

Opis choroby *

Definicja

A rare soft tissue sarcoma characterized by a slowly growing, painless space-occupying lesion, composed of large, uniform, epithelioid cells arranged in solid nests and/or alveolar structures, separated by thin, sinusoidal vessels. The tumor mostly affects adolescents and young adults. Early metastasis, most commonly to the lung, bones, and brain, is a characteristic feature and relevant prognostic factor, together with age at presentation and tumor size, while histological features have no prognostic significance.

Dane

Klasyfikacja

Choroba

Synonimy

ASPS

Mięsak pęcherzykowy części miękkich

Alveolar soft part sarcoma

Kod ORPHA

163699

Kod OMIM

606243

Kod ICD10

C49.9

Kod ICD11

XH8V95

*Źródło

orphanet