cutis® photo quiz

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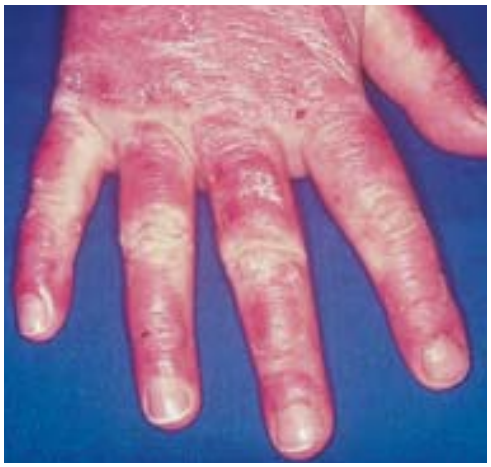
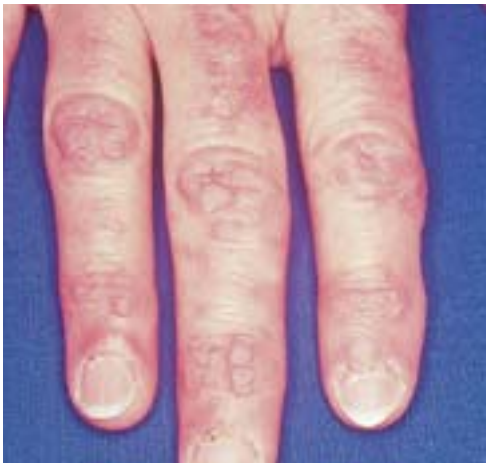


Patient 1



Patient 2

Patient 1: A 56-year-old white female presents with scaling, erythematous patches across her chest and arms. Her hands have erythematous, scaling lesions with clearing over the metacarpophalangeal and distal interphalangeal joints. The lesions have persisted for 3 weeks despite treatments with moisturizing creams and topical steroids. She is fair-skinned and reports marked sun sensitivity. She has noted increased fatigue over the past few months.

Patient 2: A 53-year-old white male presents with pruritic, erythematous lesions on the backs of both hands and small papules across the knuckles. He also reports increased fatigue and muscle weakness.

What are your diagnoses?

PLEASE TURN TO PAGE 77 FOR DISCUSSION

The Diagnoses: Lupus Erythematosus and Dermatomyositis

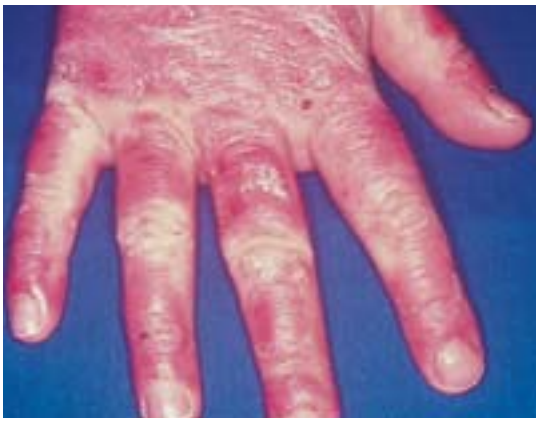


FIGURE 1. Lupus erythematosus.

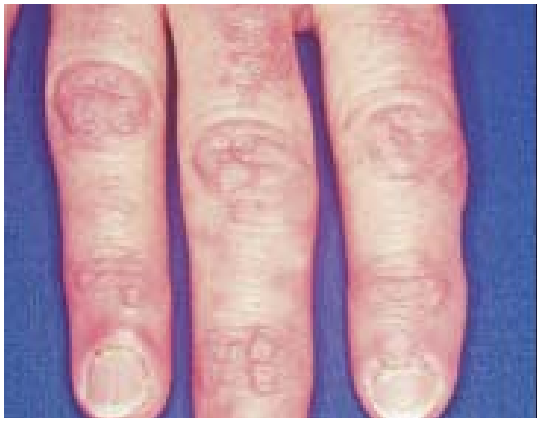


FIGURE 2. Dermatomyositis.

Connective tissue disorders are characterized by autoimmune injury of multiple organ systems. They may have diverse presentations and be difficult to differentiate because of a wide degree of overlap. It is critical to distinguish these diseases, however, as they have very different management and ultimate prognoses. In the examples presented, cutaneous hand manifestations of connective tissue disease, specifically lupus erythematosus (Figure 1) and dermatomyositis (Figure 2), can be used by the clinician as diagnostic clues to elucidate the underlying connective tissue disease process.

Lupus erythematosus (LE) is the most common autoimmune connective tissue disorder, with a prevalence of about 5 per 10,000 in North America and Europe.[1] Although it may develop at any time, it is most often seen between the ages of 20 and 40 years.[2] There is a higher prevalence of LE among women (female:male ratio of 9:1) and among certain ethnic groups such as African Americans and Hispanics.[1,2]

LE is a heterogeneous disease with multiple manifestations. Four of the 11 criteria for the diagnosis of LE are cutaneous (eg, malar rash, photosensitivity, discoid rash, oral ulcers).[3] In fact, most patients with lupus will have cutaneous manifestations at some point during their disease.[2,4]

Acute cutaneous LE is seen in the setting of systemic LE. Classically, the patient presents with a "butterfly rash," a pattern of malar erythema that extends over the bridge of the nose and spares the nasolabial folds.[2,5] Patients may exhibit extreme photosensitivity and develop a widespread rash over the neck, arms, and back of their hands. These cutaneous hand manifestations may appear as a coalescence of nonpruritic, erythematous plaques and patches that classically spare the metacarpophalangeal, proximal interphalangeal, and distal interphalangeal joints.[3] Patients may also exhibit periungual erythema; nailfold microscopy reveals tortuous, disorganized capillary loops without dilatation or areas of avascularity.[4,6] Systemic manifestations may include Raynaud's phenomenon; arthritis; fatigue; and serositis, which may manifest as pulmonary or pericardial effusions.[1-3,5]

Patients with subacute cutaneous LE exhibit skin manifestations without the systemic symptoms. The lesions may be either annular or papulosquamous, and present in a photosensitive distribution along the trunk, arms,

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and hands.[3,4] Acute cutaneous LE and subacute cutaneous LE may have identical cutaneous manifestations on the hands. Differentiation with history, physical examination, and serologic testing is required.

The prognosis for patients with LE is quite good. With frequent monitoring of disease activity and treatment during flares, the average survival at 5 years is greater than 90%.[1] Cutaneous manifestations usually can be well controlled, and most LE deaths are related to renal complications, vascular, neurologic events, or infection.[1]

Dermatomyositis (DM) is an immune-mediated, inflammatory myopathy with characteristic cutaneous manifestations; polymyositis is a related condition affecting the muscles but lacking any skin findings.[7,8] DM has a bimodal age distribution, occurring with peaks in childhood and again between the ages of 45 and 65 years.[6] In adults, DM has been found to be associated with malignancy in 5% to 50% of all cases.[6,7] DM also may commonly overlap with the presence of other connective tissue diseases (eg, lupus, scleroderma, mixed connective tissue disorder).[6,9]

The diagnosis of DM is made based on 5 criteria: symmetrical proximal muscle weakness, increased serum muscle enzymes, abnormal electromyography studies, abnormal muscle biopsy, and cutaneous findings.[6] The pathognomonic cutaneous features include a heliotrope rash and Gottron's papules.[3,6,7,9] A heliotrope rash is a violaceous discoloration around the eyes with periorbital edema. Gottron's papules consist of round, smooth, flat-topped, red to violaceous papules smaller than 1 cm that are found over bony prominences such as the metacarpophalangeal, proximal interphalangeal, and distal interphalangeal joints of the hands, as well as the elbows and knees.[3,6,9] Patient's nails often exhibit periungual erythema with cuticular hypertrophy, and capillary microscopy reveals dilated, irregular loops interrupted by areas of avascularity.[3,10] Less specific cutaneous features of DM include a pruritic, violaceous discoloration with scaling present in a photo distribution pattern over the arms and neck, poikiloderma, and scalp dermatitis.[3,8]

A work-up for malignancy should be completed when considering the diagnosis of DM. Often times the DM will resolve with treatment of the underlying cancer; a recurrence of DM may signal a recurrence of malignancy.[6] The overall prognosis for DM is estimated to be between 70% and 80% survival at 5 years, with lower survival rates associated with those cases affected by malignancy.[3]

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