Progressive nodular histiocytosis

INSERM

Source

INSERM. (1999). Orphanet: an online rare disease and orphan drug data base. Progressive nodular histiocytosis. ORPHA:158022

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.