Opis choroby *

Definicja

A rare neuroendocrine neoplasm of pancreas characterized by a high-grade malignant epithelial tumor with neuroendocrine differentiation. Based on histopathologic appearance, a small cell (composed of diffuse sheets of cells) and a large cell type (showing a nesting/trabecular pattern) are distinguished. Synaptophysin and chromogranin are positive on immunohistochemistry. The Ki-67 proliferation index is typically very high (>60 - 80%). Patients present with back pain, jaundice, and/or non-specific abdominal symptoms. Serum hormone activity is unusual. The tumor is highly aggressive with poor prognosis.

Dane

Klasyfikacja
Choroba

Synonimy
Pancreatic NEC
Pancreatic neuroendocrine carcinoma
Poorly-differentiated NEN of pancreas
Poorly-differentiated neuroendocrine neoplasm of pancreas
Poorly-differentiated pancreatic NEN
Poorly-differentiated pancreatic neuroendocrine neoplasm
Pancreatic NEC
Pancreatic neuroendocrine carcinoma
Poorly-differentiated NEN of pancreas
Poorly-differentiated neuroendocrine neoplasm of pancreas
Poorly-differentiated pancreatic NEN
Poorly-differentiated pancreatic neuroendocrine neoplasm

Kod ORPHA
506098

Kod OMIM
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Kod ICD10
C25.9

Kod ICD11
2C10.1

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*Źródło

orphanet