Opis choroby *

Definicja

A rare mature T-cell neoplasm characterized by proliferation of small to medium-sized prolymphocytes with a mature post-thymic T-cell phenotype, involving the peripheral blood, bone marrow, lymph nodes, liver, spleen, and sometimes the skin. T-cell receptor genes are clonally rearranged. Patients typically present with hepatosplenomegaly, generalized lymphadenopathy, high leukocyte count with normal serum immunoglobulins, anemia, and thrombocytopenia. HTLV-1 serology is negative. The disease course is aggressive with generally poor prognosis.

Dane

Klasyfikacja
Choroba

Synonimy
T-PLL
Przewlekła białaczka limfocytarna z komórek T
T-plL
T-cell chronic lymphocytic leukemia

Kod ORPHA
86871

Kod OMIM
-

Kod ICD10
C91.6

Kod ICD11
2A90.0

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*Źródło

orphanet