Opis choroby *

Definicja

A rare non-Langerhans cell histiocytosis characterized by infiltration of lymph nodes or extranodal tissues by non-malignant histiocytes displaying emperipolesis, a non-destructive phagocytosis of lymphocytes or erythrocytes. Most typical presentation is as a massive cervical lymphadenopathy in adolescents and young adults. Most frequent sites of extranodal disease are skin, soft tissue, bones, paranasal sinuses, orbit, salivary glands, and central nervous system. Symptoms are related to mass effect in the affected organs.

Dane

Klasyfikacja
Choroba

Synonimy
Destombes-Rosaï-Dorfman disease
Choroba Destombesa, Rosaï i Dorfmana
choroba Rosaï, Dorfmana i Destombesa
Histiocytoza zatokowa z masywną limfadenopatią
SHML
Rosaï-Dorfman-Destombes disease
SHML
Sinus histiocytosis with massive lymphadenopathy

Kod ORPHA
158014

Kod OMIM
-

Kod ICD10
D76.3

Kod ICD11
EK92

---

*Źródło

orphanet