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Poikilodermatomyositis☆

Poiquilodermatomiositis

Diana Eugenia Perales-Martínez,[a] Benjamín Moncada,[b] Antonio Rolón-Padilla,[c] Mario Alfredo Chávez-López[a,*]

[a] Departamento de Medicina Interna, Centenario Hospital Miguel Hidalgo, Aguascalientes, Mexico
[b] Departamento de Dermatología, Hospital Central Dr. Ignacio Morones Prieto, San Luis Potosí, Mexico
[c] Departamento de Patología, Centenario Hospital Miguel Hidalgo, Aguascalientes, Mexico

We present a rare case of a woman diagnosed with dermatomyositis associated with interstitial lung disease as a manifestation preceded the onset of poikilodermatosis. This presented as a generalized skin rash characterized by confluent lesions, which later was replaced by hyperpigmented and hypopigmented macules (Fig. 1), dryness and scaling, as well as alopecia. Four months later melanonychia appeared, as well as heliotrope erythema, periorbital edema, and Gottron's papules on the proximal and distal interphalangeal joints of both hands (Fig. 2), progressive muscle weakness with a symmetric, proximal distribution and weight loss of 10 kg.

The patient then presented dysphagia and dyspnea progressing to orthopnea, so she was admitted to the hospital. The most interesting laboratory results showed: creatinine 203 U/l, lactic dehydrogenase 698 U/l, aspartate transaminase 304 U/l, erythrocyte sedimentation rate 20 mm/h, rheumatoid factor 57 IU/l, C-reactive protein 3.7 mg/dl, complement fractions C3 55 mg/dl and C4 11.1 mg/dl.

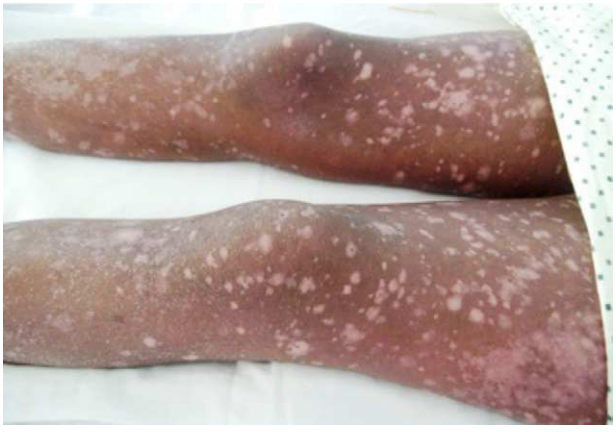

Fig. 1. Image of the legs with the characteristic alternating hypo and hyperpigmented macules of the poikilodermatosis.

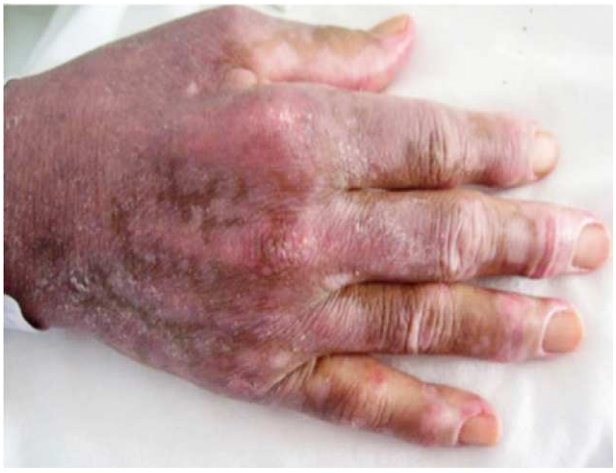

Fig. 2. Image of the hands showing Gottron's papules on the metacarpophalangeal joints superimposed on the poikilodermatosis hypopigmented lesions.

Computed tomography revealed: interstitial lung disease, emphysematous bullae, bronchiectasis and areas of pulmonary fibrosis. During the hospital stay she developed pneumonia associated with mechanical ventilation and septic shock leading to her death.

The poikilodermatosis is a rare sign of dermatomyositis and may be the initial manifestation of the disease,[1–5] as the case above described.

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* Corresponding author.
E-mail address: drmariochavez@yahoo.com (M.A. Chávez-López).

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