Acral Persistent Papular Mucinosis and Lichen myxedematosus

A.		Rebora
F.		Rongioletti

Department of Dermatology, University of Genoa, Italy

Dr. Alfredo Rebora, Department of Dermatology, Università di Genova, Viale Benedetto XV, Genoa (Italy)

The paper by Borradori et al. in this issue adds much to our knowledge of acral persistent papular mucinosis (APPM) and contributes to clarify its nosography.

APPM was proposed as a distinct form of cutaneous mucinosis in 1986 [1], and other patients have been reported since in the literature, confirming its easily recognizable clinical and histological features.

APPM mainly affects women and is characterized by multiple, symmetric, ivory or flesh-colored, 2- to 5-mm-wide papules located exclusively on the back of the hands and on the extensor surface of the wrists, extending sometimes to the distal forearms. Histologically, a large, focal, well-circumscribed deposit of mucin is found in the upper reticular dermis, interspersed by thin collagen fibers and sparing a subepidermal grenz-zone. Only occasionally is an increased number of fibroblasts present.

The main problem with APPM consists in its nosographical allocation. Is it a variant of lichen myxedematosus (LM) or is it a distinct entity?

In fact, a discrete form of LM (DPLM) had been described long ago in a handful of cases [2], and APPM cases have been said to belong to it. Some differences, both clinical and histopathological, exist, however. In DPLM, both sexes are equally affected, and the papules are larger,

erythematous, asymmetrically located, sometimes coalescing in plaques and variably distributed over the face, trunk, axillary folds and especially on the knees and elbows. Histologically, the deposits of mucin are diffuse among large collagen bundles and not focal as in APPM. Finally, there are a remarkable fibroblastic proliferation and an irregular arrangement of collagen bundles.

Yet, the distinction between APPM and DPLM is not simply a matter of semantics but has much to do with prognosis. Up to date, all patients with APPM had lacked the monoclonal paraprotein which is almost constantly found in LM and is considered essential for its diagnosis. Now, though occasional, the finding by Borradori et al. of paraprotein in a patient with unequivocal APPM suggests that APPM may be related to the group of LM, it being one of its possible presentations.

What is intriguing, however, is that only 4 cases of DPLM had been reported in the about 40 years that preceded the original description of APPM, whereas about 10 cases of APPM have ensued in less than 6. Such a frequency, however relative, depends on peculiar clinical and histopathological presentations of APPM that facilitate its identification and justify the suggestion of Naeyaert et al. [3] to retain the term APPM in any case.

References

1 Rongioletti F, Rebora A, Crovato F: Acral persistent papular mucinosis: A new entity? Arch Dermatol 1986;122:1237-1239.

Montgomery H, Underwood LJ: Lichen myxedematosus (differentiation from cutaneous myxedema or mucoid states). J Invest Dermatol 1953;20:213-236.

Naeyaert JM, Geerts ML, Kudsi S, Kint A: Acral persistent papular mucinosis: A peculiar variant of the discrete papular' form of lichen myxedematosus. Arch Dermatol 1990;126: 1372-1374.

© 1992 Karger AG, Basel 1018-8665/92/1852-0081 \$ 2.75/0