

Opis choroby *

Definicja

A rare soft tissue tumor characterized by a slow-growing, usually painless, subcutaneous nodule, predominantly located in the extremities, less frequently the trunk or head and neck region. Histopathologically, the lesion is well-circumscribed, lobulated, and composed of epithelioid, ovoid, or spindle cells arranged in a nodular and often syncytial pattern, with pseudoangiomatoid spaces and a peripheral fibrous pseudocapsule with a prominent lymphoplasmacytic cuff. The tumor is most common in the first two decades of life and usually follows an indolent course, although local recurrence may occur, while metastasis is rare.

Dane

Klasyfikacja

Choroba

Synonimy

AFH

AFH

Kod ORPHA

569164

Kod OMIM

-

Kod ICD10

D21.9

Kod ICD11

-

*Źródło

orphanet