Opis choroby *

Definicja

Progressive nodular histiocytosis is a rare, normolipemic, non-Langerhans cell histiocytosis characterized by progressive growth of multiple to disseminated, asymptomatic skin lesions that range in appearance from yellow plaques to coalescence-prone red-brown papules, nodules and pedunculated tumors up to 5 cm in size, located typically on the face, trunk and extremities (and rarely on conjuctiva and mucous membranes). Characteristic microscopic findings include a storiform spindle cell infiltrate in the deep dermis with xanthomatized macrophages and some Touton cells in the upper dermis. It is usually not associated with systemic disease.

Dane

Klasyfikacja

Choroba

Kod ORPHA

Kod OMIM

Kod ICD10 D76.3

158022 **Kod ICD11**

EE81

*Źródło

orphanet