Opis choroby *

Definicja

A rare nervous system tumor characterized by cells phenotypically representing Schwann cells but containing melanosomes and expressing melanoma markers. The entity occurs as a non-psammomatous (typically affecting spinal nerves) or a psammomatous (also involving nerves of the intestinal tract and heart) variant. About 50% of psammomatous tumors are associated with Carney complex. Slightly over 10% of melanotic schwannomas follow a malignant course.

Dane

Klasyfikacja Synonimy

Choroba Isolated melanocytic schwannoma

Isolated melanocytic schwannoma

 Kod ORPHA
 Kod OMIM
 Kod ICD10

 590539
 D36.1

Kod ICD11 2A02.1Y

*Źródło

orphanet