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Leukaemia Section

Short Communication

Intestinal T-cell lymphoma

Antonio Cuneo, Francesco Cavazzini, Gian Matteo Rigolin

Hematology Section, Dept. of Biomedical Sciences, University of Ferrara, 44100 Ferrara, Italy (AC, FC, GMR)

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Identity

Alias : Enteropathy-type T-cell lymphoma

Clinics and pathology

Phenotype/cell stem origin

The disease originates from a CD3+, CD7+ T-lymphocyte lacking CD4 and CD8 expression.

Epidemiology

The disease affects mainly the adult, with male predominance, and it is frequently associated with gluten-sensitive entheropathy.

Pathology

The disease consists of ulcerated lesions involving the small intestine. Perforation may occur. Small and larger atypical lymphocytes with pale cytoplasm infiltrate the epithelial mucosa of the villi. The TCR-Beta and TCR-Gamma genes are clonally rearranged.

Treatment

Multiagent chemotherapy (CHOP or CHOP-like regimes) was used.

Evolution

The disease may derive from patients with coeliac disease not responding to gluten-free diet. The lymphoma may spread to regional lymph nodes.

Prognosis

Response to chemotherapy is suboptimal and patients are vulnerable to toxicity of treatment due to intestinal symptoms and malnutrition preceding the diagnosis of lymphoma. Survival at 2 years was 28% in a study (Daum et al., 2003).

Cytogenetics

Cytogenetics molecular

Extra copies of chromosome 9q centered around the 9q33-34 region was detected by CGH and FISH studies (Zettl et al., 2002). 9p deletion with p16 loss was found in 18% of the cases, and loss of heterozygosity at 9p21 with loss of p16 expression was documented in approximately half of those cases with a large cell component (Obermann et al., 2004). DNA gains may involve the 5q33-34 and 7q31 regions, with a 30% frequency. Loss of chromosome material was detected at 6p24; 7p21, 17q23-25 and 17p13 (Baumgartner et al., 2003).

References

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