

Opis choroby *

Definicja

A rare hematologic neoplasm characterized by origin from precursors of plasmacytoid dendritic cells, with frequent cutaneous, bone marrow, and lymph node involvement, as well as leukemic dissemination. Most common clinical presentation is with asymptomatic solitary or multiple skin lesions (either isolated purplish nodules, isolated bruise-like papules, or disseminated purplish nodules/macules/papules), although some patients may present with leukemia. Skin biopsy shows a diffuse, monomorphous infiltrate of medium-sized blast cells resembling either lymphoblasts or myeloblasts, with massive involvement of the dermis. The clinical course is aggressive, and age has an adverse impact on prognosis.

Dane

Klasyfikacja

Choroba

Synonimy

BPDCN

BPDCN

Chłoniak z blastycznych komórek NK

Jednopostaciowy chłoniak z komórek NK

Limfoblastoidalny wariant chłoniaka z komórek

NK

Nowotwór z blastycznych plazmacytoidalnych

komórek dendrytycznych

Blastic NK-cell lymphoma

Blastic plasmacytoid dendritic cell neoplasm

Lymphoblastoid variant of NK-cell lymphoma

Monomorphic NK-cell lymphoma

Nowotwór z blastycznych plazmocytydnych

komórek dendrytycznych

Kod ORPHA

86870

Kod OMIM

-

Kod ICD10

C86.4

Kod ICD11

2A60.5

[*Źródło](#)

orpho:net