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Localized Myxoedematous lichen: A rare subtype of cutaneous Mucinosis clinical and dermoscopic features

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Abstract
Cutaneous mucinoses, also called lichen myxoedematosus, are a group of diseases characterized by mucin deposit on the skin. They may be associated to systemic diseases or be primary ones. The discrete papular form (DPLM) is a very rare entity that is located exclusively in the skin and is not associated to any systemic disease. We present the case of a 67-year-old healthy woman who had multiple discrete papillae of normal skin color with a smooth surface at the neck, the nape and the upper part of the trunk and whose histological study confirmed an (DPLM) , and we also report the dermoscopic aspect of this entity not previously found in the literature

Keywords: Cutaneous mucinoses, Mucins , Papular mucinosis , Discrete papular form (DPLM), self- limited skin disease.

Introduction
Lichen myxedematosus (LM) is an idiopathic cutaneous mucinosis [1];. Discrete papular lichen myxoedematous (DPLM) is an uncommon subtype included in the primary cutaneous mucinoses. Contrary to secondary mucinoses, the deposit of mucin in this subtype is the main feature that determines its clinical appearance.

Case report
A 67-year-old woman with no previous medical history , Admitted to dermatology for a slightly itchy rash that has been evolving for 3 years.The dermatological examination had objectified multiple discrete papillae of 2 to 4 mm, of normal skin color with a smooth surface, confluent by some places on a large ill-defined cupboards sitting at the neck, the nape and the upper part of the trunk (figures 1, 2), without other lesions in the rest of the body, and no associated systemic signs, Dermoscopy has shown the presence of whitish homogeneous areas without structure between normal skin giving a cloudy appearance. (Figure 3 a, b)

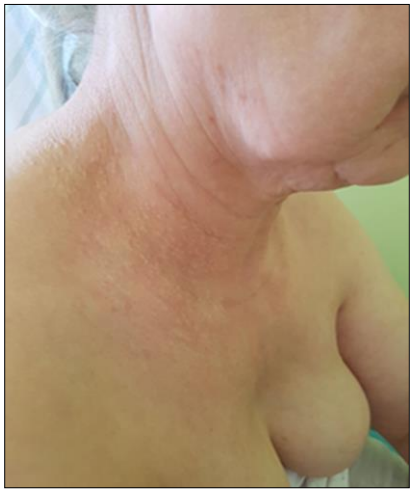

Fig 1

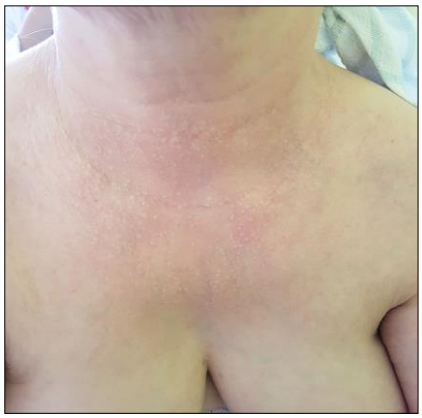

Fig 2

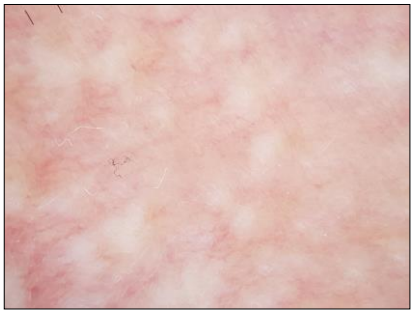

Fig 3 A

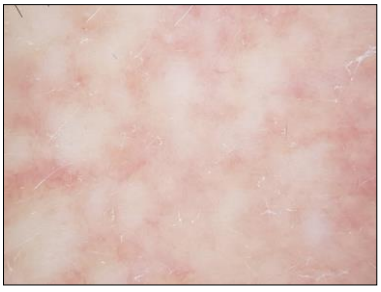

Fig 3B

Histological examination showed a normal epidermis with mucin deposition on the upper papillary and reticular dermis, sparing the deep dermis. There was increased spacing between the collagen bundles, but the number of fibroblasts was not increased. Laboratory tests, including blood count, liver enzymes, kidney function, thyroid hormones, serum protein analysis and immunoglobulins; were normal. Serologic tests for human immunodeficiency virus (HIV) and hepatitis B and C viruses (HBV, HCV) were all negative. The patient was treated with a topical corticosteroid for 6 months, with complete improvement without recurrence after 2 years of follow-up.

Discussion

Papular mucinosis, also known as localized lichen myxedema, or localized mucinous, a été classée en 5 sous-types [1] : The discrete papular form (DPLM), as seen in our patient, is a very rare entity affecting both sexes, with a slight male predominance [2] , characterized by the presence of waxy, flesh-colored or reddish, violaceous papules, 2–5 mm in size, affecting the trunk and limbs in a symmetrical pattern. They can vary in number, and commonly appear in proximal sites [3]. The affected skin is not indurated and the face is usually spared [1] The lesions progress slowly without induration or systemic involvement. Rarely, they resolve spontaneously [4] There have been only 14 cases unrelated to HIV infection reported previously in the literature [5]. The aetiology of this disease remains unknown. An over stimulation of fibroblasts has been implicated in patients with human immunodeficiency virus infection, but not in other cases [6] , DPLM may also be associated with the hepatitis C virus (HCV), particularly among Japanese [7, 8].

By definition, there is no associated paraproteinemia [1],

Dermoscopy of the discrete papular form has not yet been mentioned in the literature

In contrast P.Mendes Bastos *et al* reported dermoscopy of scleromyxoedema by describing a roundish and clustered white-ivory homogenous areas similar to rice grains in between normal skin [9].

Regarding the dermoscopy of Acral persistent papular mucinosis, C. Navarrete-Dechent *et al* described a reddish-orange background color with multiple red dotted and globular vessels [10]. Histologically, the upper and mid dermis shows edema and a diffuse or focal mucinous deposit under a normal epidermis. Fibroblast proliferation is variable, but there is neither collagen deposition nor sclerosis [5].

It is a disease that remains difficult to treat, but a transition to scleromyxedema has not yet been reported [3].

Given that this disorder shows a benign course,and usually self-limitedto the skin and have very little or no morbidity, adopting a no-treatment approach is an acceptable option [11].

Many treatments have been tried including dermabrasion, CO2 laser, intralesional corticosteroids or hyaluronidase injections, oral retinoids and psoralen ultraviolet, A, pimecrolimus, with variable results [5], Topical tacrolimus 0.1% has recently been suggested as an efficient alternative given its antimodulator activity [12], concerning our patient she had a good response under topical corticosteroids alone, after 6 months of treatment without recurrence after 2 years of follow-up.

Conclusion

DPLM is a rare variant of localized LM. It is a self- limited skin disease, and prognosis is generally good. It is important for the clinicians to exclude any possible underlying disease such as gammopathies or HIV infection in such cases

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