Opis choroby *

Definicja

A rare dendritic cell tumor characterized by an aggressive, high-grade neoplasm derived from Langerhans cells, most commonly extranodal and multifocal, involving the skin and underlying soft tissue, as well as lung, liver, spleen, and bone. Primary nodal involvement is seen in a minority of patients. Immune-phenotyping and the presence of Birbeck granules on ultrastructural examination reveal the Langerhans cell derivation of the neoplastic cells. Prognosis is generally poor.

Dane

Klasyfikacja
Choroba

Kod ORPHA
86897

Kod OMIM
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Kod ICD10
C96.4

Kod ICD11
2B31.3

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*Źródło

orphanet