Acquired ichthyosis (AI) in association with systemic lupus erythematosus (SLE) is a rare dermatologic finding, with only 7 previously published cases worldwide. We report a 25-year-old black woman with AI associated with SLE. A skin biopsy specimen from the lower extremity showed histologic changes consistent with both ichthyosis vulgaris and SLE, a unique finding that has not been previously reported. We also review the world literature on AI and SLE.

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Case Report
Systemic lupus erythematosus (SLE) was diagnosed in a 25-year-old black woman 2 years prior to hospital admission. She was hospitalized for acute steroid psychosis versus a primary psychotic disorder and was treated with olanzapine and divalproex sodium. Her prior medical history included iron deficiency anemia, osteoporosis, and osteomyelitis. The patient reported a new skin eruption in the lower extremity that coincided with her most recent SLE exacerbation.

Results of a physical examination revealed malar rash, oral ulcers, and diffuse alopecia. The lower extremities demonstrated bilateral ichthyosiform scaling (Figure 1). Pertinent laboratory findings included an erythrocyte sedimentation rate of 99 mm/h (reference range, 0–20 mm/h), anemia, and lymphopenia. Laboratory tests were positive for antinuclear antibodies (titer 1:2560) with a speckled pattern and anti-Sm (Smith) antibodies. The serum C3 level was within reference range and the C4 level was <10 mg/dL (reference range, 20–50 mg/dL). Results of a laboratory test for thyroid-stimulating hormone were negative. A skin biopsy specimen from the left shin demonstrated a hyperkeratotic stratum corneum with a diminished stratum granulosum (Figure 2). Colloidal iron stain revealed extensive dermal mucin deposition (Figure 3). Direct immunofluorescence was not performed.

The patient was treated for an SLE exacerbation due to malar rash, alopecia, arthritis, anemia, psychosis, hypocomplementemia, and elevated antinuclear antibody titers. She was managed with high-dose prednisone for 3 weeks and hydroxychloroquine sulfate 200 mg twice daily. The patient was treated for acquired ichthyosis (AI) with emollients and lactic acid lotion 12% and showed improvement at follow-up examination.
# Review of Published Case Reports of Patients With Systemic Lupus Erythematosus (SLE) and Acquired Ichthyosis (AI)

<table>
<thead>
<tr>
<th>Case Report</th>
<th>Author (Year)</th>
<th>Age, y; Race; Sex</th>
<th>Description of AI</th>
<th>Location of AI</th>
<th>Histologic Features</th>
<th>Treatment</th>
<th>Disposition of AI</th>
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<tbody>
<tr>
<td>1</td>
<td>Duvic and Jegasothy(^4) (1980)</td>
<td>26; female</td>
<td>Hyperpigmented, diamond-shaped, scaly areas with fine diffuse scaling</td>
<td>Anterior shins, arms</td>
<td>Hyperkeratosis without parakeratosis, decreased granular cell layer thickness, normal dermis</td>
<td>Increase in prednisone dose</td>
<td>Resolved rapidly but recurred 5 mo later with flare-up of SLE while on tapered prednisone dose</td>
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<tr>
<td>2</td>
<td>Font et al(^3) (1990)</td>
<td>31; female</td>
<td>Hyperpigmented, well-demarcated, scaly areas with fine diffuse scaling</td>
<td>Trunk, arms, thighs</td>
<td>Marked epidermal atrophy, hyperkeratosis with mild parakeratosis, absence of a granular layer</td>
<td>Addition of prednisone</td>
<td>Resolved after 1 mo</td>
</tr>
<tr>
<td>3</td>
<td>Yamada et al(^9) (1990)</td>
<td>75; male</td>
<td>Pigmentation, cornification</td>
<td>Entire body</td>
<td>N/A</td>
<td>Prednisolone acetate</td>
<td>Still present at time paper was submitted</td>
</tr>
<tr>
<td>4</td>
<td>Labauge et al(^8) (1992)</td>
<td>51; female</td>
<td>Rapidly progressive diffuse ichthyosis</td>
<td>Trunk, extremities</td>
<td>Hyperkeratotic epidermis with the absence of a granular layer, mild dermal edema</td>
<td>Addition of prednisolone acetate</td>
<td>Resolved within 2 y</td>
</tr>
<tr>
<td>5</td>
<td>Roger et al(^7) (1993)</td>
<td>25; female</td>
<td>Hyperpigmented, well-demarcated, scaly areas with fine diffuse scaling</td>
<td>Trunk, arms, thighs</td>
<td>N/A</td>
<td>Pulse therapy with prednisolone acetate with continuation of cyclophosphamide</td>
<td>Resolved after 2 wk but recurred 1 mo later with another flare-up of SLE</td>
</tr>
<tr>
<td>6</td>
<td>Tlacuilo-Parra et al(^5) (2004)</td>
<td>30; mestizo Mexican; female</td>
<td>Hyperpigmented, well-demarcated, scaly areas with fine diffuse scaling</td>
<td>Back, arms, thighs</td>
<td>Epidermal atrophy, marked hyperkeratosis with decreased granular cell layer thickness, normal dermis</td>
<td>Increase in prednisone dose with continuation of cyclophosphamide</td>
<td>Resolved after 2 mo</td>
</tr>
</tbody>
</table>
Comment

Adult AI may be associated with malignancy, medication, and, rarely, autoimmune disease. Dermatomyositis, mixed connective tissue disease, and SLE have infrequently been reported with AI. Although skin lesions are found in as many as 85% of patients with SLE, AI, which is clinically indistinguishable from ichthyosis vulgaris, is a rare cutaneous finding. AI may be the only presenting cutaneous finding of SLE and may vary in intensity with the waxing and waning of the underlying disease. Treating SLE usually results in improvement of AI.4

A comprehensive world literature search was conducted without language restriction using a PubMed/MEDLINE search of articles referenced in Index Medicus and revealed only 7 other cases of AI associated with SLE (Table). Five cases were published in English,1-7 1 in French,8 and 1 in Japanese.9 Our case is the only one with histologic findings of AI and SLE demonstrated in a single skin biopsy specimen. Similar to ichthyosiform sarcoidosis, with the epidermal changes of ichthyosis vulgaris and the dermal granulomas of sarcoidosis, our case had the same epidermal change and dermal mucin associated with SLE.10

The histologic features of AI are identical to those of ichthyosis vulgaris and include thickening of the stratum corneum and absence or thinning of the granular layer.11 The pathogenesis of AI is unclear. Diminution of the granular layer may be caused by an abnormal host response targeted against keratohyalin granules. Antiprofilaggrin antibodies are thought to connect the autoimmune nature of SLE with the development of the secondary process of AI.5

The primary treatment for AI associated with SLE is treatment of the underlying connective tissue...
disorder. Keratolytics including salicylic acid, urea, and propylene glycol also can be employed. These agents activate keratinocyte disaggregation and induce the removal of scales. Hydration with lactic acid lotion 12% promotes the disaggregation of corneocytes by increasing susceptibility to mechanical forces and increasing the release of hydrolytic enzymes.

REFERENCES