

Opis choroby *

Definicja

Malignant peripheral nerve sheath tumor with perineurial differentiation is a rare soft tissue sarcoma composed predominantly of spindle-shaped neoplastic cells showing perineurial differentiation and displaying abundant cellular pleomorphism or anaplasia, frequent mitoses, tumor necrosis and high metastatic potential. It often presents as a soft, painless, solid mass in subcutaneous tissues of the trunk or limbs, but tumors have also been described in the facial area, mediastinum, retroperitoneum, pancreas, paravertebral column and the pelvic soft tissues. Frequent local recurrence and distant metastatic spread has been reported.

Dane

Klasyfikacja Podtyp histopatologiczny	Synonimy Malignant perineurioma
--	---

Kod ORPHA 252128	Kod OMIM -	Kod ICD10 C47.9
----------------------------	----------------------	---------------------------

Kod ICD11
-

*Źródło

orphanet