Opis choroby *

Definicja

A rare benign peripheral nerve sheath tumor characterized by a well-demarcated intraneural or diffusely infiltrative extraneural 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
-

Kod ICD10
D36.1

Kod ICD11
2F3Y

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*Źródło

orphanet