

Postępująca histiocytoza guzowata

Kod Orpha: 158022 Kod OMIM:

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 conjunctiva 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

158022

Kod OMIM

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Kod ICD10

D76.3

Kod ICD11

EE81

[*Źródło](#)

orphanet

Rozszerzony opis choroby

Brak opisu rozszerzonego dla tej choroby. Opracowanie w toku.

Dostępna na stronie www.orphanet.pl