

## Leukaemia Section

### Short Communication

## T-cell/histiocyte-rich large B cell lymphoma

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Published in Atlas Database: July 2010

Online updated version : <http://AtlasGeneticsOncology.org/Anomalies/TrichLargBCLID2071.html>

DOI: 10.4267/2042/45004

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### Clinics and pathology

#### Disease

T-cell/histiocyte-rich large B cell lymphoma (THRLBCL) is a distinct entity of aggressive lymphoma, recognized by the WHO classification as a separate entity (2008), in which few scattered neoplastic cells (usually <5%) are surrounded by reactive T-lymphocytes and histiocytes. THRLBCL shares several morphological and immunophenotypic similarities with nodular lymphocyte-predominant Hodgkin's Lymphoma (Pittaluga et al., 2010). These two entities may share initial transforming events that occur at germinal center B cell, followed by early divergence in the evolution of the neoplastic process (Franke et al., 2002).

#### Phenotype/cell stem origin

The postulated normal counterpart is a germinal centre B cell.

The immunophenotype of the neoplastic component in pan-B positive, BCL6+, CD15-, CD30-.

#### Epidemiology

It accounts for a minority of diffuse large B-cell lymphoma (<10%).

#### Clinics

The disease runs an aggressive course and is usually associated with poor outcome in those patients presenting at an advanced stage.

#### Pathology

The lymph node section shows scattered large cells surrounded by many lymphocytes and histiocytes. The disease must be distinguished from nodular

lymphocyte-predominant Hodgkin's lymphoma, which has distinct clinical features.

#### Treatment

Chemoimmunotherapy using anti CD20 monoclonal antibody rituximab in combination with CHOP or CHOP-like regimens is the standard of care (El Weshi et al., 2007).

#### Prognosis

A >80% overall response rate was obtained by chemoimmunotherapy, with a 5-year overall survival of approximately 50% (El Weshi et al., 2007).

### Cytogenetics

#### Cytogenetics molecular

Because of the low number of neoplastic cells, there are technical problems in obtaining evaluable metaphase chromosomes in THRLBCL. Tetraploid clones with complex aberrations, including a 14q32 translocation, were described in the absence of BCL6, BCL2 or c-myc involvement (La Starza et al., 1996; Wang et al., 2005; de Leval et al., 2006).

The presence of the PAX5/IGH gene rearrangement was demonstrated by fluorescence in situ hybridization (FISH) to represent a recurrent aberration (Poppe et al., 2005).

An average of 4.7 genomic imbalances/case were detected by comparative genomic hybridization in 17 cases of THRLBCL. The most frequently detected imbalances included gain of Xq (59%, minimal overlapping region Xq12q13), 4q (41%, minimal overlapping region 4q25q26), Xp (29%, minimal overlapping region Xp21p11), and 18q (24%, minimal overlapping region 18q21), as well as loss of 17p (24%) (Franke et al., 2002).

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- This article should be referenced as such:*
- Cuneo A, Rigolin GM, Cavazzini F. T-cell/histiocyte-rich large B cell lymphoma. *Atlas Genet Cytogenet Oncol Haematol.* 2011; 15(4):358-359.
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