

# Atypical Lichen Myxedematosus Associated with Multiple Myeloma

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**Abstract:** Lichen myxedema (LM) represents a group of rare skin diseases, which falls under chronic mucinosis. LM, previously described as localized scleromyxedema, however, is not associated with systemic involvement. Among the subtypes, atypical LM is rare, with few reported cases associated with multiple myeloma (MM). We present a case of MM, positive for lambda chains, with acute-onset clinical symptoms, and diagnosed with atypical LM. The patient was managed with topical corticosteroid with improvement of lesions after one month of treatment.

**Keywords:** Lichen myxedematous; Mucinosis; Scleromyxedema

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## 1. Introduction

Lichen myxedema (LM) is a chronic mucinosis of idiopathic origin, and its incidence is unknown <sup>[1]</sup>. It can be classified into generalized, localized, and atypical LM, with the latter being very rare. There are several cases reported to be associated with multiple myeloma (MM) and monoclonal gammopathy <sup>[2,3]</sup>. We present a case of atypical LM associated with MM to further understand this pathology, identify the clinical features, and highlight the acute onset that can occur as well as the excellent response to treatment with topical corticosteroid.

## 2. Case

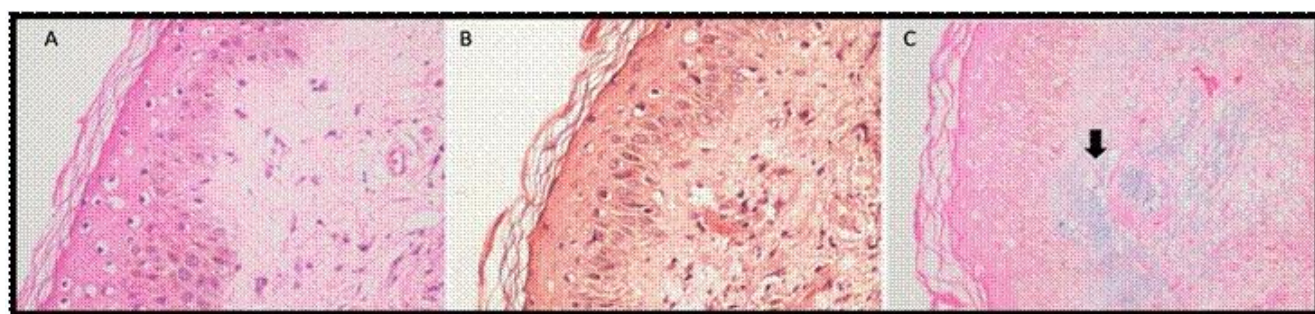
A 65-year-old gentleman who was diagnosed with MM, positive for lambda chains (IgG 340), and received hematopoietic precursor transplantation presented with febrile neutropenia and *Escherichia coli* (*E. coli*) multi-drug resistant bacteremia during hospitalization. He was managed with piperacillin/tazobactam and referred to the Dermatology Unit upon developing mildly pruritic lesions on the pinna, neck, and upper part of the anterior chest over a course of 1 day.

On physical examination, euchromatic, slightly shiny, cupular papules with an erythematous base, which converged giving a cobblestone appearance were observed on the postauricular region, pinna, cheeks, neck, anterior chest, suprascapular region, dorsum of hands, and lateral side of fingers (**Figure 1A–C**).



**Figure 1.** Skin lesion. (A) Back of the neck and interscapular area. (B) Neckline. (C) Left lateral aspect of the neck showing slightly shiny cupuliform papules, with an erythematous base.

LM or lichen amyloidosis was suspected, so a biopsy was performed, which confirmed the diagnosis of LM (**Figure 2**). The patient tested negative for human immunodeficiency virus (HIV), hepatitis B virus (HBV), and hepatitis C virus (HCV); his thyroid-stimulating hormone (TSH) was within normal limits.



**Figure 2.** Histopathology. (A) Hematoxylin and eosin (H&E) staining, x40: lymphocyte exocytosis toward the epidermis, with dermal papillae oedema and slight perivascular inflammatory infiltrate. (B) Congo red staining, x40: negative staining for amyloid deposits in the dermis. (C) Alcian blue staining, x40: positive for mucin deposits in the dermis (thick black arrow).

Subsequently, topical corticosteroid was initiated, with almost-complete resolution of the lesions after one month.

### 3. Discussion

LM is a rare cutaneous mucinosis, predominantly in men, and characterized by slightly translucent euchromatic papules, nodules, or plaques, with a lichenoid appearance due to mucin deposition and varying degrees of fibrosis, in the absence of thyroid disease <sup>[1,2]</sup>. The cause of the deposition remains unclear, but various pathophysiological mechanisms such as paraproteinemia and viral infections (HIV and HCV) have been proposed <sup>[3]</sup>. The most widely accepted hypothesis is associated with the aforementioned entities is an elevation of factors such as interleukin 1 (IL-1), tumor necrosis factor alpha (TNF- $\alpha$ ), and transforming growth factor beta (TGF- $\beta$ ), which stimulate glycosaminoglycan synthesis and fibroblast proliferation <sup>[2]</sup>. The latest revised Rongioletti classification divided LM into 3 subgroups, generalized LM, localized LM, and atypical LM, with the latter sharing characteristics of the first two <sup>[2]</sup>. In the clinicopathological classification, localized LM presents as localized papular rash, with focal and sparse mucin deposits on

histology and variable fibroblast proliferation; there is no association with monoclonal gammopathy or thyroid disease <sup>[4]</sup>. For generalized LM, it presents as generalized papular rash (scleromyxedema) and the classic microscopic triad of mucin deposition, fibroblast proliferation, and fibrosis; it is associated with monoclonal gammopathy in the absence of thyroid disorders <sup>[4]</sup>.

Patients who do not meet the criteria for any of the above subtypes are classified as atypical LM. This entity is very rare, and there are only a few cases reported that are unrelated to HIV <sup>[2-4]</sup>, alike the case of our patient, whose condition was associated with MM and immunoglobulin (Ig) G lambda monoclonal gammopathy in the absence of HIV infection. In the case of our patient, there was focal and scanty mucin deposition in the papillary dermis, as evidenced by Alcian blue staining.

Recently, Nofal *et al.* <sup>[5]</sup> proposed the diagnostic criteria for LM, consisting of two arms. The first arm includes clinical features, such as the presence of firm, waxy papules that coalesce to form nodules or plaques, and typical histopathological features, such as diffuse dermal mucin deposition and fibroblast proliferation. The second arm includes associated findings that have been reported in literature, such as monoclonal gammopathy, thyroid disorder, HIV and HCV infections, peripheral eosinophilia, thymic carcinoma, and hepatocellular carcinoma <sup>[5]</sup>.

A classification system was also proposed for LM severity, which includes three categories: (i) grade 1 (mild) patients with limited (localized) pure skin lesions; (ii) grade 2 (moderate) patients with extensive (generalized) skin lesions; and (iii) grade 3 patients, which include patients with localized or generalized skin lesions and extracutaneous involvement <sup>[5]</sup>.

Although there is no known effective treatment for this pathology, studies have shown that patients respond well to corticosteroids and topical calcineurin inhibitors, although spontaneous remission is not uncommon <sup>[4,6]</sup>. If it is associated with monoclonal gammopathy, immunoglobulin therapy (IVIg) is an option <sup>[6]</sup>; other treatments described include psoralen plus ultraviolet A (PUVA) photochemotherapy, intralesional steroids, carbon-dioxide (CO<sub>2</sub>) laser, and intralesional hyaluronidase <sup>[3]</sup>. In the case of our patient, topical steroid treatment was indicated, with almost-complete resolution of the lesions after one month of treatment.

Atypical LM associated with MM is a rare cutaneous entity, with very few reported cases, of which several aspects are unknown. We present a case in which its acute onset and favorable prognosis are evident; through this case report, more is known about the clinical features and the adequate response to treatment with topical corticosteroid.

## Disclosure statement

The authors declare no conflict of interest.

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