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
-

Kod ICD10
C96.8

Kod ICD11
2B31.1

---

*Źródło

orphanet