

Widespread Poikilodermatous Dermatomyositis Associated With Chronic Lymphocytic Leukemia

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PRACTICE POINTS

- Poikiloderma, even with an unusual clinical presentation, can be a useful clinical clue for the diagnosis of dermatomyositis or other collagen vascular disease.
- Dermatomyositis can be paraneoplastic and though often associated with epithelial malignancies and solid tumors can also be associated with leukemias.

To the Editor:

Dermatomyositis represents a rare idiopathic inflammatory process presenting with cutaneous lesions and muscular weakness. It often represents a paraneoplastic syndrome. We report the case of a 62-year-old man with a history of total-body poikiloderma and a recent diagnosis of chronic lymphocytic leukemia (CLL). Despite lacking typical features of the disease, a diagnosis of dermatomyositis was made. Our patient may represent a distinct poikilodermatous variant of dermatomyositis, sharing the generalized distribution of the erythrodermic subtype.

A 62-year-old man presented with pruritic poikiloderma involving the neck, arms, legs, abdomen, chest, and back of 2 years' duration (Figure). He also experienced dysphagia and weakness of the legs. The rash was previously treated by other dermatologists with a combination of high-potency topical steroids and topical tacrolimus 0.1% without success. His history was notable for CLL, which had been diagnosed by a dermatologist 6 months prior to the current

presentation. Prior to his visit to the dermatologist, the patient had received 6 chemotherapeutic sessions with a combination of rituximab and cyclophosphamide for the treatment of CLL. The rash did not improve with chemotherapy.

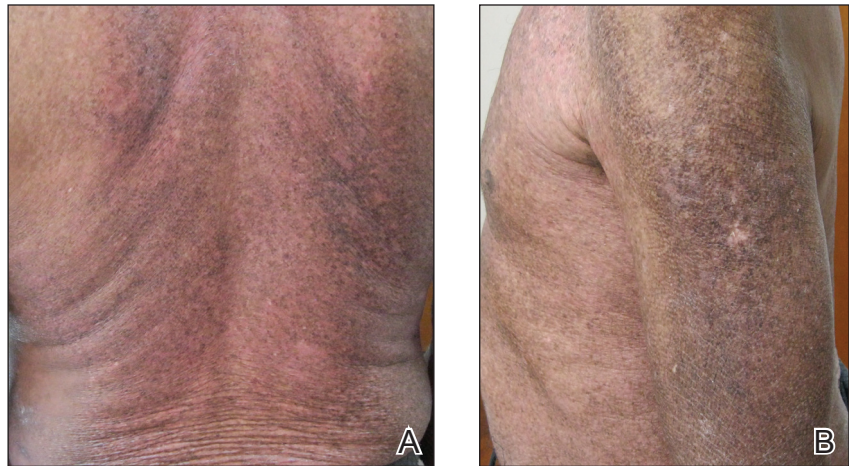
Repeat biopsies of affected regions only demonstrated features of mild interface dermatitis. Direct immunofluorescence studies showed scattered colloid body fluorescence for IgM. Because of bilateral weakness of the legs, a muscle biopsy was taken, which demonstrated severe atrophy and interstitial fibrosis, with neurogenic abnormalities detected in areas of lesser atrophy via abnormal muscle fiber-type grouping. Metabolic panel showed elevated muscle enzymes in the blood: creatine kinase, 243 U/L (reference range, 10–225 U/L); serum aldolase, 16 U/L (reference range, ≤ 8.1 U/L); lactate dehydrogenase, 314 U/L (reference range, 60–200 U/L). An autoimmune panel was negative for Jo-1, Scl-70, U1 ribonucleoprotein, DNA, desmoglein 1 and 3, and antiacetylcholine receptor antibodies. An elevated erythrocyte sedimentation rate was measured at 16 mm/h (reference range, 0–10 mm/h). Given these findings, the lesions were confirmed as a widespread poikilodermatous variant of dermatomyositis.

The patient was placed on a daily 50-mg dose of prednisone, which produced rapid improvement in scaling and erythema. Creatine kinase and serum aldolase levels normalized and motor strength

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The authors report no conflict of interest.

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Poikiloderma distributed on the back (A) as well as the abdomen and arms (B).

increased. After 1 week the prednisone dosage was reduced to a daily 30-mg dose, and then 20 mg a week later. The skin lesions completely resolved within 4 to 5 months and the patient is currently on a prednisone dose of 5 mg, alternating with 2.5 mg of prednisone and rituximab infusion every 2 months.

Dermatomyositis is a rare entity with an incidence of approximately 0.5 to 1 per 100,000 individuals.¹ It presents with a characteristic rash composed of Gottron papules; pathognomonic flat violaceous papules on the dorsal interphalangeal joints, elbows, or knees; and a heliotrope rash, a violaceous erythema involving the eyelids. Poikiloderma frequently is reported to present in a shawl-like distribution, encompassing the shoulders, arms, and upper back.^{1,2} Dermatomyositis of the poikilodermatous type can present in nonphotoexposed areas and photoexposed areas. The unusual feature is the total-body involvement, which is analogous to erythroderma.³

Our case may represent a distinct poikilodermatous manifestation sharing the distribution of the erythrodermic subtype. We believe that the skin lesions may have represented a paraneoplastic event presenting prior to diagnosis with CLL. Dermatomyositis has a strong association with cancer, with patients 3 times more likely to develop internal malignancy.⁴ Association is strongest for non-Hodgkin lymphoma, as well as ovarian, lung, colorectal, pancreatic, and gastric cancer. When associated with malignancy, symptoms of dermatomyositis or myositis typically precede the discovery of malignancy by an average of 1.9 years.⁵ Dermatomyositis has been previously reported to present as a paraneoplastic manifestation of CLL.⁶ One case has been reported of a patient with CLL who developed leukemia cutis presenting with poikiloderma in the characteristic dermatomyositis shawl-like distribution.⁷ The lack of dermal infiltration with leukemic cells in our patient, however, makes a paraneoplastic etiology much more likely.

Our patient's rash did not initially improve with treatment of CLL, but dermatomyositis associated with hematological malignancy may precede, occur simultaneously, or follow the diagnosis of malignancy.⁸ Additionally, symptoms of dermatomyositis do not always parallel the course of hematological malignancy outcome. However, rituximab has been used as a treatment of dermatomyositis and may have contributed some synergistic effect in combination with prednisone in our patient.⁹

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