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Case Report
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A RARE CASE OF T CELL-LYMPHOBLASTIC LYMPHOMA: IHC PROFILE STUDY

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ABSTRACT

Background: *Tcell-Lymphoblastic lymphoma is a rare subtype of Non-Hodgkin's lymphoma with an incidence of approximately 2% occurring most commonly in young men. The usual presentation is an anterior mediastinal mass and cervical lymphadenopathy.* ***Case report:*** *We report a case in a 15 year old male presenting with generalized lymphadenopathy with bilateral cervical lymph node enlargement, fever and general fatigue. After physical examination, Fine Needle Aspiration and subsequent excisional biopsy of cervical lymph node was performed.* ***Macroscopy*** *(Excision Biopsy): A single nodular tissue measuring 2x1.5 cm was received. Cut section showed grey white areas.* ***Microscopy****:* Sections studied showed *lymph node with completely effaced architecture, replaced by monotonous population of lymphoid cells in a diffuse pattern. Cells were moderately large having hyperchromatic nuclei with granular chromatin and scant cytoplasm. Also seen were scattered macrophages. A starry sky pattern was seen. Capsule showed tumor cell infiltration. On further confirmation by IHC: Tumor cells were diffusely immunopositive for CD45, Tdt, CD4, CD8, CD7 and partially immunoreactive for CD3 and CD45RO.Focal immunoreactivity for CD43 and CD20 for residual B cells noted. Ki-67 proliferative index was 40%.* ***Conclusion:*** *T cell-lymphoblastic lymphoma is a rare subtype of Adult Non-Hodgkin's lymphoma. It has a 90% cure rate in children with standard ALL chemotherapy regimen but has a poor prognosis in adults.*

KEYWORDS

Lymph node, Non-Hodgkin's lymphoma, T Cell-Lymphoblastic lymphoma

INTRODUCTION

T cell-lymphoblastic lymphoma is a rare subtype of Non- hodgkin's lymphoma with an incidence of approximately 2% occurring most commonly in adoloscents and young adults. Overall, 80%-90% of all lymphoblastic lymphomas are of T-cell lineage. The Male: Female ratio is 2:1.The patients present with respiratory difficulties due to a large mediastinal mass, often accompanied by symptoms of Superior Vena Cava obstruction, pleural effusion and supraclavicular lymphadenopathy [1]. There is a tendency of early dissemination to the bone marrow and central nervous system.

CASE REPORT

A 15 year old male presented with fever, weakness and generalized lymphadenopathy with bilateral cervical lymph node enlargement of 15-20 days duration. Hematological investigations revealed mild leukocytosis. Chest X-ray was normal. All other routine investigations were within normal limits. Bone marrow examination was within normal limits with normal myeloid to erythroid ratio. After

Physical examination, Fine needle aspiration cytology and subsequent excisional biopsy of right cervical lymph node was performed.

PATHOLOGICAL FINDINGS:

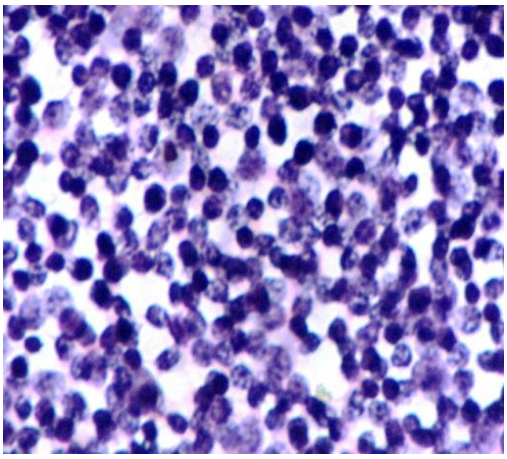
FNAC finding

Smears studied showed uniform population of cells with intermediate sized nuclei having convoluted and cerebriform shapes with fine granular chromatin, inconspicous nucleoli and scanty cytoplasm. Many mitotic figures seen (**Fig1a, b**).

Macroscopy: Received a single nodular tissue measuring 2x1.5 cm. Cut section showed grey white areas.

Microscopy: Sections studied show lymph node with completely effaced architecture, replaced by monotonous population of lymphoid cells in a diffuse pattern. Cells were moderately large having hyperchromatic nuclei with granular chromatin and scant cytoplasm. Also seen were scattered macrophages. A starry sky pattern was seen (**Fig2**). Capsule showed tumor cell infiltration.

IHC study: Tumor cells were diffusely immunopositive for CD45, Tdt, CD4, CD8, CD7 (**Fig.3, 4, 5, 6, 7**) and partially immunoreactive for CD3 and CD45RO (**Fig8,9**). Focal immunoreactivity for CD43 and CD20 for residual B cells noted (**Fig 10, 11**). Ki-67 proliferative index was 40% (**Fig.12**).



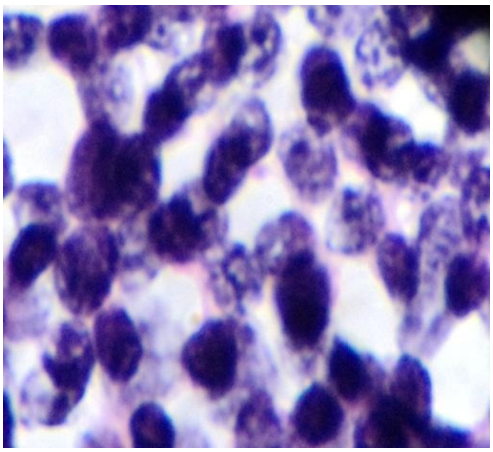


Fig. 1a & 1b- Cerebriform and convoluted nucleated cells (Giemsa 100X, 400X)

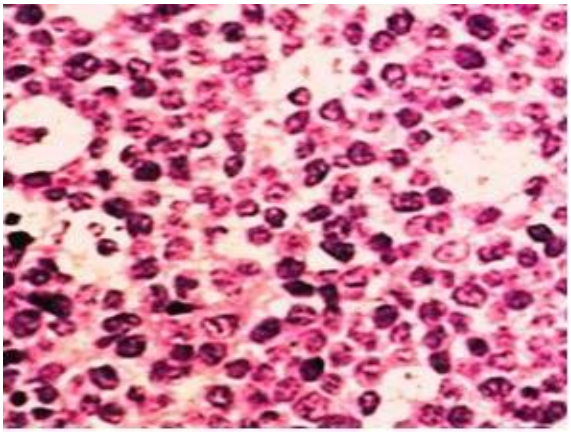


Fig. 2- "Starry sky pattern" seen in the section (H&E 100X)

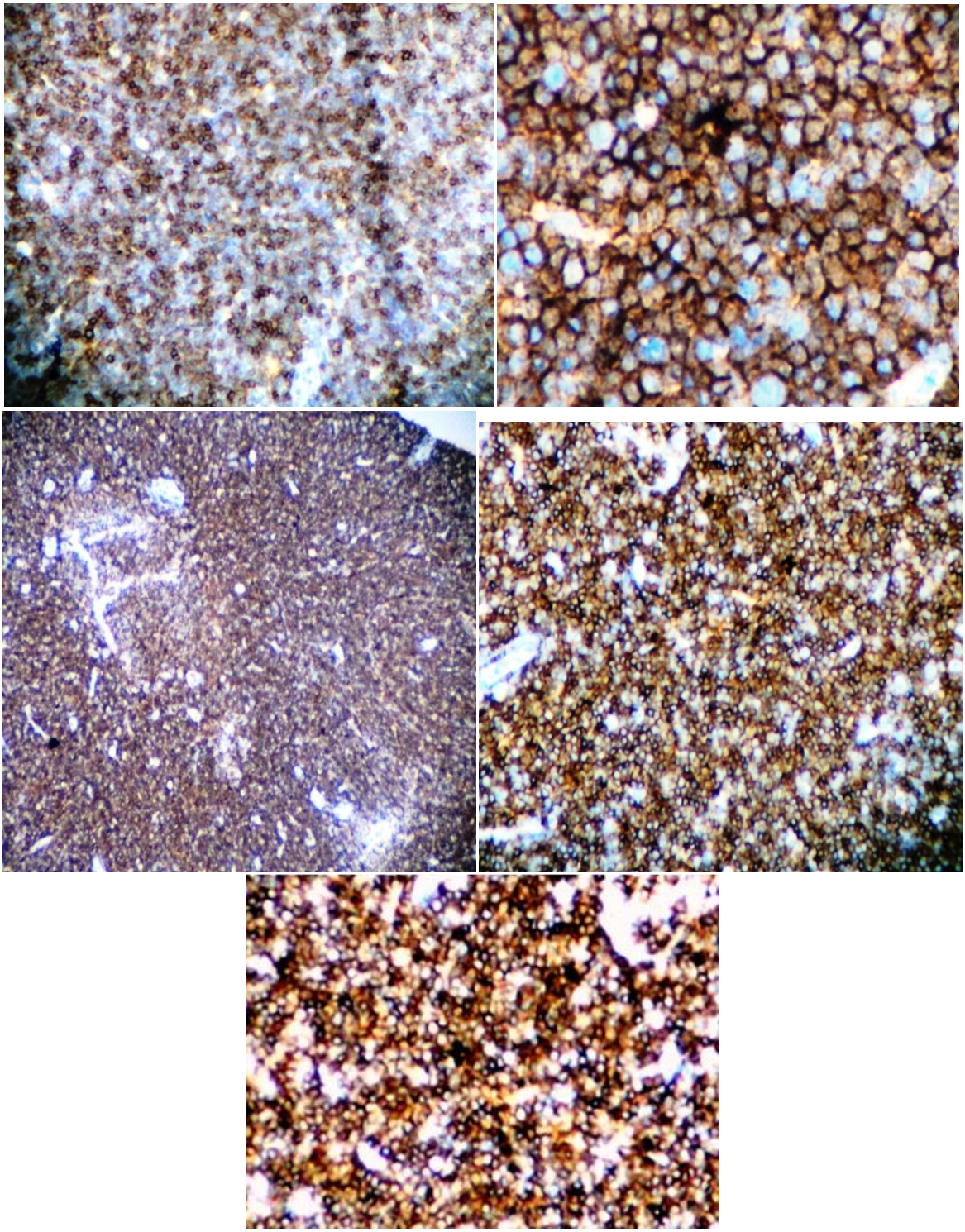


Fig. 3, Fig.4, Fig.5, Fig.6 & Fig.7- Diffuse immunopositivity of tumor cells for CD45,TdT CD4,CD7 & CD8 (100X,400X,100X,100X & 100X)

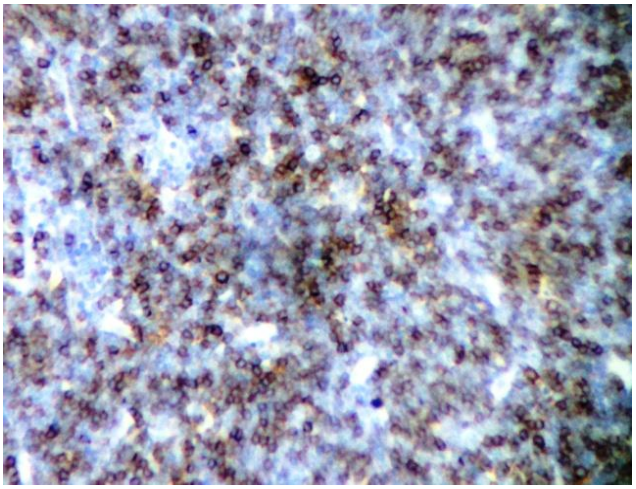
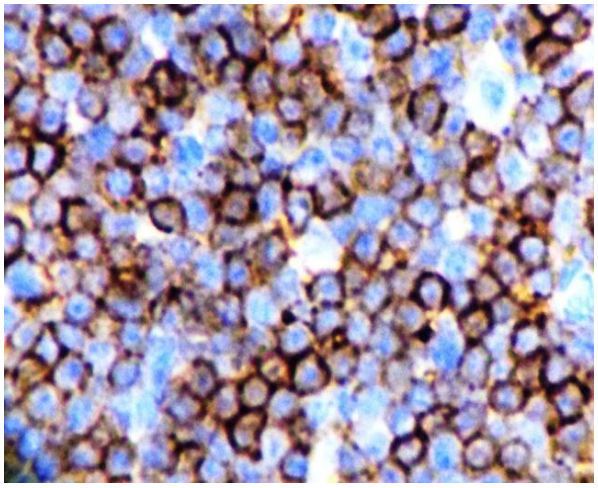



Fig. 8 & 9- Partial immunoreactivity of tumor cells for CD3 & CD45RO (100X & 400X)

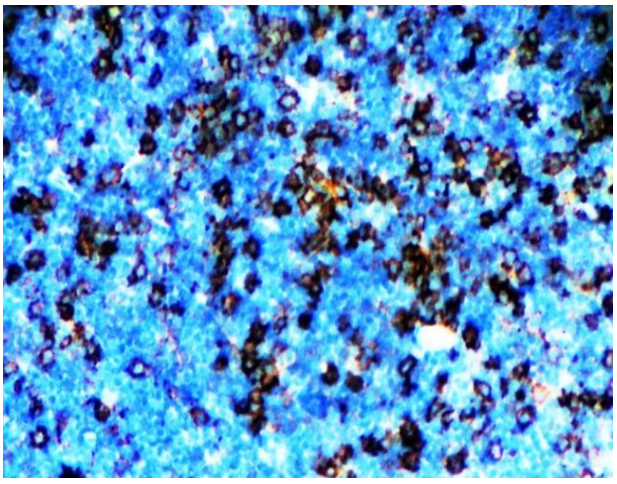
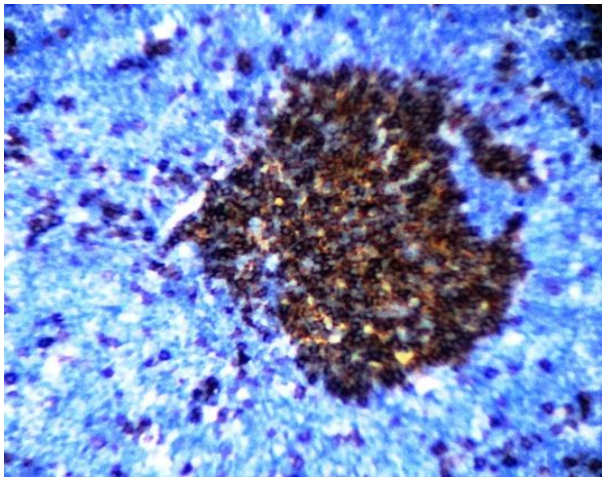



Fig. 10 & Fig. 11- Focal immunoreactivity of tumor cells CD43& CD20 (100X & 100X)

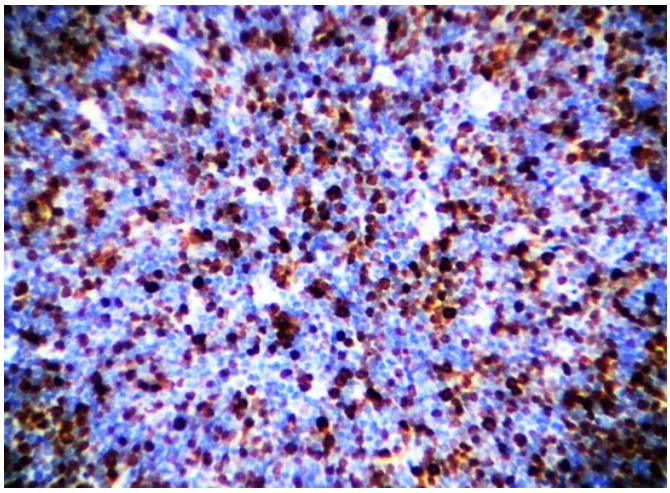


Fig. 12-Tumor cells positive for Ki-67(100X)

DISCUSSION

T cell - lymphoblastic lymphoma is a rare form of Non Hodgkin's Lymphoma representing 1.8-2% of all lymphomas in the NHL Classification project [1]. Over 90-95% of patients have Ann Arbor stage III-IV disease. The disease can involve a wide number of extranodal sites known as 'Sanctuary sites' such as the CNS and Reproductive system. Peripheral blood involvement by lymphoblasts is observed in one-third of patients. Conversion to full blown Leukemia is common. It follows an aggressive clinical course. Cytological examinations reveal diffuse population of cells with intermediate sized nuclei having convoluted and cerebriform shapes with fine granular chromatin, inconspicous nucleoli and scanty cytoplasm. Atypical mitotic figures are present. The histopathological and immunophenotypic features help distinguish from other subset of lymphomas. The characteristic findings such as lymph node architecture effacement replaced by monotonous population of lymphoblasts in a diffuse pattern characterized by high N: C ratio, convoluted or nonconvoluted nuclei, dusty chromatin and inconspicous nucleoli. These are positive for Pan T cell markers such as CD2, CD3, CD4, CD7and Tdt. CD1a, CD10, CD43, CD 99 and CD45RO are variably expressed [2,3]. The clinical features, cytological features histopathological findings and immunohistochemistry correlated with our findings. This entity should be differentiated with Myeloid sarcoma, Burkitt lymphoma, Thymoma, Mantle cell lymphoma, blastoid variant, Follicular lymphoma etc. with the help of immunohistochemistry as they may closely resemble clinically and based on histopathological features [4]. The initial diagnostic difficulty which was faced was absence of any mediastinal mass or related symptoms. Thus initial assessment was difficult, but cytological examinations, histopathology and further confirmation by IHC helped clinch the diagnosis.

CONCLUSION

T cell-lymphoblastic lymphoma is a rare subtype of Adult Non-Hodgkin's lymphoma. It has a 90% cure rate in children with standard ALL chemotherapy regimen but has a poor prognosis in adults. IHC plays a very important role in early diagnosis and helps in prevention of delay in the treatment thus increasing the survival rates.

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