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## Case Presentation

### Mucin in the dermis: a case of tender tumors

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## Abstract

We present an original case report of a 45-year-old woman with a five-month history of sporadic, tender, nodules present on the right upper abdomen, bilateral dorsal wrists, right upper arm, and left flank. Biopsy revealed a mild perivascular infiltrate, increased dermal mucin, and no significant increase in fibroblasts. Presentation and histology were most consistent with nodular lichen myxedematosus (NLM), a rare primary mucinosis. Only four previous cases are reported in the literature to our knowledge. Management of NLM and other subtypes of lichen myxedematosus is not well described. Our patient failed systemic steroids and was unable to tolerate hydroxychloroquine, but subsequently improved with oral methotrexate. This suggests that methotrexate may be of benefit for NLM.

**Keywords:** mucinosis, clinical evaluation and treatment, dermatopathology

## Case synopsis:

### History

A 45-year-old woman presented to the dermatology clinic for a five-month history of sporadic, tender, nodules. The first nodule appeared over the ribs followed by nodules on the arms and trunk. The pain associated with nodule onset subsided over several weeks while the nodules persisted. The patient also complained of pre-existing shoulder, neck, and back pain.

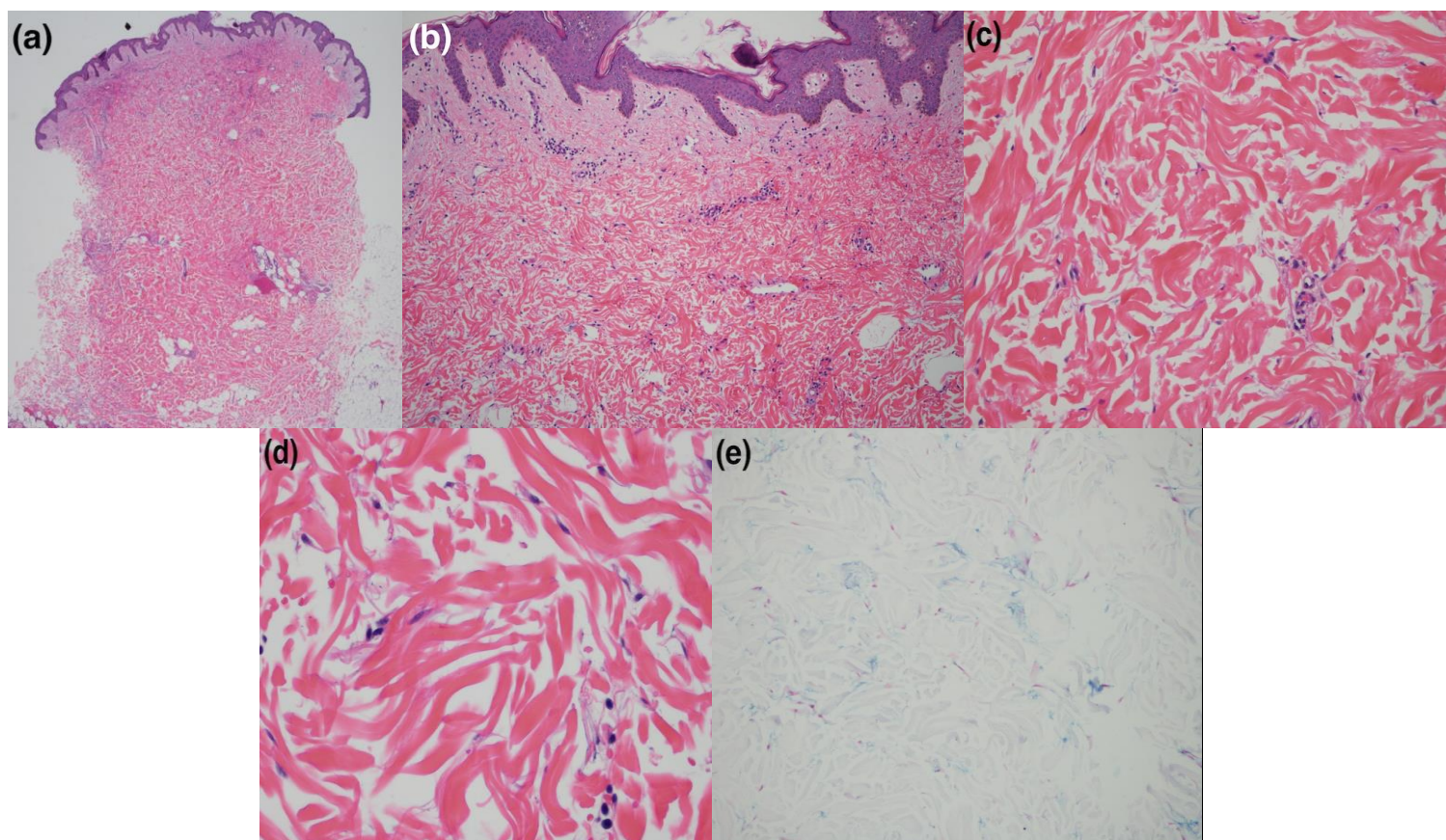
### Physical Examination

At presentation, physical examination revealed multiple tender nodules affecting the right upper abdomen, bilateral dorsal wrists (Figure 1), right upper arm, and left flank.



**Figure 1.** Bilateral dorsal wrists. Symmetric subcutaneous nodules with surrounding soft tissue edema (arrows).

The latter nodule was biopsied (Figure 2).



**Figure 2.** Biopsy, left flank. (a) Cross-section through nodule. Hematoxylin-eosin stain; original magnification: x20. (b) Mild perivascular infiltrate without significant inflammation of the adipose tissue. Hematoxylin-eosin stain; original magnification: x100. (c,d) Increased mucin deposition in dermis with separation of collagen fibers. No significant increase in fibroblasts was evident. Hematoxylin-eosin stain; original magnification: (c) x200, (d) x400. (e) Alcian blue stain highlighting the increased dermal mucin; original magnification: x200.

## Laboratory

Serum thyroid stimulating hormone, C-reactive protein and serum protein electrophoresis were within normal limits. Sedimentation rate was 20mm/hr. Autoimmune serology including ANA, RNP, SSA, SSB, ANCA, ANA, Anti-proteinase 3, and anti-myeloperoxidase antibodies were negative.

## Histopathology

There was mild perivascular inflammatory infiltrate without significant inflammation of the adipose tissue. Increased mucin deposition was noted in the dermis with separation of collagen fibers and no significant increase in fibroblasts. No interface changes were noted. An Alcian blue stain highlighted the increased dermal mucin. In-situ hybridization for kappa and lambda cytoplasmic light chains and an immunostain for CD138 were negative.

## Discussion

### Diagnosis

Nodular lichen myxedematosus (NLM)

### Comment

Nodular lichen myxedematosus is a subtype of localized lichen myxedematosus (LM). LM is a primary mucinosis characterized by papular or nodular eruptions in the absence of thyroid dysfunction [1]. The generalized form of LM, known as scleromyxedema, is associated with a monoclonal gammopathy and other systemic features. In contrast, the localized forms do not share these systemic associations [1]. Four subtypes of localized LM have been described, including (1) a discrete papular form, (2) an acral persistent form, (3) cutaneous mucinosis of infancy, and (4) a nodular form [2]. The nodular form, diagnosed in our patient, is extremely rare with only four previous cases reported to our knowledge [3–6]. However, some cases may have been previously named atypical tuberous myxedema, making our count potentially underestimated [3].

Our patient's presentation with nodules on the dorsal wrists, arms and trunk is most consistent with NLM. The histologic finding of abundant mucin deposition between dermal collagen bundles is also compatible with the diagnosis of LM [1,2]. Absence of serum monoclonal protein further supported this diagnosis [1,2].

Other pathologic considerations include scleredema and reticular erythematous mucinosis (REM). Scleredema is unlikely given the absence of a thick dermis with swollen collagen fibers and clear spaces within the mucin deposition [7]. Although some features of REM appear in the case presented, REM is associated with perifollicular infiltrates and presents as erythematous macules or papules in a reticular pattern on the chest and back, which were not present [8,9]. Jessner's lymphocytic infiltrate and lupus erythematosus tumidus are other considerations, but the lymphoid infiltrate is denser, compared to the slight perivascular pattern seen in the current case [10].

Management of NLM is not well described. One report suggested topical corticosteroids may provide benefit [2], yet a pediatric report showed lack of response to both mometasone and tacrolimus [6]. Another report described a patient with NLM who was successfully treated with intralesional triamcinolone [5].

Given that our patient presented with concurrent arthralgias, systemic therapy was initiated by her rheumatologist. She failed dexamethasone and was unable to tolerate hydroxychloroquine owing to side effects. She then started methotrexate 5 mg weekly. Despite improvement, symptoms flared by the end of each week. The dose was increased to 10 mg, and after two months, the patient noted some improvement in the frequency and severity of her flares. During the next two months the patient continued to have positive response to the methotrexate the first four to five days after each dose. However, she reported progressive swelling and tenderness two to three days prior to the next scheduled methotrexate dose suggesting that the medication, although effective, was not sufficient to provide complete relief. Thus, the dose was titrated to 17.5 mg weekly, the current dose at the time of this publication. Since initiating methotrexate, the patient's symptoms have steadily improved and no new nodules have formed. Although the most effective therapeutic dose for our patient is yet to be established, this clinical picture suggests that methotrexate may be of benefit for NLM.

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