t(6;14)(p25.3;q11.2) TRA/IRF4

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Clinics and pathology

Disease
Peripheral T-cell lymphoma, not otherwise specified (PTCL, NOS)

Phenotype/cell stem origin
Mature (peripheral) cytotoxic alpha-beta T-cell origin.

Etiology
No etiologic factors are known.

Epidemiology
Adult males (age range, 67-82 years).

(A) Atypical lymphocytes in bone marrow smear from patient with PTCL, NOS with t(6;14)(p25.3;q11.2) (Wright-Giemsa, original magnification x1000). (B) Bone marrow trephine biopsy (H&E, x400). Tumor cells are (C) positive for CD2, (D) negative for CD5, (E) positive for granzyme B, and (F) positive for nuclear IRF4/MUM1 (x400).
Clinics
Presentation with cytopenias in the absence of lymphadenopathy, sometimes with skin involvement.

Pathology
The bone marrow is hypercellular with the normal marrow elements replaced by an extensive infiltrate of atypical, mostly medium-sized lymphoid cells with irregular nuclear outlines. Admixed plasma cells and a background of reticulin fibrosis are present. The tumor cells display an abnormal T-cell phenotype with expression of CD3, the cytotoxic marker TIA1 (+/− granzyme B), and T-cell receptor-beta (beta-F1), but without coexpression of CD5. Most cases express CD4 and lack expression of CD25 and CD30. IRF4/MUM1 protein is expressed in tumor cell nuclei. Cases tested for EBV by in situ hybridization have been negative.

Treatment
No treatment data are available.

Prognosis
The prognosis has not been established.

Cytogenetics
The rearrangement can be detected in standard G-banded karyotype.

Cytogenetics molecular
The rearrangement can be detected using dual-fusion fluorescence in situ hybridization with probes to the IRF4 and TCR@ loci.

Genes involved and proteins

IRF4
Location
6p25.3