

Opis choroby *

Definicja

A rare soft tissue sarcoma characterized by a malignant, fibroblastic lesion with variably myxoid stroma, pleomorphism, and a distinctively curvilinear vascular pattern. The majority of tumors arise in the limbs including the limb girdles, more often in dermal/subcutaneous tissues than in the underlying fascia and skeletal muscle, and usually present as a slowly growing, painless mass. Depth of the lesion and tumor grade do not influence the high rate of local recurrence, while the percentage of metastasis and tumor-associated mortality are much higher in deep-seated and high-grade neoplasms.

Dane

Klasyfikacja

Choroba

Synonimy

Fibromyxosarcoma

Śluzakowata złośliwa włóknista histiocytoma

Włókniakośluzakomięsak

Myxoid malignant fibrous histiocytoma

Kod ORPHA

79105

Kod OMIM

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Kod ICD10

C49.9

Kod ICD11

2B53.0

*Źródło

orphanet