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Short Communication

Characteristics of T-cell Prolymphocytic Leukemia

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DESCRIPTION

T-Cell Prolymphocytic Leukemia (T-PLL) is an aggressive T-cell leukemia that involves the blood, bone marrow, lymph nodes, liver, spleen, and skin. T-PLL is an extremely rare leukemia which mostly affects adults over the age of 30. It represents for 2% of all adult small lymphocytic leukemia's. Other names for T-cell lymphocytic leukemia include "knobby" T-cell leukemia and T-prolymphocytic leukemia/T-cell lymphocytic leukemia [1].

Symptoms and signs

T-cell prolymphocytic leukemia typically presents with systemic disease, including enlargement of the liver and spleen, extensive enlargement of the lymph nodes, and skin infiltrates. Leukemic cells can be found in peripheral blood, lymph nodes, bone marrow, spleen, liver, and skin due to the disease's systemic nature [2,3]. A high lymphocyte count (>100 x 10^9/L) and low levels of red blood cells and platelets in the blood are common findings. Serologies of HTLV-1 are negative, and serum immunoglobins are within normal ranges with no paraproteins present. The initiating cell line for T-cell prolymphocytic leukemia is assumed to be a mature (post-thymic) T-cell.

Diagnosis: T-PLL is consists of medium-sized lymphocytes with single nucleoli and basophilic cytoplasm with occasional blebs or projections in the peripheral blood. The nuclei are usually round to oval in shape, with the exception of few patients who have cells with a more irregular nuclear outline, similar to the cerebriform nuclear shape seen in Sezary syndrome. A tiny cell variant provides for 20% of all T-PLL cases, and the Sezary cell-like (cerebriform) variant account for 5%. Bone marrow involvement is typically diffuse, with morphology similar to peripheral blood [4]. Leukemic cells infiltrate both the red and white pulps of the spleen, and lymph node involvement is typically diffuse through the paracortex. Skin infiltrates are seen in 20% of patients, and they are often dense and confined to the dermis and skin appendages.

T-PLL has the immunophenotype of a mature (post-thymic) T-lymphocyte, with the neoplastic cells is positive for pan-T antigens CD2, CD3, and CD7 and negative for TdT and CD1a. The immunophenotype $CD4^+/CD8^-$ is present in 60% of cases, the immunophenotype $CD4^+/CD8^+$ is present in 25% of cases, and the immunophenotype $CD4^-/CD8^+$ is present in 15% of cases.

Genetic discoveries: Clonal TCR gene rearrangements for the chains are frequent. The most common chromosomal abnormality is chromosome 14 inversion, or inversion of 14(q11;q32). This occurs in 80% of cases, with the other 10% showing a reciprocal translocation of chromosome 14 (t(14;14)(q11;q32)). In addition, 75% of patients have chromosome 8 abnormalities, including such idic (8p11), t(8;8)(p11-12;q12), and trisomy 8.

Treatment: The majority of people with T-cell prolymphocytic leukemia require immediate treatment. T-cell prolymphocytic leukemia is difficult to treat, and most available chemotherapeutic drugs do not respond. Purine analogues pentostatin, fludarabine, cladribine, chlorambucil, and other forms of combination chemotherapy regimens, including cyclophosphamide, doxorubicin, vincristine, prednisone (CHOP), etoposide, and bleomycin, have all been used with limited success in some patients (VAPEC-B) [5]. Alemtuzumab (Campath), an anti-CD52 monoclonal antibody that attacks white blood cells, has been shown to be more effective in treatment than previous options. People who had a complete response to alemtuzumab survived a median of 16 months after treatment in one study of previously treated people with T-PLL [6].

CONCLUSION

In some of patients who respond to well treatment may also benefit from stem cell transplantation to enhance the response. T-PLL is an extremely rare, aggressive disease in which patients are unable to live normal lives. Prior to the recent introduction of new treatments, such as alemtuzumab, the median survival time after diagnosis was 7.5 months. Some patients have recently survived five years or more, whereas the median survival remained poor. For every three women, four men are diagnosed with this disease. Despite its rarity, mature T cell leukemia is the most common type.

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REFERENCES

1. Elaine SJ, Nancy Lee H. Pathology and genetics of tumours of haematopoietic and lymphoid tissues. World Health Organization Classification of Tumors IARC Press (2001).
2. Matutes E, Brito-Babapulle V, Swansbury J, J Ellis, R Morilla, C Dearden, et al. Clinical and laboratory features of 78 cases of T-prolymphocytic leukemia. Blood. 1991;78(12):3269-3274.
3. Matutes E, Garcia Talavera J, O'Brien M, Catovsky D. The morphological spectrum of T-prolymphocytic leukaemia. Br J Haematol. 1986;64(1):111-124.
4. Brito-Babapulle V, Catovsky D. Inversions and tandem translocations involving chromosome 14q11 and 14q32 in T-prolymphocytic leukemia and T-cell leukemias in patients with ataxia telangiectasia. Cancer Genet Cytogenet. 1991;55(1):1-9.
5. Maljaei SH, Brito-Babapulle V, Hiorns LR, Catovsky D. Abnormalities of chromosomes 8, 11, 14, and X in T-prolymphocytic leukemia studied by fluorescence *in situ* hybridization. Cancer Genet Cytogenet. 1998;103(2):110-116.
6. Sorour A, Brito-Babapulle V, Smedley D, Yuille M, Catovsky D. Unusual breakpoint distribution of 8p abnormalities in T-prolymphocytic leukemia: a study with YACS mapping to 8p11-p12. Cancer Genet. 2000;121(2):128-132.