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

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Definition

T-cell/histiocyte-rich large B-cell lymphoma (THRLBCL) is characterized by a limited number of scattered, large B cells embedded in a background of abundant T cells and histiocytes. THRLBCL may arise de novo; however, more recent data suggest the possibility of a closer relationship with progression forms of nodular lymphocyte predominant Hodgkin lymphoma (NLPHL) than previously thought, indicating that NLPHL may proceed to or contain areas indistinguishable from THRLBCL. In small biopsies in particular, differentiating between a progression form of NLPHL (i.e. NLPHL with THRLBCL-like transformation) and de novo THRLBCL may be difficult, if not impossible.

ICD-O code

9688/3

Synonyms

T-cell–rich B-cell lymphoma; B-cell lymphoma rich in T cells and simulating Hodgkin disease; histiocyte-rich/T-cell–rich large B-cell lymphoma; T-cell–rich large B-cell lymphoma; T-cell–rich/histiocyte-rich large B-cell lymphoma; histiocyte-rich large B-cell lymphoma

Epidemiology

THRLBCL mainly affects middle-aged men. It accounts for < 10% of all DLBCLs.

Localization

THRLBCL mainly affects the lymph nodes, but bone marrow, liver, and spleen involvement is frequently found at diagnosis.

Clinical features

Patients present with fever, malaise, splenomegaly, and/or hepatomegaly. At diagnosis, almost half of cases are at an advanced Ann Arbor stage, with an intermediate-risk to high-risk International Prognostic Index (IPI) score (Table 13.24). The disease is often refractory to the chemotherapy regimens currently in use.

Imaging

Because of the important differential diagnosis with NLPHL, clinical staging procedures (including imaging analyses) are important. NLPHL usually affects one or two regions, whereas THRLBCL frequently manifests as systemic disease. Because THRLBCL is more PET-avid than is NLPHL, staging procedures such as FDG-PET and CT may facilitate the differential diagnosis {246}.

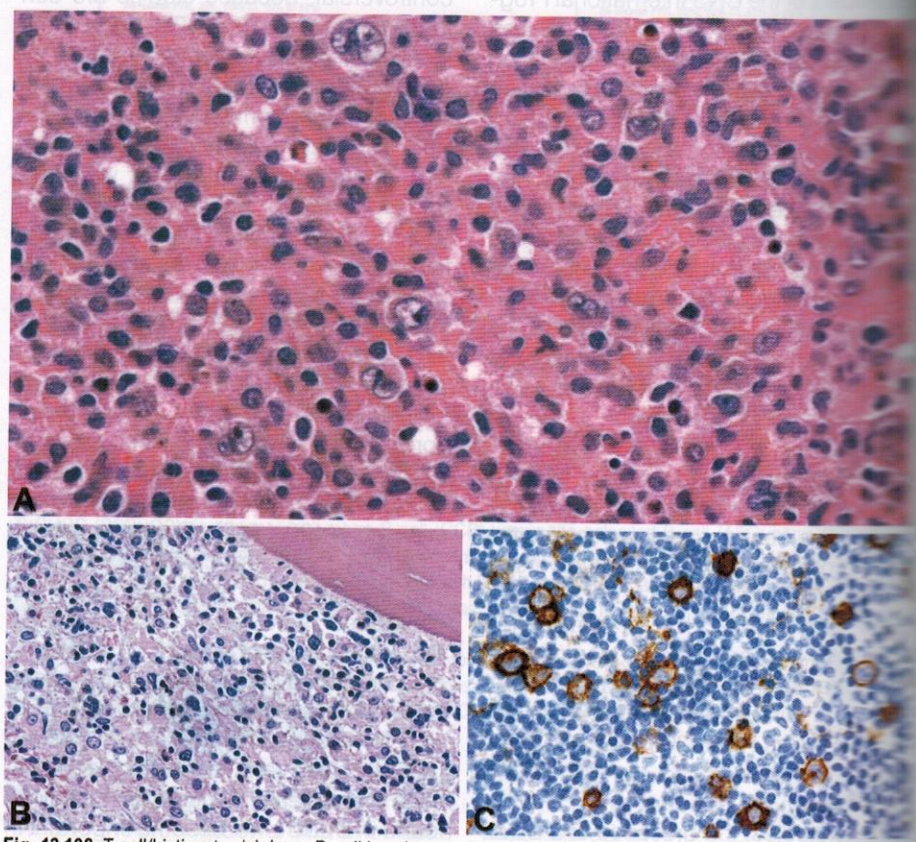

Fig. 13.108 T-cell/histiocyte-rich large B-cell lymphoma. **A** Lymph node. **B** Bone marrow. **C** CD20 stain highlights the large neoplastic B cells.

Microscopy

THRLBCL has a diffuse or less commonly vaguely nodular growth pattern replacing most of the normal lymph node parenchyma. It is composed of scattered, single large B cells embedded in a background of small T cells and variable numbers of histiocytes. The tumour cells are always dispersed and do not form aggregates or sheets. These cells may mimic the neoplastic lymphocyte predominant (LP) cells of NLPHL, but usually show greater variation in size and, in some cases, may resemble centroblasts or more pleomorphic cells, mimicking Reed-Sternberg or Hodgkin cells {19,3299}. They are typically found within clusters of bland-looking non-epithelioid histiocytes that may not be obvious on conventional examination. These histiocytes are a main and distinctive component of THRLBCL and are useful for the diagnosis {1448}. Nearly all of the background lymphocytes are of T-cell lineage, with typically only very few scattered B cells. Meshworks of follicular dendritic cells are absent. Eosinophils and plasma cells are not found. De novo THRLBCLs are usually diffuse and do not show the typical small B-cell background of NLPHL. However, there are cases of NLPHL in which the small B cells are diminished in number, and in

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