Nasal T cell lymphoma

Antonio Cuneo, Francesco Cavazzini, Gian Matteo Rigolin

Hematology Section, Dept. Of Biomedical Sciences, University of Ferrara, Ferrara Italy (AC, FC, GMR)

Published in Atlas Database: May 2008
Online updated version: http://AtlasGeneticsOncology.org/Anomalies/NasalTCellLymphomaID2100.html
DOI: 10.4267/2042/44475

This work is licensed under a Creative Commons Attribution-Noncommercial-No Derivative Works 2.0 France Licence.
© 2009 Atlas of Genetics and Cytogenetics in Oncology and Haematology

Identity

Alias: Angiocentric T-cell lymphoma; Polymorphic reticulosis; Lethal midline granuloma

Clinics and pathology

Disease
Extranodal NH/T-cell lymphoma, nasal type.

Phenotype/cell stem origin
This lymphoma derives from the transformation of NK lymphocytes and, less frequently, T-lymphocytes.

Epidemiology
It is seen most frequently in China, Japan, Korea and other Asian countries and in Central America.

Clinics
Middle aged adults are most frequently affected, with slight male predominance. The disease involves the nasal cavity and may spread to the pharynx, palate and larynx. Less frequently, orbital and cranial nerve involvement was described. Spreading to the skin, soft tissue and gastrointestinal tract may occur. Bone marrow involvement is unusual at presentation. Hemophagocytic syndrome was described in some cases. The tumor is locally invasive and destructive (Liang et al., 2006).

Pathology
The cellular infiltrate is polymorphic, typically associated with an angioinvasive growth pattern, with consequent angiodestruction, ischemia and tissue necrosis. Neoplastic cells are CD56 positive, with negativity for surface CD3. The TCR gene is usually germline, even though some cases with a clonally rearranged TCR were reported (Yoon et al., 1999).

Epstein-Barr virus infection in this lymphoma was well documented by molecular methods (Chiang et al., 1997).

Treatment
Combination regimens such as CHOP or other aggressive schedules followed by local radiotherapy are the mainstay of treatment. Autologous bone marrow transplantation has been used.

Prognosis
Prognosis is severe, with less than 50% of the patients achieving durable complete response after intensive chemotherapy and local radiotherapy. The disseminated forms of the disease are almost uniformly fatal.

Cytogenetics

Cytogenetics morphological
Three out of seven cases studied by Wong et al (1997), including one nasal, one extranasal and one leukemic form, showed a common region of deletion at 6q21-q25, suggesting that this may be a nonrandom chromosomal aberration. Other non-random abnormalities include +X, i(1q), i(7q), +8, del(13q), del(17p), i(17q), and 11q23 rearrangement (Wong et al., 1999).

Cytogenetics molecular
P73 gene methylation was described in 94% of the cases; other methylated genes included hMLH1 (63%), p16 (63%), p15 (48%), and RAR beta (47%) (Siu et al., 2002). P53 gene overexpression was documented (Quintanilla-Martinez et al., 1999).

Comparative genomic hybridization studies identified del(6q), del(13q), del(17p), del(1p), del(12q), and partial gain of Xp, 2p, 10q as recurrent abnormalities (Siu et al., 1999; Ko et al., 2001). Some of these
abnormalities (i.e. 17p deletion and 1p deletion) may be associated with the aggressive leukemic variant of the disease (Nakashima et al., 2005). Genome-wide array-based comparative genomic hybridization identified recurrent regions of imbalances: gain of 2q and loss of 6q16-27, 11q22-23, 5p14, 5q34, 1p36, 2p16, 4q12, 4q31 (Nakashima et al., 2005).

References


Wong KF, Zhang YM, Chan JK. Cytogenetic abnormalities in natural killer cell lymphoma/leukaemia--is there a consistent pattern? Leuk Lymphoma. 1999 Jul;34(3-4):241-50


This article should be referenced as such: