

Opis choroby *

Definicja

A rare dendritic cell tumor characterized by a neoplasm composed of spindle to ovoid cells with phenotypic features similar to those of interdigitating dendritic cells. Solitary lymph node involvement is common, although extranodal localization (in particular skin and soft tissue) has also been reported. Patients usually present with an asymptomatic mass, sometimes with systemic symptoms such as fatigue, fever, and night sweats. Generalized lymphadenopathy, splenomegaly, or hepatomegaly may be seen in rare cases. The clinical course is generally aggressive.

Dane

Klasyfikacja

Choroba

Synonimy

Interdigitating cell sarcoma
Reticulum cell sarcoma
Interdigitating cell sarcoma
Reticulum cell sarcoma

Kod ORPHA

86900

Kod OMIM

267730

Kod ICD10

C96.4

Kod ICD11

2B31.4

*Źródło

orphanet