

Opis choroby *

Definicja

A rare soft tissue tumor characterized by distinctive perivascular epithelioid cells, often arranged radially around a vascular lumen, as well as spindled cells in variable proportion. Melanocytic and muscle markers are typically positive. The tumors have been reported in the uterus, fallopian ligament, and large and small intestine, among others. Depending on their location, they may present as a painful or painless mass, or with vaginal bleeding. Tumors displaying infiltrative growth, marked hypercellularity, nuclear enlargement and hyperchromasia, high mitotic activity, atypical mitotic figures, and/or coagulative necrosis should be regarded as malignant.

Dane

Klasyfikacja

Choroba

Synonimy

PEComa

Guz z nabłonkowatych komórek
przynaczyniowych

Perivascular epithelioid tumour

Kod ORPHA

595133

Kod OMIM

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Kod ICD10

D21.9

Kod ICD11

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*Źródło

orphanet