Leukaemia Section

Short Communication

T-cell/histiocyte-rich large B cell lymphoma

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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).
References


This article should be referenced as such: