

Leukaemia Section

Short Communication

Primary Cutaneous CD8 Aggressive Epidermotropic Cytotoxic T Cell Lymphoma

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Published in Atlas Database: November 2019

Online updated version : <http://AtlasGeneticsOncology.org/Anomalies/AggrEpidCytotoxTLymphomaID1753.html>

Printable original version : <http://documents.irevues.inist.fr/bitstream/handle/2042/70779/11-2019-AggrEpidCytotoxTLymphomaID1753.pdf>
DOI: 10.4267/2042/70779

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Abstract

Review on Primary Cutaneous CD8 Aggressive Epidermotropic Cytotoxic T Cell Lymphoma, with data on clinics, and the genes involved.

Keywords

Primary Cutaneous CD8+ Epidermotropic Cytotoxic T Cell Lymphoma.

Other names

CD8+ AECTCL

Clinics and pathology

Disease

Primary cutaneous CD8+ aggressive epidermotropic cytotoxic T cell lymphoma (AECTCL) had been considered a provisional disease until the 2016 World Health Organization Classification of Cutaneous lymphomas (Guitart J et al, 2017). It is a rare cutaneous lymphoma, representing less than 1% of all cutaneous lymphomas and it has aggressive behavior and a poor prognosis (Willemze R et al, 2005).

Etiology

AECTCL is a proliferation of epidermotropic CD8+ cytotoxic T cells with the expression of TIA-1 marker.

Epidemiology

AECTCL is rare, but most patients are adult with a median survival among 12 months (Berti E et al, 1999).

Clinics

The clinical course usually begins with a prodrome of localized or disseminated eruptive papules, nodules, and tumors with a central ulceration/necrosis or hyperkeratotic patches/plaques in the skin and cutaneous adnexa. The disease rapidly disseminates to organs such as lungs, testis, CNS and oral mucosa in just weeks or months (Willemze R et al, 2008). Lymph nodes are not infiltrated by the disease. B symptoms (fever, night sweats, and weight loss) are seen in most patients. Some cases may be associated with hemophagocytic syndrome (Toro JR et al, 2003).

Pathology

Histologically the disease shows infiltrates of T cells in full epidermal thickness with a different degree of spongiosis, intraepidermal blistering and necrosis. In the early stages a lichenoid pattern with pagetoid epidermotropism and subepidermal edema is seen. In advanced stages, diffuse dermal infiltrates in nodular and tumor-like lesions are characteristic. Epidermal necrosis and ulceration or destruction of skin structures are commonly found (Berti E et al, 1999; Santucci M et al, 2003; Robson A et al, 2015). The neoplastic cells showed a high Ki-67 proliferation index.

Immunophenotype AECTCL express a peripheral T cell phenotype with CD3+, CD8+, CD7+/-, CD45RA+, beta-F1+, Granzyme B+ (g-B), perforin (PF), T-intracytoplasmic antigen (TIA-1) and CD45RO-. CD2, CD4 and CD5 are frequently lost and CD30 is rarely expressed but they have a high

proliferation, expressed with Mib-1 marker. Epstein-Barr virus is negative (Robson A et al, 2015; Guitart J et al, 2017; Quintanilla-Martinez L et al, 2013). AECTCL express clonal T cell receptor gene rearrangements.

Genes

The neoplastic T cells that are representative of this disease show clonal T cell receptor gene rearrangements. Specific genes abnormalities have not been described.

Treatment

Treatment remains unclear since no randomized trials are available. Peripheral T Cell lymphoma combination chemotherapy is usually tried but response rates remain poor (Berti E et al, 1999).

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This article should be referenced as such:

Corazón-Monzón A, Juárez-Salcedo LM, Dalia S. Primary Cutaneous CD8 Aggressive Epidermotropic Cytotoxic T Cell Lymphoma. *Atlas Genet Cytogenet Oncol Haematol.* 2020; 24(8):323-323.
