Opis choroby *

Definicja

A rare lichen myxedematosus characterized by a progressive, generalized, papular, sclerodermoid cutaneous eruption usually occurring in association with monoclonal gammopathy, but in the absence of thyroid disease. Histological hallmark is the triad of dermal mucin deposition, fibroblast proliferation, and fibrosis. Patients present with relatively sudden onset of numerous closely spaced, waxy, firm papules and plaques predominantly involving the head, neck, trunk, and dorsal aspects of the extremities, on the background of thickened, edematous, erythematous skin with sclerodermoid appearance. Systemic involvement with cardiovascular, gastrointestinal, pulmonary, musculoskeletal, renal, or nervous system complications is common.

Dane

Klasyfikacja
Choroba

Synonimy
Arndt-Gottron disease
Choroba Arndta i Gottrona
Uogólniona wysypka grudkowa liszajowata
Generalized lichenoid papular eruption
Generalized papular and sclerodermoid lichen myxedematosus

Kod ORPHA
167635

Kod OMIM
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Kod ICD10
L98.5

Kod ICD11
EB90.11

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*Źródło

orphanet