A Case of Acral Persistent Papular Mucinosis

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Acral persistent papular mucinosis is a subtype of the localized papular mucinosis. Clinically, ivory to flesh-colored papules develop exclusively on the back of the hands, the extensor surface of the wrists, and occasionally the distal forearms. Histologically, mucin accumulates in the upper reticular dermis, typically sparing a subepidermal zone and fibroblasts are not increased in number. On experiencing a case of numerous papules on both hands and forearms of a fifty year-old woman, we present it as acral persistent papular mucinosis.
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Key Words : Acral persistent papular mucinosis

Lichen myxedematosus, also called papular mucinosis is a disorder characterized by lichenoid papules, nodules or plaques due to dermal mucin deposition, and a variable degree of fibrosis without thyroid dysfunction. It was classified into four clinical types in 1953 by Montgomery for the first time; generalized lichenoid papular eruption, discrete papular form, localized to generalized lichenoid plaques, and urticarial plaques and nodular eruptions[1]. However, this classification was based on small number of cases and it did not explain the clinical differences such as lesional distribution, histology, associated systemic symptoms and prognosis.

Recently, taking these clinical manifestations into consideration, a new classification was introduced[2]. Lichen myxedematosus can be classified into two major types; generalized papular and sclerodermoid form and localized papular form. The latter is subdivided into five subtypes and acral persistent papular mucinosis is one of them.

We experienced a case of acral persistent papular mucinosis and report it with review of literature.

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CASE REPORT

A 50-year-old woman had pruritic numerous papular eruption on the dorsa of the hands, and the extensor surface of the forearms for 3 years (Fig.1). There was no significant previous medical history or associated systemic complaint. Physical examination revealed multiple, 3 to 5 mm sized, white-ivory colored papules on the dorsum of hand and extensor surface of distal forearm. The papules had a smooth surface and firm consistency (Fig.2). There was no associated sclerosis or induration of the skin adjacent to the papules. The thyroid gland was normal in both size and consistency and the remainder of the physical examination was not significant. Results of laboratory tests for complete blood count, liver function test, urinalysis, thyroid function test and protein electrophoresis were either normal or negative.

Routine hematoxylin-eosin stain revealed a domed-shaped and relatively well circumsciribed lesion situated in the upper reticular dermis sparing subepidermal zone in which most of the collagen fibers were split and replaced by a lightly stained material and few fibroblasts (Fig. 3). This material stained positive for alcian blue at a pH of 2.5 (Fig. 4). The overlying epidermis was normal and there was no evidence of either granulomas or vasculitis.

She received lesional triamcinolone injections

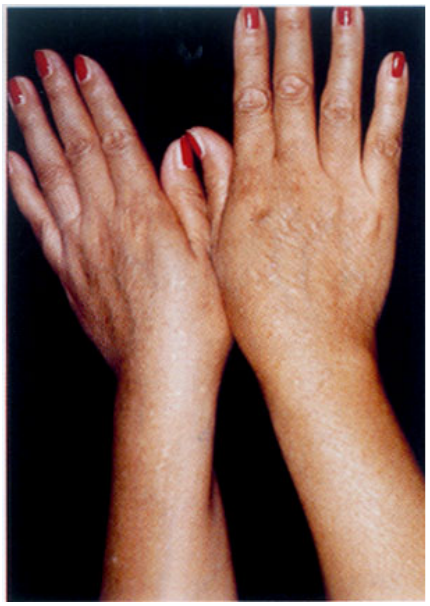

Fig. 1. Numerous papules on the dorsa of the hands and the extensor surface of the distal forearms.

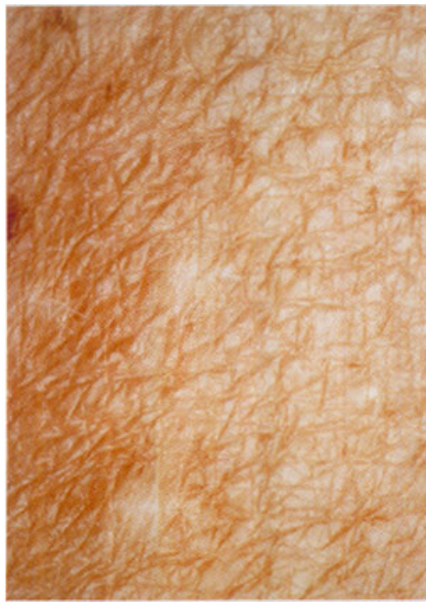

Fig. 2. Multiple 3 to 5 mm sized, firm, white-ivory colored papules on the dorsum of hand.

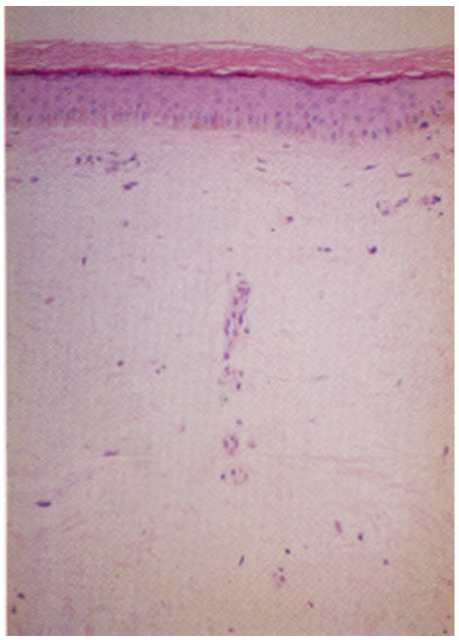

Fig. 3. Focal mucin deposition between the collagen bundles of the upper reticular dermis (H&E stain, × 100).

Fig. 4. Material stained positive for alcian blue at a pH of 2.5 (× 100).

in a few skin lesions and is being observed without gross interval change after two months' of outpatient visits.

DISCUSSION

Lichen myxedematosus or papular mucinosis can be classified as follows: a generalized papular and sclerodermoid form, also called scleromyxedema,

and a localized papular form[2]. The localized form can be further divided into 5 subtypes: 1) discrete papular form in which papules involve any site; 2) acral persistent papular mucinosis in which papules affect only the extensor surfaces of the hands and wrist; 3) self-healing papular mucinosis, in which lesions spontaneously resolve after a few weeks or several months; 4) papular mucinosis of infancy, which is the pediatric variant of the discrete form or acral persistent papular mucinosis; and 5) nodular form[2].

Acral persistent papular mucinosis(APPM) was established by Rongioletti in 1986 and characterized by its exclusive location on the dorsum of hand and wrist and occasionally on the forearm extensor surface without systemic disease[3]. Patients with this diagnosis are typically women and have bilaterally symmetrical flesh-coloured papules always on the hands and wrists. The trunk and face are spared. A monoclonal gammopathy, sclerosis and visceral involvement are absent. It has a good prognosis and course is of persistence and slow progression. Histologically. mucin accumulates focally in the upper reticular dermis, typically sparing a subepidermal zone. Fibroblast are not increased in number[2,3].

Other papular form of mucinosis are most likely to be confused with APPM. The generalized papular and sclerodermoid form, commonly refered to as scleromyxedema, present with wide spread papules and extensive thickening of affected areas, and in most cases an IgG paraproteinemia with λ light chain is associated. It has a chronic, progressive disabling course and is associated with systemic, even fatal manifestations[4,5]. The discrete papular lichen myxedematosus(DPLM) is the most likely to be confused with APPM. DPLM occurs with equal frequency in men and women, papules may confluent into nodules or plaques and involve limbs and trunk. In addition DPLM is larger than that of APPM and may be compared to lesions of sarcoid, xanthomas, or granuloma annulare[1,5,6].

Excision can be tried if the numbers are a few. Topical corticosteroids may be of some benefit. But APPM does not require therapy and a wait-and-see approach is recommended.

On experiencing a case of numerous papules on both hands and forearms of a fifty year-old woman, we present it as acral persistent papular mucinosis, a subtype of papular mucinosis.

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