

Leukaemia Section

Mini Review

Primary cutaneous CD30+ anaplastic large cell lymphoma

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Identity

Note

Formerly referred to as "regressive atypical histiocytosis".

Clinics and pathology

Phenotype/cell stem origin

This entity is regarded as a T-cell lymphoma. The neoplastic cells express the CD30 antigen, and show variable expression of T-cell markers. Clonal rearrangement of the T-cell receptor is usually found in approximately 60% of the cases.

Epidemiology

It accounts for approximately 9% of all cutaneous lymphomas. Median age at onset is 60 years.

Pathology

Usually the disease arises de novo in the skin and presents with a solitary reddish nodule, which may become ulcerated. Rarely, distinct nodules may be appreciated in a cutaneous area, or in multiple sites. The risk of disease dissemination at extracutaneous sites is very low in the presence of a solitary nodule. The neoplastic nodule is composed of large anaplastic cells with pleomorphic nucleus. Some multinucleated cells are also present, resembling Reed-Sternberg cells. Reactive neutrophils and macrophages infiltrate the lymphomatous lesion. Unlike systemic CD30+ anaplastic large cell lymphoma, the expression of the ALK protein is not found in primary cutaneous tumours.

Treatment

Approximately 25% of the cases may show spontaneous regression of the cutaneous nodule. Surgical excision with or without subsequent radiotherapy to the involved cutaneous site is curative in the majority of cases. Patients at risk of developing recurrence at extracutaneous sites (i.e. with multiple nodules at distant cutaneous sites) may require multiagent chemotherapy.

Prognosis

The prognosis is excellent in those patients with a solitary nodule undergoing complete surgical resection. Adequate systemic treatment usually attains durable complete responses in patients with disseminated disease, with a 90% overall survival at 5 years.

Cytogenetics

Note

The tumour cells are difficult to grow in culture and few data are available. The t(2;5), usually associated with systemic anaplastic large cell lymphoma, is not found in this disease. Single cases with variant ALK translocations were described. In patients with extranodal anaplastic large cell lymphoma, recurrent breakpoints occurred at 1p36; 6p25 and 8q24; a 6q-chromosome was found in one patient.

Cytogenetics molecular

Comparative genomic hybridization studies disclosed chromosomal imbalances in approximately 40% of the cases. The most frequent gains involved chromosome 1/1p and 5 (50% of the cases), 6,7,8/8p, and 19 (38%).

Gains involving chromosome 9 were also described. Other gains occurred less frequently at 3/3p, 7/7q, 10, 12, 13, 15q and 20/20p. Recurrent losses were detected at 6q21 and 18p11. There is a positive correlation between the presence of chromosomal imbalances and clonal T-cell receptor gene rearrangement. Recurrent oncogene copy number gains involve FGFR1 at 8p11; NRAS at 1p13.2, RAF1 at 3p25, >CC: TXT: MYCN ID: 112> at 2p24.1, CTSB at 8p22, FES at 15q26.1 and CBAF2 at 21q22. The majority of cases investigated by real time PCR revealed amplification of the following genes: CTSB, RAF1, REL at 2p13; JUNB at 19p13. Gains involving chromosome 9 and losses involving chromosome 6q and 18p were seen in relapsing patients.

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