express an αβ phenotype and therefore the name was changed to Hepatosplenic T cell lymphoma

❑Epidemiology

- Very rare (<1% of all non-Hodgkin lymphoma)
- Peak incidence in adolescents and young adults (Median age: 35 years)

❑Etiology:

- Long-term immunosuppressive therapy in patients with solid organ transplant
- Prolonged antigenic stimulation in children treated with azathioprine and infliximab for crohn’s disease

❑Sites of involvement

- Patient’s present with splenomegaly and hepatomegaly. No lymphadenopathy seen
- Bone marrow always constantly involved

❑ Clinical features

- Splenomegaly+
- Hepatomegaly+/-
- Anemia, leucopenia, thrombocytopenia

❑Morphology

Spleen- diffuse involvement of red pulp

Liver - diffuse involvement

❑Histopathology

- Spleen- Cells are monotonous with medium-sized nuclei and a rim of pale cytoplasm. Nuclear chromatin is loosely condensed with small inconspicuous nucleoli . They involve cords and sinuses of splenic red pulp with atrophy of the white pulp.
- Liver - predominant sinus infiltration
- Bone marrow-Neoplastic cells are almost constantly present with intra-sinusoidal distribution

❑Immunophenotype

- Neoplastic cells are CD3+,TCRδ1+,TCR□β+,CD 56+/-,CD4-,CD8+/-,CD5-

❑Prognosis and predictive factors

- Course is aggressive. Patients may respond initially to chemotherapy, but relapses are seen in the vast majority of cases .
- The median survival is <2 years