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

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Clinics and pathology

Disease
Peripheral T-cell lymphomas not otherwise specified (PTCL-NOS) include a heterogeneous group of diseases involving lymph nodes and extra nodal sites deriving from the clonal expansion of mature T-lymphocytes bearing clonally rearranged TCR genes.

Phenotype/cell stem origin
The cell of origin is an activated mature CD4+ lymphocyte. The phenotype is usually CD4+/CD8-, TCRβ+ whereas the expression of CD7 and CD5 may be low. Occasionally, CD30 may be positive.

Epidemiology
There is geographic variation in the incidence of T-cell lymphoma. PTCL-NOS accounts for approximately 4-7% of all non Hodgkin's lymphomas and for 30-70% of all mature T-cell lymphomas.

Clinics
The disease runs an aggressive clinical course.

Pathology
The proliferation effaces the lymph node architecture, with paracortical or diffuse growth pattern. The cells are medium-to-large sized, with irregular nucleus, distinct nucleoli. Mitotic figures may be numerous.

Treatment
Anthracycline-based regimes such as CHOP yields unsatisfactory results with lower CR rates than in B-cell diffuse large cell lymphomas and high relapse rate.

Intensive regimens such as hyperCVAD with or without autologus bone marrow transplantation may be effective in this type of lymphoma, though the superiority of this approach over conventional treatment has not been definitely proven.

Prognosis
Reported failure free survival rates ranged between 12 and 45% (Armitage, 2006).

Cytogenetics

Cytogenetics molecular
Complex karyotypes are reported in 70-90% of the cases (Rizvi et al., 2006). Recurrent chromosome gains were described to involve 7q, 8q, 17q and 22q, whereas recurrent regions of loss of chromosome material were represented at 4q, 5q, 6q, 9p, 10q, 12q and 13q (Pileri et al., 2008). In a recent study, frequent gains involved 7q22q31 (33%), 1q (24%), 3p (20%), 5p (20%) and 8q24qter (22%). Losses occurred at 6q22q24 (26%) and 10p13pter (26%). Complex karyotypes were predictive of an inferior outcome, but no association was noted between specific aberrations and survival (Nelson et al., 2008).

Array comparative genomic hybridization (CGH) for high-resolution analysis of PTCL-NOS identified a region with high copy number gain at 14q32.2, and a region with homozygous loss at 9p21.3. Gains of 7p and 7q and loss of 9p21.3 showed a significant association with poor prognosis (Nakagawa et al., 2009).

p53 protein overexpression and mutation of p53 may be found in 30% of the cases and may correlate...

Significantly with treatment failure and worse overall and disease-free survival (Pescarmona et al., 2001). Recurrent copy number gain may also involve chromosomes 8, 9 and 19. Other genomic imbalances may include overexpression of CARMA1 at 7p22 and of MYCBP2 at 13q22, both genes being localized within regions of frequent copy number gain. LOH was found at 2q34 (Fujiwara et al., 2008).

References


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