GENETICS AND GENODERMATOSES

# HYALINOSIS CUTIS ET MUCOSAE. A FOLLOW-UP OF 20 YEARS

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Background: Hyalinosis cutis et mucosae (HC) is a rare hereditary disorder characterized by hyaline deposits in the dermis and other internal organs. We report a case of HC followed-up for 20 years.

Observation: A 15-year-old woman, born of non-consanguineous parents, has been presenting a variety of cutaneous and mucosal lesions: verrucous plaques on the elbows, knees and wrist and shiny-white papules on the neck and axillae and along the eyelids and nostrils. The entire tegument appeared thickened with a waxy texture. The buccal mucosa was also infiltrated. Lesions on the scalp, axillae and pubic region caused alopecia. Laryngoscopy revealed a thickened larynx explaining a permanent hoarseness. Histological examination of cutaneous biopsies showed a PAS positive deposition of a hyaline material of the papillary dermis, surrounding blood vessels and adnexial epithelia, leading to the diagnosis of HC. Twenty years later, our examination showed the same clinical and histopathological aspects.

Key message: In our report, the clinical and histopathological features remained unchanged for 20 years which highlights the slowly-progressing course of the disease.