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# Ambient or Static-T lymphoblastic Leukaemia/Lymphoma

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**Editorial**

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**Abbreviations:** TLBL: T Lymphoblastic Lymphoma; T ALL: T Acute Lymphoblastic Leukaemia; ALL: Acute Lymphoblastic Leukaemia; LBL: Lymphoblastic Lymphoma; CNS: Central Nervous System; PCR: Polymerase Chain Reaction; TCR: T cell receptor; FAB: French-American-British; ECOG: Eastern Cooperative Oncology Group; KPS: Karnofsky Performance Scales; TDT: Terminal Deoxynucleotidyl Transferase.

T lymphoblastic leukaemia/lymphoma is a neoplasm engendered from lymphoblast of T cell lineage which may configure lymphomatous tumefaction or incriminate peripheral blood and bone marrow. T lymphoblastic lymphoma (T LBL) appears to arise from thymocytes.

Additionally designated as pre T cell acute lymphocytic leukaemia / lymphoma (pre T ALL), T lymphoblastic lymphoma / leukaemia (T LBL) or acute lymphoblastic leukaemia with aberrant myeloid antigen expression, the condition incriminates adolescents and young adults. Majority of T lymphoblastic lymphoma commence following birth. A slight male predominance is observed. In contrast to T lymphoblastic lymphoma, T acute lymphoblastic leukaemia (T ALL) may be derived from T cell progenitors confined to bone marrow, depicts an immature immuno-phenotype, immune reactive CD47, an absence of chromosome 11q23 rearrangement and variable genetic expressions. T acute lymphoblastic leukaemia is categorized wherein ≥25% of lymphoblast appear to replace bone marrow cells. In the absence of aforesaid diagnostic criterion, the condition is designated as T lymphoblastic lymphoma.

T acute lymphoblastic leukaemia/ lymphoma may be segregated from B acute lymphoblastic leukaemia/ lymphoma with cogent immunohistochemistry or flow cytometry [1,2]. T acute lymphoblastic leukaemia/ lymphoma configure ~15% of childhood and ~25% of adult instances of acute lymphoblastic leukaemia (ALL). Also, T lymphoblastic lymphoma constitutes an estimated ~95% instances of lymphoblastic lymphoma (LBL). T lymphoblastic lymphoma represents with a mediastinal mass along with absent or minimal incrimination of bone marrow. Untreated T lymphoblastic lymphomas may manifest central nervous system (CNS) involvement [1,2]. Intra-thymic differentiation encountered within T acute lymphoblastic leukaemia/ lymphoma demonstrates cellular categorization denominated as •Pro-T cells delineating an immuno-phenotype of cCD3+, CD7+, CD2-, CD1a-, CD34+/-, CD4- / CD8-. •Pre-T cells exhibiting an immuno-phenotype of cCD3+, CD7+, CD2+, CD1a-, CD34+/-, CD4-/ CD8-. •Cortical T enunciating an immuno-phenotype of cCD3+, CD7+, CD2+, CD1a+, CD34-, CD4+/ CD8+. •Medullary T exemplifying an immuno-phenotype of cCD3+, CD7+, CD2+, CD1a-, CD34-, surface CD3+, either CD4+ or CD8+.

In contrast to B acute lymphoblastic leukaemia, T lymphoblastic leukaemia/ lymphoma demonstrates cryptic cytogenetic abnormalities which may be appropriately discerned by fluorescent in situ hybridization (FISH) or polymerase chain reaction (PCR) [1,2]. Generally, chromosomal translocation t(1;14)(p32;q11) implicating SCL (TAL1) and T cell receptor alpha/delta locus are encountered. Also, translocation t(10;14)(q24;q11) implicating HOX11 (TLX1) and T cell receptor alpha/delta locus may be discerned. Additionally, activating mutations of NOTCH1, chromosomal deletions of CDKN2A (INK4A) or non-lineage specific genetic rearrangements within T cell receptor (TCR) may occur [1,2]. One third (~30%) instances of T acute lymphoblastic leukaemia demonstrate genomic rearrangements within T cell receptor (TCR) loci as ~14q11.2 (alpha/ delta) ~7q35 (beta) ~7p(14-15) (gamma). T acute lymphoblastic leukaemia/ lymphoma manifests

chromosomal mutations within genes as ~MYC (8q24.1) ~TAL1 (1p32) ~RBTN1 (LMO1) (11p15) ~RBTN2 (LMO2) (11p13) ~HOX11 (TLX1) (10q24) ~HOX11L2 (TLX3) (5q35) ~LYL1 (19p13) ~LCK (1p34.3-35) Cytogenetic evaluation of T acute lymphoblastic leukaemia/ lymphoma exhibits monoclonal genomic rearrangements of T cell receptor (TCR) gene. Majority of instances depict an anomalous karyotype, especially within14q11.2 (a/d TCR loci), 7q35 (β) and 7p14-15 chromosomes. Frequently, T lymphoblastic lymphoma (LBL) represents with rapidly progressive tumefaction confined to anterior mediastinum which may engender pleural effusion with respiratory distress, thereby configuring a medical emergency [1,2]. In contrast to elderly subjects > 61 years, incriminated young subjects within 16 years to 60 years commonly delineate hepatosplenomegaly, mediastinal mass and regional or distant lymph node enlargement. Besides, myeloid antigens and lineage nonspecific genetic rearrangements are infrequently discerned [1,2].

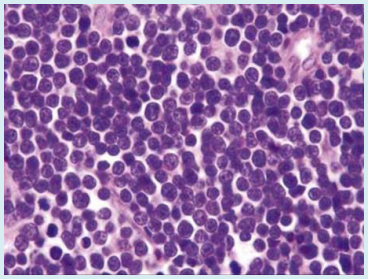

**Figure 1:** T lymphoblastic lymphoma depicting disseminated neoplastic lymphocytes with imbued with scant, basophilic cytoplasm, enlarged nuclei with delicate chromatin, convoluted nuclear membrane, nuclear grooves and indistinct nucleoli [2-5].

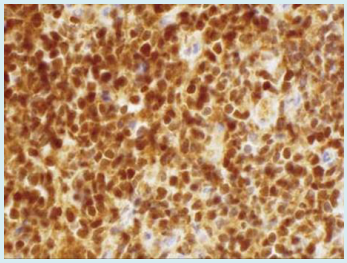

**Figure 2:** T lymphoblastic lymphoma immune reactive to terminal deoxy-nucleotidyl transferase (TdT)(5).

Cellular component of T acute lymphoblastic leukaemia/ lymphoma simulates B cell disease and is represented by neoplastic T lymphoblast pervaded with scanty cytoplasm, delicate nuclear chromatin, convoluted nuclear membrane with nuclear grooves and indistinct nucleoli. Mitotic activity is significant and configures a 'starry sky' configuration with commingling of benign macrophages. An interstitial pattern of bone marrow incrimination is observed, as encountered with lymphomas exemplifying features manifesting French-American-British (FAB) classification L1 or L2 (Figures 1 & 2). Incriminated lymph nodes demonstrate comprehensive effacement of nodal architectural or partial lymphomatous involvement with sparing of germinal centres and neoplastic infiltrate confined to para-cortex. Implication of thymus enunciates replacement of normal glandular parenchyma [1,2]. Occasionally, peripheral blood examination demonstrates eosinophilia and myeloid hyperplasia [1,2]. Chromosomal translocations as t(8;13)(p11.2;q11-22) with implication of FGFR1 gene are variably discerned wherein few instances with aforesaid translocations may engender myeloid malignancy as myelodysplastic syndrome, acute myeloid leukaemia or myeloid sarcoma. T acute lymphoblastic leukaemia/ lymphoma manifests distinctive morphological features as elevated mitotic index and capsular infiltration of implicated lymph nodes [1,2].

Prognostic outcomes and functional assessment of Non-Hodgkin's lymphoma may be evaluated with Eastern Cooperative Oncology Group (ECOG) Performance Status wherein a decimated score is indicative of superior functional status which is designated as ~grade 0 : completely active individual demonstrating pre-disease performance devoid of restriction ~grade 1: ability to walk, maintain non strenuous activities while standing or sitting as domestic work or office work with restriction of physically strenuous activities ~grade 2: ability to walk and appropriately achieve self-care with restriction of work or office activities wherein subject is mobile for > 50% of waking hours ~grade 3: capacity for limited individual care, incriminated subjects confined to bed or chair for > 50% of waking hours ~grade 4:completely disabled subjects lacking capacity for individual care and comprehensively confined to bed or chair ~ grade 5: subject ceases to exist. A decimated grade is indicative of superior functional status [2,3].

Karnofsky Performance Scales (KPS) are designated as ~grade 100: normal subjects lacking disease specific evidence and absence of complaints ~grade 90: ability to continue normal daily physical activities with minor symptoms of disease ~grade 80: ability to continue normal activities with effort, certain disease symptoms may ensue ~grade 70: incriminated subject capable of individual care although unable to continue normal daily activities or active work ~grade 60: incriminated subject requires occasional assistance although appears capable of individual care ~grade 50: incriminated subject necessitates considerable assistance and frequent medical care ~grade 40: incriminated subjects is disabled and necessitates special care and assistance ~grade 30: severely disabled individual with indications for hospitalization although mortality may ensue within distant future ~grade 20: extremely sick individual requiring hospitalization and active therapeutic

intervention ~grade 10: incriminated subject is moribund and approaching fatality with rapidly progressive, fatal disease processes ~grade 0: incriminated subject ceases to exist.

Elevated grade appears to indicate superior functional status [2,3].

T acute lymphoblastic leukaemia/ lymphoma is immune reactive to CD1a, CD2, cCD3(cytoplasmic), CD5, CD7 or terminal deoxynucleotidyl transferase (TdT). Immune reactive CD4+/CD8+ is encountered within ~one fifth (22%) of instances [3,4]. The neoplasm is variably immune reactive to CD10, CD13, CD16, CD33, CD56, CD57, CD79a or CD117. Immune reactive CD117a is associated with activating mutations within FLT3 genes, CD4 and CD8. T acute lymphoblastic leukaemia/ lymphoma is configured of lymphoblast immune reactive to CD3 , CD99, terminal deoxynucleotidyl transferase (TdT) or CD7. Variable expression of T cell markers as CD1a, CD2, CD4, CD5, CD8, CD34, CD10 and CD4/CD8 is exemplified.

T acute lymphoblastic leukaemia/ lymphoma is immune non-reactive to CD19, CD20, human leucocyte antigen DR (HLA-DR), surface immunoglobulin, CD22 or CD25 and preponderantly demonstrates a CD4-/CD8- immuno-phenotype [3,4].

T acute lymphoblastic leukaemia requires segregation from conditions such as Burkitt's leukaemia, blastoid variant of mantle cell lymphoma and thymoma [3,4]. T acute lymphoblastic leukaemia/ lymphoma may be appropriately alleviated with chemotherapy (~ 60%). However, in contrast to pre B acute lymphoblastic leukaemia, subjects may delineate preliminary disease reoccurrence, therapeutic induction failure or isolated tumour reoccurrence within the central nervous system (CNS) [3,4]. Superior prognostic outcomes may ensue with overexpression of TLX1(HOX11+), especially within incriminated adult subjects. Factors contributing to inferior prognostic outcomes are designated as ~expression of CFLAR, NOTCH2 or BTG3 genes and 3+ methylated genes ~minimal residual disease following commencement of treatment ~paediatric subjects depicting T acute lymphoblastic leukaemia ~occurrence of TAL1+ and LYL1+(3,4). In contrast to B acute lymphoblastic leukaemia, T acute lymphoblastic leukaemia exemplifying clinical features indicative of disease progression demonstrate augmented disease associated mortality. However, scenarios devoid of aforesaid clinical features exhibit concordant proportionate mortality between T acute lymphoblastic leukaemia and B acute lymphoblastic leukaemia [3,4].

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5. Image 1 and 2 Courtesy: Doctorlibinfo.com