Opis choroby *

Definicja

A rare lymphatic system anomaly characterized by multifocal congenital and progressive vascular lesions of the skin, gastrointestinal tract, and occasionally other anatomic sites, causing potentially life-threatening thrombocytopenic coagulopathy. Macroscopically, the lesions appear as round to oval, red-brown plaques, as large as a few centimeters in diameter. Histopathologically, they consist of dilated, thin-walled vessels with variable endothelial hyperplasia, positive for lymphatic endothelial cell markers, and resembling benign lymphangioendothelioma.

Dane

Klasyfikacja
Choroba

Synonimy
Cutaneovisceral angiomatosis-thrombocytopenia syndrome
Limfangioendoteliomatoza wieloogniskowa z małopłytkowością
MLT
Zespół naczyniakowatości trzewno-skórnej i małopłytkowości
MLT
Multifocal lymphangioendotheliomatosis with thrombocytopenia

Kod ORPHA
464321

Kod OMIM
-

Kod ICD10
D18.1

Kod ICD11
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*Źródło

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