

Opis choroby *

Definicja

Intravascular large B-cell lymphoma (IVLBCL) is a very rare form of diffuse large B-cell lymphoma (see this term) characterized by the selective growth of lymphoma cells within the lumina of small blood vessels (especially the capillaries) that most often presents with a wide range of clinical manifestations (as potentially any tissue can be involved), with patients from Western countries more frequently manifesting with neurological and cutaneous symptoms while patients from Asian countries more frequently displaying hepatosplenomegaly and thrombocytopenia. IVLBCL is characterized by an absence of lymphadenopathy, an aggressive clinical course and a poor prognosis.

Dane

Klasyfikacja

Choroba

Synonimy

Angioendotheliomatosis proliferans systemisata

Angiotropowy chłoniak wielkokomórkowy

Choroba Tappeinera i Pfliegera

Limfomatoza wewnątrznaczyniowa

Złośliwa angioendoteliomatoza

Angiotropic large cell lymphoma

Intravascular lymphomatosis

Malignant angioendotheliomatosis

Tappeiner-Pfleger disease

Kod ORPHA

98839

Kod OMIM

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Kod ICD10

C83.3

Kod ICD11

2A81.1

*Źródło

orphanet