Nodular Mucinosis Associated With Light-Chain Monoclonal Gammopathy of Uncertain Significance☆

Mucinosis nodular asociada a gammapatía monoclonal de cadenas ligeras de significado incierto

To the Editor:

Nodular mucinosis is a chronic primary idiopathic mucinosis that, according to the 2001 classification proposed by Rongioletti et al.,[1] belongs to the group of the localized forms of lichen myxedematosus, which includes acral persistent papular mucinosis, a mild form of papular mucinosis, self-healing papular mucinosis, papular mucinosis of infancy, and the nodular form. Localized mucinoses are characterized by the appearance of small numbers of waxy papules (or plaques or nodules due to confluence), usually on the lower limbs or trunk. These localized forms must be differentiated from the diffuse form or scleromyxedema, which is characterized by a rash of groups of small, firm waxy papules of around 2 to 3 mm in diameter, arising predominantly on the upper part of the trunk and on the neck, face, forearms, and hands.[2] The papules show a linear distribution and the perilesional skin is shiny. Muscle, joint, nervous system, gastrointestinal, pulmonary, or otohinolaryngologic manifestations may be present, and paraproteinemia is an almost constant feature. Atypical and intermediate forms also exist; these include patients with scleromyxedema but with no systemic signs or paraproteinemia, patients with localized forms associated with paraproteinemia, and combined or not otherwise specified cases.[1]

We present the case of a 72-year-old woman with no past history of interest. She was seen in our department for the appearance 3 months earlier of 2 brownish erythematous plaques with a slightly scaly surface on both elbows (Fig. 1). The lesions had grown both radially and in thickness and were slightly tender. Biopsy revealed mild orthokeratotic hyperkeratosis with papillomatosis and, in the papillary dermis, abundant interstitial mucin deposits that stained positive with alcian blue. The diagnosis was localized mucinosis (Fig. 2).

Laboratory tests including complete blood count, biochemistry, and 24-hour urine analysis were normal. Plasma protein electrophoresis showed a peak of 890 mg/dL in the gamma region that was shown by immunofixation to be an oligoclonal band in a polyclonal background. Serum free light chains were measured and a kappa chain of 30 mg/L was detected, with a kappa-to-lambda ratio of 1.88. The hematology department did not consider it necessary to perform bone marrow study. With these results, we made a diagnosis of light chain monoclonal gammopathy of uncertain significance based on the Myeloma Working Group 2014 criteria.[3] Treatment was started with topical clobetasol propionate under occlusive dressings for 1 month, with almost complete resolution of the lesions. At the time of writing, the patient continues on follow-up in the hematology department.

This was therefore an atypical form of lichen myxedematosus; only 4 such cases have been published to date.[4–7] The pathogenesis of the association is unknown. In 1 of the cases, the skin lesions disappeared 8 years after their onset and the paraproteinemia some years later, with no systemic clinical repercussions.[4] Another case occurred in a 60-year-old may with skin lesions clinically and histologically compatible with papular mucinosis. After 3 months of treatment with oral corticosteroids with no response, the patient presented disorientation and a deterioration in his general state that led to a diagnosis of immunoglobulin M type multiple myeloma. He then received standard therapy with bortezomib and dexamethasone and the skin lesions disappeared after 4 treatment cycles.[6] Finally, the interesting case of a 38-year-old woman with lesions of papular mucinosis that presented a Köebner phenomenon—a feature not previously described in the literature—and a monoclonal gammopathy of uncertain significance.[7]

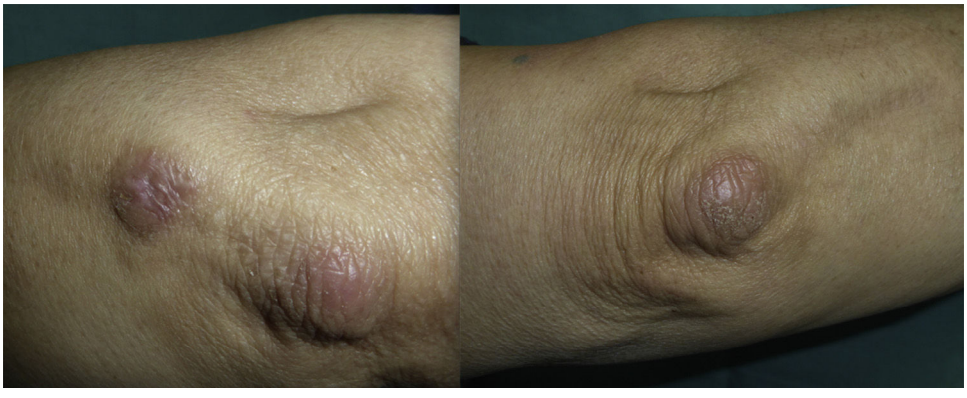

Figure 1 Brownish erythematous plaques on both elbows.

☆ Please cite this article as: Rodríguez-Jiménez P, Chicharro P, Ascensión A, de Argila D, Daudén E. Mucinosis nodular asociada a gammapatía monoclonal de cadenas ligeras de significado incierto. Actas Dermosifiliogr. 2017;108:272–273.

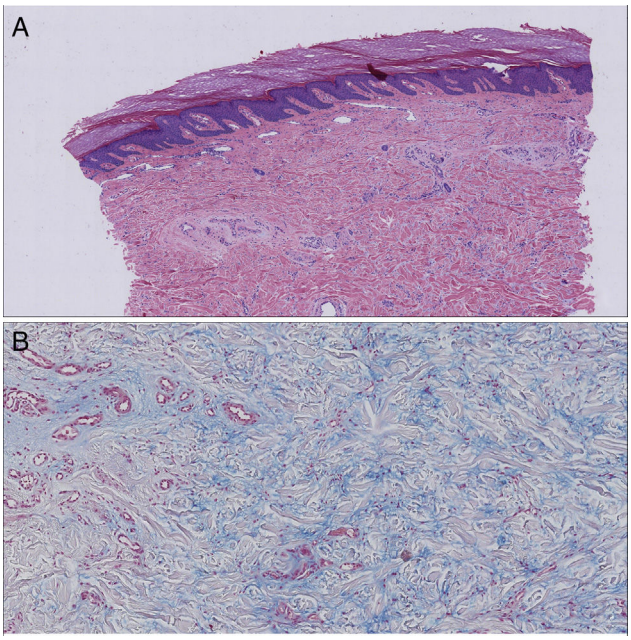

Figure 2 A, Orthokeratotic hyperkeratosis with papillomatosis. Hematoxylin and eosin, original magnification x40. B, Mucin deposits that stain positively with alcian blue. Alcian blue, original magnification x100.

It should be noted that, as previously mentioned by other authors,[1] the terms lichen myxedematosus, scleromyxedema, and papular mucinosis have tended to be used interchangeably in the literature and in daily clinical practice and, despite the 2001 reclassification,[1] confusion between the terms persists, particularly regarding the atypical forms such as ours.

In our patient, thanks to the cutaneous manifestations, we will continue to monitor her closely and, if there is progression to light chain multiple myeloma (estimated incidence of 0.3% per year[3]) or to primary amyloidosis, it should be possible to make an early diagnosis and initiate treatment, which is fundamental to the prognosis of this disease.

Conflicts of Interest

The authors declare that they have no conflicts of interest.

References

1. Rongioletti F, Rebora A. Updated classification of papular mucinosis, lichen myxedematosus, and scleromyxedema. J Am Acad Dermatol. 2001;44:273–81.
2. Dinneen AM, Dicken CH. Scleromyxedema. Am Acad Dermatol. 1995;33:37–43.
3. Rajkumar SV, Dimopoulos MA, Palumbo A, Blade J, Merlini G, Mateos MV, et al. International Myeloma Working Group updated criteria for the diagnosis of multiple myeloma. Lancet Oncol. 2014;15:e538–48.
4. Hardie RA, Hunter JAA, Urbaniak S, Habeshaw J. Spontaneous resolution of lichen myxoedematosus. Br J Dermatol. 1979;100:727–30.
5. Borradori L, Aractingi S, Blanc F, Verola O, Dubertret L. Acral persistent papular mucinosis and IgA monoclonal gammopathy: Report of a case. Dermatology. 1992;185:134–6.
6. Rather PA, Hussain M, Bagdadi F. Localized cutaneous mucinosis associated with multiple myeloma: A rare presentation. Indian J Dermatol. 2014;59:422.
7. Thatte S, Dongre A, Chikhalkar S, Khopkar U. Papular mucinosis associated with monoclonal gammopathy of unknown significance. Indian J Dermatol Venereol Leprol. 2015;81:213.

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Initial Evaluation of the Adult Patient with Condylomata Acuminata☆

Valoración inicial del paciente adulto con condilomas acuminados

To the Editor:

Condylomata acuminata (genital warts) is the sexually transmitted infection (STI) that most often prompts patients to seek medical attention.[1–3] In Spain, the annual incidence of genital warts is estimated to be 160.4 new cases per 100 000 inhabitants.[3]

Genital warts are mainly diagnosed by visual inspection. In the initial clinical assessment of patients who seek care for genital warts, an exhaustive clinical history should be taken and a complete physical examination should be carried out.[1–10]

Various international bodies have published standardized guidelines on the management of genital warts.[4–10] At present, there is no protocol on how to carry out an adequate medical history and physical examination. The following is the routine approach that we follow during a patient's first visit to the STI unit at Centro Sanitario Sandoval in Madrid:

- Demographic data: age, sex.
- Pregnancy and lactation status.

☆ Please cite this article as: Imbernón-Moya A, Ballesteros J. Valoración inicial del paciente adulto con condilomas acuminados. Actas Dermosifiliogr. 2017;108:273–275.