Angioimmunoblastic T-cell lymphoma

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Identity

Alias
Angioimmunoblastic lymphadenopathy with disprotidemia.

Clinics and pathology

Phenotype/cell stem origin
The lymphoma cell is a peripheral T lymphocyte in various stages of differentiation. The neoplastic clone expresses T-cell antigens and is usually CD4+. The malignant T-cells are believed to secrete cytokines responsible for the polyclonal B-cell hyperplasia observed in involved nodes. Clonality studies demonstrated a monoclonal rearrangement of the β-chain of the T-cell receptor (TCR) in the majority of cases. In some cases clonality could not be demonstrated. This led some authors to postulate the existence of at least two types of AILD, namely a reactive and benign type and a lymphomatous form.

Etiology
The disease is rare.

Clinics
The disease preferentially affects elderly males (male-to-female ratio 3:1, median age around 60 years). Most patients present with generalized lymphadenopathy, hepatosplenomegaly, skin rash and general symptoms (fever, weight loss). Polyclonal hypergammaglobulinemia is a common finding.

Pathology
The lymph node architecture is effaced and no reactive germinal centres are usually observed. The infiltrate may involve the perinodal fat. There is a proliferation of high endothelial venules with clusters of follicular dendritic cells. The lymphoid infiltrate consists of small-to-large cells resembling immunoblasts and atypical clear cells with round nucleus and abundant pale cytoplasm. The latter cells may occur in small aggregates or sheets.

Treatment
Some patients respond to steroids; in steroid-unresponsive patients multiagent chemotherapy usually produces short lasting responses.

Evolution
Few patients present spontaneous or steroid-induced remission; the majority of cases feature an aggressive disease with short survival despite chemotherapy. Most patients die with infection and active disease.

Prognosis
Median survival is about 1-3 years.

Cytogenetics

Note
A mixture of normal and abnormal cells is usually seen in the vast majority of cases. The cytogenetic picture at disease presentation may be normal in some cases which may develop clonal abnormalities during the course of the disease. The following karyotype pattern can be found.

Cytogenetics morphological
Clonal abnormalities defining a stemline, with one or more sidelines (approximately 30-50% of the cases). Normal karyotype (10-30% of the cases). Single cells with unrelated chromosome anomalies (10-20%). Unrelated clones with aberrant karyotypes, each carrying single unrelated additional anomalies (10-20%). Recurrent chromosome changes in those cases with an abnormal clone include trisomy 3, trisomy 5 and
trisomy X A 14q+ chromosome is a recurrent structural defect. Recurrent breakpoints include 1p31-32; 3p24-25; 4p13; 9q21-22; 12q13; 14q11; 14q32

The presence of abnormal metaphases in unstimulated cultures was associated with failure to respond to therapy and with shorter survival, as was the case with +X, structural aberrations of chromosome 1, and complex karyotype. The latter cytogenetic parameter maintained prognostic predictivity at multivariate analysis.

Cytogenetics molecular

Using probes for the detection of +3, +5 and +X, the vast majority of cases can be shown to carry aneuploidy.

References


This article should be referenced as such: