Opis choroby *

Definicja

A rare soft tissue sarcoma characterized by a lesion in the deep soft tissues of the proximal extremities and limb girdles, composed of malignant chondroblast-like cells arranged in cords, clusters, or networks, and an abundant myxoid matrix. The tumor is typically encased by a pseudocapsule and divided into multiple nodules by fibrous septa. Patients present with a soft tissue mass which can be painful and may ulcerate the skin or restrict range of motion if located next to joints. Despite prolonged survival, local recurrence and metastasis are frequent.

Dane

Klasyfikacja
Choroba

Kod ORPHA
209916

Kod OMIM
612237

Kod ICD10
C49.9

Kod ICD11
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*Źródło

orphanet