

Opis choroby *

Definicja

A rare histiocytic tumor characterized by a malignant proliferation of cells showing morphological and immunophenotypic features of mature tissue histiocytes. Most cases occur in extranodal sites, most commonly the intestinal tract, skin, and soft tissue. Patients may present with a solitary mass, lymphadenopathy, a skin rash or numerous tumors on the trunk and extremities, lytic bone lesions, hepatosplenomegaly with pancytopenia, intestinal obstruction, and/or systemic symptoms. The neoplasm is aggressive with typically poor therapy response.

Dane

Klasyfikacja

Choroba

Kod ORPHA

86896

Kod OMIM

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Kod ICD10

C96.8

Kod ICD11

2B31.1

*Źródło

orphanet