

Nerwiakowłókniak

Kod Orpha: 252183 Kod OMIM:

Opis choroby *

Definicja

A rare benign peripheral nerve sheath tumor characterized by a well-demarcated intraneuronal or diffusely infiltrative extraneuronal space-occupying lesion consisting of Schwann cells, perineurial-like cells, and fibroblasts. It presents as a cutaneous nodule, a circumscribed mass in a peripheral nerve, a plexiform enlargement of a major nerve trunk, or with diffuse but localized involvement of skin and subcutaneous tissue. Multiple neurofibromas are typically associated with neurofibromatosis 1. Malignant transformation occurs almost exclusively in plexiform neurofibromas and neurofibromas of major nerves.

Dane

Klasyfikacja

Choroba

Kod ORPHA

252183

Kod OMIM

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Kod ICD10

D36.1

Kod ICD11

2F3Y

*Źródło

orphanet

Rozszerzony opis choroby

Brak opisu rozszerzonego dla tej choroby. Opracowanie w toku.