

Leukaemia Section

Short Communication

Hepatosplenic T-cell lymphoma (HSTCL)

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Identity

Alias

Hepatosplenic Gamma delta T-cell lymphoma; Diffuse small cleaved cell lymphoma, unclassified (WF classification); Pleomorphic small cell lymphoma, medium size cell (HTLV-1 negative) (Kiel classification).

Clinics and pathology

Phenotype/cell stem origin

This lymphoma entity originates from T-lymphocytes expressing the Gamma Delta subunits of the TCR. Rarely, cases expressing the Alpha Beta subunits were reported.

Epidemiology

Young males are affected predominantly.

Clinics

The disease presents with hepatosplenomegaly, in the absence of lymphadenopathy. Bone marrow involvement and cytopenias are frequently encountered (Cooke et al., 1996).

Pathology

The proliferation consists of medium-sized lymphocytes with a rim of pale cytoplasm. The nuclei show condensed chromatin with inconspicuous nucleoli (Feldman et al., 2006). Typically these cells show an intrasinusoidal pattern of growth sparing the portal triads and the white pulp. Intrasinusoidal bone marrow involvement may occur. The neoplastic lymphocytes are CD3+, CD4- and may express CD8 and CD56. These features, along with negativity for granzyme B and for perforin, indicate a proliferation of Gamma delta resting T lymphocyte.

Treatment

Multiagent chemotherapy, including anthracyclines is the treatment of choice. Autologous or allogeneic transplantation may have a role in selected patients (Cooke et al., 1996).

Prognosis

The patients usually respond to chemotherapy, but relapses occur frequently and median survival is around three years. Allogeneic BMT may cure some patients.

Cytogenetics

Cytogenetics morphological

Extra-copies of the long arm of chromosome 7q deriving from one or more isochromosome 7q are frequently found in this lymphoma. Trisomy 8 may also occur. Multiple copies of 7q were also documented by FISH. An increased number of 7q signals was found in cases with cytologic features of progression, indicating a tendency of HSTCL to multiply the i(7)(q10) chromosome during evolution. (Wlodarska et al., 2002).

References

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