Improvement of plaquelike cutaneous mucinosis after intravenous immunoglobulins treatment

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INTRODUCTION

Plaquelike cutaneous mucinosis is a chronic disease characterized by hyperpigmented well-demarcated papules or plaques over the midline of the back and chest, although bilateral and symmetrical paraxial plaques have also been described.[1] Spontaneous resolution is rare and, given the absence of randomized clinical trials, treatment is based on isolated cases or small case series. Reported interventions include antimalarial medications, antihistamines, topical and systemic corticosteroids, treatment of underlying systemic disorders, or therapeutic abstention.[2] We present a case of plaquelike cutaneous mucinosis with an excellent and sustained response to intravenous immunoglobulins, without recurrences after 3 years of follow-up.

CASE REPORT

A 45-year-old woman consulted for a 2-month history of intensely indurated and painful erythematous and violaceous plaques on the submammary and intermammary regions (Fig 1). She had received treatment with potent topical and systemic corticosteroids, with slight improvement. The patient associated constitutional symptoms consisted of weight loss, anorexia, and asthenia. Her medical history included type 2 diabetes that was adequately controlled with dapagliflozin. Blood analysis and punch biopsy were performed.

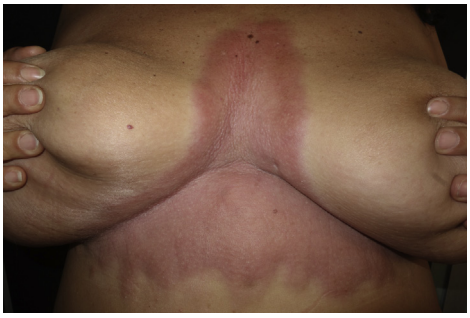

Fig 1. Erythematous and violaceous plaques on the submammary and intermammary regions.

Laboratory examination revealed a globular sedimentation rate of 109 mm/h (normal range 1-20 mm/h), without alterations in her blood cell count or biochemistry. Proteinogram, immunoelectrophoresis, autoimmunity profile, and serology results were normal or negative. Skin biopsy demonstrated a sparse perivascular lymphocytic infiltrate, a discrete thickening and increased number of collagen fibers, and mucin deposits throughout the dermis (Fig 2, *A* and *B*). Cutaneous lesions were clinically compatible with scleromyxedema, but the absence of associated monoclonal gammopathy or fibrosis on histologic examination made this diagnosis unlikely. Because of the location, we considered reticular erythematous mucinosis as a possible diagnosis, although her lesions did

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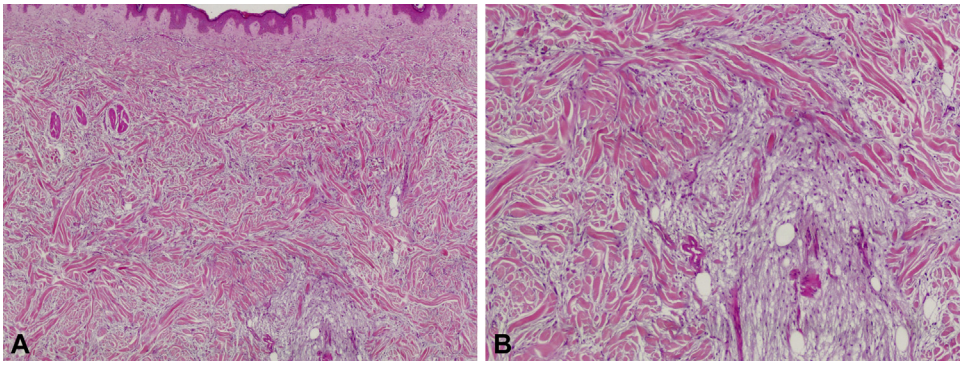

Fig 2. **A** and **B**, Skin biopsy showing dermal mucin deposition, a discrete thickening and increased number of collagen fibers, and a sparse perivascular lymphocytic infiltrate. No dilated blood vessels or fibroblast proliferation was observed. (Hematoxylin-eosin stain; original magnifications: **A**, ×20; **B**, ×40.)

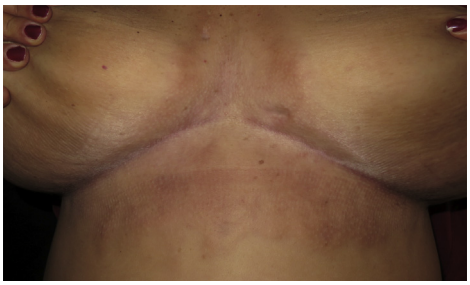

Fig 3. Residual hyperpigmentation after intravenous immunoglobulins treatment.

not have the clinical appearance of a netlike erythema. Consequently, a final diagnosis of plaquelike cutaneous mucinosis was made. Treatment with hydroxychloroquine was started. However, the patient did not tolerate it because of gastrointestinal symptoms. Treatment with intravenous immunoglobulins 2 g/kg (200 g) for 4 consecutive days (50 g/d) once a month for 3 months was started. The patient had a good response, with disappearance of the plaques and induration, but a slight residual hyperpigmentation persisted (Fig 3). As an adverse effect, she complained of self-limited mild headache. The patient has remained asymptomatic for 3 years after treatment completion, and has not developed blood dyscrasias or other systemic abnormalities.

DISCUSSION

Cutaneous mucinoses are a heterogeneous group of disorders characterized by dermal mucin deposition. They are classified as primary or secondary, in which mucin simply represents an associated histologic finding. Plaquelike cutaneous mucinosis is an infrequent variant of lichen myxedematosus that presents with features that are atypical or intermediate between diffuse (scleromyxedema) and localized lichen myxedematosus.[3] It is more frequent in middle-aged women and is characterized by multiple erythematous or hyperpigmented papules coalescing into well-demarcated plaques on the back, chest, or both. Histologically, it presents with a mild to moderate perivascular and perifollicular lymphocytic infiltrate and interstitial dermal mucin.[2]

Differential diagnosis is wide and primarily includes other cutaneous mucinosis, such as reticular erythematous mucinosis or scleromyxedema. The latter is characterized by skin induration and numerous firm papules, fibrosis, and fibroblast proliferation. Nearly all patients have an associated monoclonal gammopathy and sometimes have systemic manifestations that can be fatal.[4] In our patient, the presence of discrete collagen thickening made us consider the diagnosis, although the absence of a monoclonal gammopathy and fibrous papules made this diagnosis unlikely. Conversely, some authors have considered plaquelike cutaneous mucinosis and reticular erythematous mucinosis as a different clinical presentation of the same rare syndrome. Clinically, patients with reticular erythematous mucinosis develop reticular or netlike pink to red macules and vascular dilation is present in the biopsy. There are reports of autoimmune diseases (eg, lupus, dermatomyositis), hypothyroidism, and common internal malignancies in patients with reticular erythematous mucinosis and plaquelike cutaneous mucinosis.[5] Wriston et al[2] reviewed the literature and collected 15 published cases of plaquelike cutaneous mucinosis, of which 2 (13%) were associated with an internal neoplasm and another 2 (13%) with an autoimmune process (hyperthyroidism). The remaining cases were not

associated with an underlying process (as in our patient) or the information was not available.

Plaquelike cutaneous mucinosis treatments are mostly based on case reports. Antimalarial drugs and topical or systemic corticosteroids are the most frequently used.[2] In our patient, systemic corticosteroids did not lead to improvement and antimalarial drugs were not tolerated. Despite the potential for an exacerbation in reticular erythematous mucinosis,[5] improvement after sun exposure has been reported in plaquelike cutaneous mucinosis.[6] Some cases have shown improvement after treatment of underlying diseases (thyroid ablation, thyroid hormone replacement, and radiochemotherapy).[2] To our knowledge, there are no reports of patients with plaquelike cutaneous mucinosis treated with intravenous immunoglobulins. However, European guidelines recommend its use as a first-line treatment for other cutaneous mucinoses such as scleromyxedema, alone or associated with other drugs.[7] Intravenous immunoglobulins can be an alternative treatment in cases of plaquelike cutaneous mucinosis that are recurrent or nonresponsive to more classical therapies.

REFERENCES

1. Owen JL, Word AP, Vandergriff T, Desai SR. Paraneoplastic plaque-like cutaneous mucinosis: a case report. *Dermatol Online J*. 2014;21(2).
2. Wriston CC, Rubin AI, Martin LK, Kossard S, Murrell DF. Plaque-like cutaneous mucinosis: case report and literature review. *Am J Dermatopathol*. 2012;34(4):e50-e54.
3. Cárdenas-Gonzalez RE, Ruelas MEH, Candiani JO. Lichen myxedematosus: a rare group of cutaneous mucinosis. *An Bras Dermatol*. 2019;94(4):462-469.
4. Rongioletti F, Merlo G, Carli C, et al. Histopathologic characteristics of scleromyxedema: a study of a series of 34 cases. *J Am Acad Dermatol*. 2016;74(6):1194-1200.
5. Rongioletti F, Merlo V, Riva S, et al. Reticular erythematous mucinosis: a review of patients' characteristics, associated conditions, therapy and outcome in 25 cases. *Br J Dermatol*. 2013;169(6):1207-1211.
6. Perry HO, Kierland RR, Montgomery H. Plaque-like form of cutaneous mucinosis. *Arch Dermatol*. 1960;82:980-985.
7. Enk AH, Hadaschik EN, Eming R, et al. European guidelines (S1) on the use of high-dose intravenous immunoglobulin in dermatology. *J Eur Acad Dermatol Venereol*. 2016;30(10):1657-1669.