

Opis choroby *

Definicja

A rare dendritic cell neoplasm characterized by a proliferation of spindle to ovoid cells with morphological and immunophenotypic features of follicular dendritic cells. Conventional follicular dendritic cell sarcomas are negative for EBV. The tumor arises as a painless, slow-growing mass in lymph nodes (most often cervical), extranodal sites (such as tonsils, gastrointestinal tract, soft tissue, mediastinum, or lung, among others), or both. Paraneoplastic pemphigus may occur in rare cases. Predictive factors are tumor size, presence of coagulative necrosis, mitotic count, and presence of significant cytological atypia.

Dane

Klasyfikacja

Choroba

Kod ORPHA

86902

Kod OMIM

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Kod ICD10

C96.4

Kod ICD11

2B31.5

*Źródło

orphonet