Discrete Papular Form of Lichen Myxedematosus: A Case Report and Review of the Literature

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GOAL
To gain a thorough understanding of lichen myxedematosus (LM)

OBJECTIVES
Upon completion of this activity, dermatologists and general practitioners should be able to:
1. Discuss the clinical presentation of LM.
2. Explain the histologic features of LM.
3. Describe the various forms of LM.

CME Test on page 92.

The discrete papular form of lichen myxedematosus (LM) is a rare idiopathic skin disorder. We present a case of this type in an 80-year-old African American woman. She was treated with pimecrolimus cream, which resulted in symptomatic relief. To our knowledge, this is the first report of the discrete papular form of LM occurring in an African American, as well as the first report on the disorder’s response to pimecrolimus therapy. We also review the English medical literature on this rare disease and examine and summarize the findings.

Citer. 2005;75:105-112.

The discrete papular form of lichen myxedematosus (LM) is a rare idiopathic skin disorder. We present a case of this type in an 80-year-old African American woman. She was treated with pimecrolimus cream, which resulted in symptomatic relief. To our knowledge, this is the first report of the discrete papular form of LM occurring in an African American, as well as the first report on the disorder’s response to pimecrolimus therapy. We also review the English medical literature on this rare disease and examine and summarize the findings.

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Underwood in 1953. LMs are a group of localized or generalized papular eruptions of unknown etiology categorized as cutaneous mucinoses in which dermal mucin deposition is the distinctive histologic feature.\textsuperscript{3-5} LM has no relation to endocrine gland disorders, particularly thyroid disease.\textsuperscript{2,3,6}

In their landmark paper, Montgomery and Underwood\textsuperscript{2} classified 4 clinical types of LM: (1) generalized lichenoid papular eruption, which is also called scleromyxedema, (2) discrete papular form, (3) localized to generalized lichenoid plaques, and (4) urticarial plaques and nodular eruptions that usually evolve into the generalized lichenoid form. In 2001, Rongioletti and Rebora\textsuperscript{6} updated the classification system. The articles by Montgomery and Underwood\textsuperscript{2} and Rongioletti and Rebora\textsuperscript{6} both agree with the existence of the discrete papular form, also called discrete papular LM (DPLM). According to Rongioletti and Rebora, DPLM has only been reported in the medical literature 8 times.\textsuperscript{1,2,7-12} This is unlike scleromyxedema, the most common type of LM, which has been documented in more than 110 cases.\textsuperscript{6} In this case report, we describe an interesting case of DPLM

\textbf{Figure 1.} Right (A) and left (B) neck with multiple 2- to 3-mm discrete, firm, flesh-colored papules.
that was localized to the neck of our patient, and we review the English medical literature.

**Case Report**

An 80-year-old black woman presented to our clinic with a 2-month history of a pruritic eruption localized to her neck. Multiple topical steroids and antihistamines had been used without any decrease in the eruption or the pruritus. The woman's medical history was significant for gastric cancer status post a partial gastrectomy 7 years prior, pseudotumor cerebri, hypertension, and osteoporosis. She is followed regularly for these conditions, which are stable. Her medications included acetazolamide, atenolol, nifedipine, rabeprazole sodium, fluoxetine, oxybutynin chloride, alendronate sodium, docusate calcium, and a multivitamin. Prior to the onset of this eruption, the patient had a change in the acetazolamide formulation, which was initially thought to be the culprit. Although she returned to the original formulation for longer than a month, she had no resolution of the symptoms. At that time, she sought evaluation in our clinic.

On physical examination, multiple 2- to 4-mm discrete, flesh-colored shiny papules without scale were present over the patient's bilateral and posterior neck (Figure 1). Results of 2 punch biopsies showed splaying of the collagen with mucin present diffusely throughout the dermis. No increase in the number of fibroblasts was seen (Figure 2). Results of colloidal iron stain showed positive results in the papillary dermis and showed mucin mildly throughout the deep dermis (Figure 3), supporting the diagnosis of LM. Complete blood count, chemistry panel, liver function, thyrotropin, and serum protein test results were all within reference range. Pimecrolimus cream has been effective in treating our patient's pruritus. We have followed her for 14 months, and the lesions have remained localized to her neck, with continued normal thyrotropin and serum protein values.

**Comment**

In 2001, Rongioletti and Rebora\(^6\) revised the classification system of LM, categorizing it into 3 broad subsets: (1) generalized papular and sclerodermoid, (2) localized LM, and (3) atypical forms.

The first subset, generalized papular and sclerodermaid, represents scleromyxedema. Diagnosis requires a generalized papular and scleroderma eruption, monoclonal gammopathy (paraproteinemia), no evidence of thyroid dysfunction, and a histologic triad of fibroblast proliferation, fibrosis, and mucin deposition.\(^6\) Scleromyxedema is associated with many systemic disorders that may include numerous organ systems.\(^6,13\) Although spontaneous resolution has been reported,\(^14\) scleromyxedema typically is a long-term and disfiguring disease\(^6,13\) associated with variable morbidity and mortality.\(^15,16\)

The criteria for diagnosing the subset of localized LM requires a papular eruption, deposits of mucin with variable fibroblast growth, absence of paraproteinemia, and absence of thyroid dysfunction.\(^6\) There are 5 subtypes\(^6\): DPLM,\(^2,7,9,17-21\) acral persistent papular mucinosis (APPM),\(^22-27\) cutaneous mucinosis of infancy,\(^28-31\) self-healing papular mucinosis (SHPM),\(^32-36\) and nodular LM.\(^37,38\) All subtypes show small, firm, waxy papules limited to a few areas of the skin, which may coalesce and form nodules or plaques. DPLM can involve any site on the body. APPM exclusively involves both extensor surfaces of the distal upper extremities. Cutaneous mucinosis of infancy is a pediatric variant of DPLM or APPM. SHPM has spontaneous resolution. Nodular LM is characterized by a

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**Figure 2.** Results of a 4-mm punch biopsy show spaces between the collagen fibers and no increase in the number of fibroblasts (H&E, original magnification ×4).
predominance of nodules. All subtypes except for SHPM typically persist long-term. Systemic symptoms were only reported in SHPM. In general, localized LM is self-limited and associated with a good prognosis.

The histologic features of LM are summarized in Table 1. The pathology lies within the dermis for all types of this disease, and the epidermis is essentially normal. The types of LM can be distinguished from one another by 3 histologic clues: (1) mucin distribution pattern, (2) dermal level of mucin deposits, and (3) some extra findings. Differentiation can be additionally aided by observing the number of fibroblasts. Common mucin stains include colloidal iron, mucicarmine, and alcian blue at pH 2.5 (but not pH 0.5); common metachromatic mucin stains include toluidine blue, thionine, and methylene blue.

DPLM is a subtype of localized LM. Although the lesions can involve any site, DPLM is typically distributed symmetrically to the trunk and limbs. The skin lesions have been described as a variable number of firm, smooth, waxy, or flesh-colored papules 2 to 5 mm in size. Normal serum protein and thyroid function test results verify the diagnosis of DPLM. Because Montgomery and Underwood originally defined LM as having no relation to any endocrine gland disturbance, we recommend additional random chemistry panel or fasting blood glucose laboratory tests if there is any question that a patient may have diabetes mellitus.

Histologically, DPLM may have a diffuse or local pattern of mucin distribution involving the upper and mid reticular dermis. The involved dermis typically shows edema. The amount of fibroblast proliferation is variable. When compared with scleromyxedema, DPLM has no collagen deposition or sclerosis and a lesser amount of fibroblast proliferation.

Some argue that DPLM and APPM are closely related variants. However, we believe that DPLM and APPM are distinct subtypes of localized LM. DPLM typically affects men (Table 2) and its lesions may be erythematous, larger, and include areas other than the distal upper extremities. In comparison, APPM has an overwhelming female-to-male ratio of 4.7:1. APPM also exclusively involves the back of the hands, extensor surface of the wrists, and sometimes the distal forearms. Histologically, APPM typically has more focal mucin deposits, which spare a subepidermal grenz zone, and a normal number of fibroblasts; DPLM has mucin deposits that are more diffuse (compared with APPM), have a variable number of increased fibroblasts, and have an irregular arrangement of collagen bundles.

Table 2 summarizes the main features of the 10 DPLM cases reported in the English medical literature. Our criteria required there be no evidence of paraproteinemia, no history or laboratory evidence of any endocrine disease, no history of human immunodeficiency virus infection, histologic proof of dermal mucin deposits, and specific gross descriptions consistent with DPLM. It should be noted that only 4 of the 8 DPLM cases originally cited by Rongioletti and Rebora were included in Table 2. Four cases were excluded because 2 reports were written in French, 1 report included a patient with diabetes mellitus, and another did not perform a necessary laboratory test to rule out thyroid disease. Other cases were excluded because they did not fulfill our criteria.

Figure 3. Biopsy specimen stained with colloidal iron exhibits ample mucin between the collagen fibers (original magnification ×10).
Among the 10 DPLM patients summarized, 7 were men. The mean age of the group was 51.3 years. Only 3 of the 10 cases reported information on the patient’s ethnicity: one was Caucasian, one was Asian, and our patient was African American. Nine of the 10 cases reported no evidence of paraproteinemia. The single case that did not report this finding was published before an association between LM and paraproteinemia was known in the 1960s. All 10 cases reported thyroid function test results within reference range. Only 3 of the 10 patients had symptomatic skin lesions, which were mainly pruritus. No systemic symptoms were noted. Three of the 10 patients had solitary skin lesions on areas other than the typical trunk and limbs. One patient had skin lesions only on his lumbar region, another had facial lesions, and our patient had lesions on her neck. Seven of the 10 patients had comorbid medical disorders, including hypertension, migraines, psoriatic erythroderma, psoriasis, seizures, gastric cancer in remission, pseudotumor cerebri, osteoporosis, and hepatitis C. Only hepatitis C was comorbid with more than one DPLM patient. This may be a coincidence because one patient developed DPLM after contracting hepatitis C, while the other patient developed it before contracting hepatitis C. Psychiatric illness was limited to depression and was found in 2 cases. No DPLM cases progressed to scleromyxedema, and none have been reported in the literature.

DPLM rarely resolves on its own. Spontaneous resolution did not occur in any of the 10 reported DPLM cases, and only 2 patients were treated successfully. In Reynolds et al, the patient responded to therapy with intralesional corticosteroid injections and flurandrenolide-impregnated tape. In Kaymen et al, a patient was treated with a CO2 laser and postoperative intralesional corticosteroid injections. There was no growth after one year. All the other DPLM cases did not have effective therapy, lacked specific details of improvement, or did not report this.
<table>
<thead>
<tr>
<th>Reference</th>
<th>Age, y (Sex)</th>
<th>Symptoms</th>
<th>Location of Lesions</th>
<th>Course After Initial Medical Visit</th>
<th>Treatments</th>
<th>Other Notable Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Montgomery and Underwood²</td>
<td>38 (F)</td>
<td>NR</td>
<td>Dorsal surface of forearms, wrists, hands, and medial aspects of knees</td>
<td>Mild enlargement in size of skin lesions after 10 y</td>
<td>NR</td>
<td>None</td>
</tr>
<tr>
<td>Coskey and Mehregan⁷</td>
<td>22 (M)</td>
<td>AS</td>
<td>Left deltoid, right arm</td>
<td>No change after 8 y</td>
<td>NR</td>
<td>None</td>
</tr>
<tr>
<td>Tay and Khoo⁸</td>
<td>41 (M)</td>
<td>AS</td>
<td>Trunk, shoulders, and extensor surfaces of arms and forearms</td>
<td>No change after 2 y</td>
<td>Failed therapy with oral thyroxine</td>
<td>None</td>
</tr>
<tr>
<td>Enerback and Mobacken⁹</td>
<td>46 (M)</td>
<td>Symptomatic</td>
<td>Chest, back, shoulders, neck, and upper arms</td>
<td>Stable with mild changes after 9 y</td>
<td>Failed therapy with topical corticosteroids</td>
<td>None</td>
</tr>
<tr>
<td>Rongioletti and Rebora¹⁷</td>
<td>59 (F)</td>
<td>AS</td>
<td>Both upper extremities (upper arms and forearms)</td>
<td>Spreading of new lesions to shoulders, thighs, and trunk after 1 mo of interferon therapy for hepatitis C</td>
<td>None</td>
<td>DPLM worsened with interferon therapy; onset of DPLM occurred before acquiring hepatitis C</td>
</tr>
<tr>
<td>Reynolds et al¹⁸</td>
<td>32 (M)</td>
<td>NR</td>
<td>Chest, back, upper extremities more than lower extremities</td>
<td>Stable after therapy; exact period NR</td>
<td>Intralesional and topical corticosteroids</td>
<td>Successful therapy</td>
</tr>
<tr>
<td>Kaymen et al¹⁹</td>
<td>63 (M)</td>
<td>NR</td>
<td>Face only</td>
<td>Stable 1 y after treatment with CO₂ laser</td>
<td>CO₂ laser and intralesional corticosteroids</td>
<td>Successful therapy</td>
</tr>
<tr>
<td>Montesu et al²⁰</td>
<td>70 (M)</td>
<td>Symptomatic</td>
<td>Face, neck, and clavicle area (stable for 2 y)</td>
<td>New lesions develop on back of hands and buttocks; period NR</td>
<td>Some improvement with topical corticosteroids and emollients</td>
<td>Onset of DPLM occurred after acquiring hepatitis C</td>
</tr>
<tr>
<td>Poswig et al²¹</td>
<td>62 (M)</td>
<td>AS</td>
<td>Back (lumbar region)</td>
<td>No change after 18 mo</td>
<td>NR</td>
<td>None</td>
</tr>
<tr>
<td>Our patient</td>
<td>80 (F)</td>
<td>Symptomatic</td>
<td>Posterior and lateral surfaces of neck</td>
<td>No change after 14 mo</td>
<td>Failed therapy with topical corticosteroids and antihistamines; successful symptomatic relief with pimecrolimus</td>
<td>No gross changes in skin lesions, but symptomatic relief from pruritis with topical pimecrolimus therapy</td>
</tr>
</tbody>
</table>

*F indicates female; NR, not reported; M, male; AS, asymptomatic; DPLM, discrete papular lichen myxedematosus.
information.2,7-9,17,20,21 Only Tay and Khoo,8 Enerback and Mobacken,9 and Kaymen et al19 commented on which therapies failed (oral thyroxine, topical corticosteroids, and shave excision, respectively). Our patient complained of persistent itching that did not respond to initial treatment with topical steroids and antihistamine medications. She later received pruritic relief with pimecrolimus cream therapy. However, the skin lesions remained. To our knowledge, this is the first report of pimecrolimus therapy in the treatment of pruritis secondary to DPLM.

It is difficult to treat a rare disease such as DPLM when the pathogenesis is unknown, and many treatments have failed. Fortunately, DPLM and the other localized forms of LM are usually self-limited to the skin and have very little or no morbidity, leading some experts to believe that the disorder is unnecessary to treat.8 We believe that treatment is sometimes helpful. In our patient, the DPLM lesions were pruritic and located at a cosmetic area of the neck. The patient and dermatology staff decided to pursue a treatment plan. Therefore, even though localized LM lesions are typically benign, sometimes it is beneficial to treat them, especially if they cause irritating symptoms or cosmetic issues.

Conclusion
DPLM is a rare variant of localized LM. DPLM can be diagnosed by a thorough history and physical examination; histologic proof of dermal mucin deposits; and ruling out other diseases with laboratory tests for serum protein, thyroid function, and, if the patient is at risk of diabetes mellitus, blood glucose levels. DPLM is a self-limited skin disease, and prognosis is generally good, but it typically persists long-term and may slowly progress. Treatment is usually unnecessary, but it may be recommended if the lesions are symptomatic or cause cosmetic issues. Unfortunately, few treatment plans have been shown to successfully treat DPLM.

REFERENCES
Lichen Myxedematosus


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