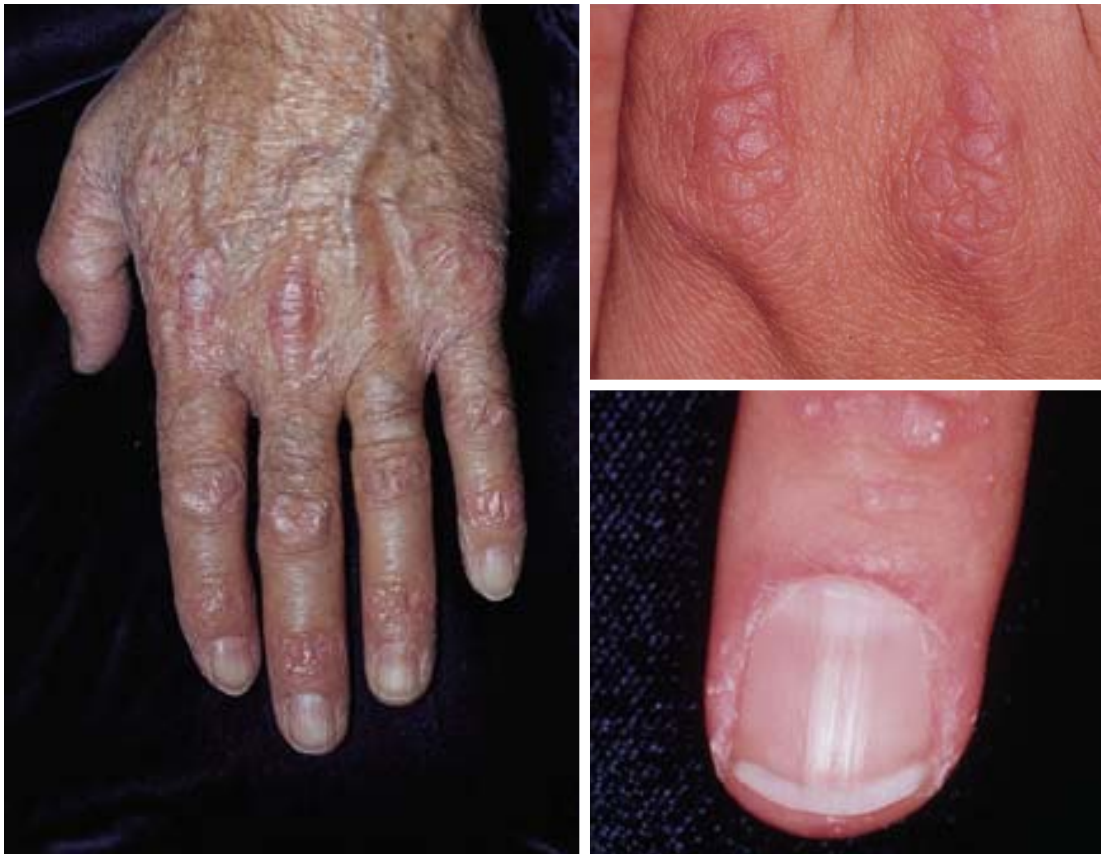
What Is Your Diagnosis?



These patients have the same condition.

PLEASE TURN TO PAGE 39 FOR DISCUSSION

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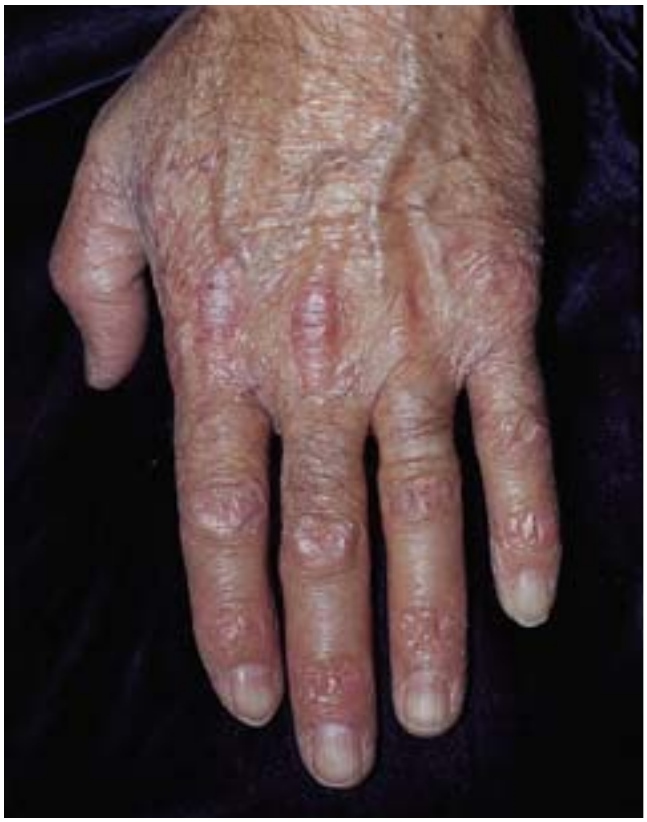
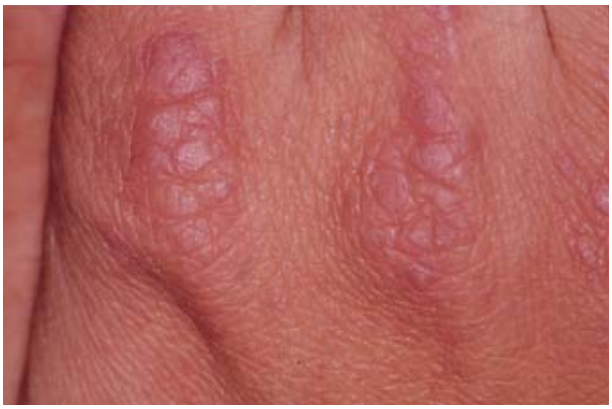
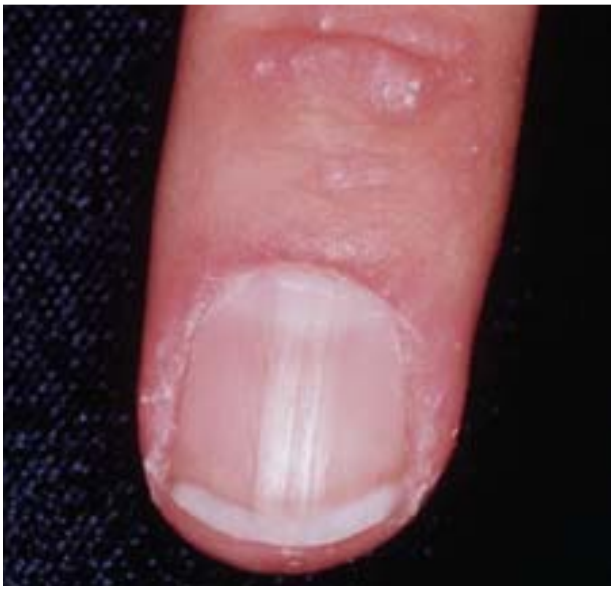
The Diagnosis: Dermatomyositis







Cutaneous signs of dermatomyositis include Gottron's papules (flat-topped pink-to-red papules overlying the interphalangeal joints) and Gottron's sign (pink-to-violaceous macular erythema, sometimes accompanied by cutaneous atrophy, overlying the interphalangeal joints and extensor surfaces). Nail folds demonstrate telangiectasia, focal avascular areas, and ragged cuticle. Additional cutaneous signs include periorbital heliotrope rash and involvement of the skin on the shoulders (shawl sign). As skin lesions evolve, they may become atrophic or mildly hyperkeratotic. Scalp involvement and pruritus are common. Usually skin and muscle disease appear concurrently; however, in approximately 30% of patients with dermatomyositis, skin disease precedes symptoms of muscle disease by up to 6 months.[1]

Gingival telangiectases, a finding analogous to nail fold telangiectases, can be especially prominent in juvenile dermatomyositis and may be accompanied by oral mucosal edema, erosions, ulcers, or white mucosal patches.[2]

The association of dermatomyositis with malignancy is controversial. Polymyositis does not appear to be independently associated with malignancy; however, dermatomyositis may be paraneoplastic. Therefore, age-appropriate screening for malignancy is advisable. Ovarian cancer deserves special mention, as this tumor is often asymptomatic until late in its course, and the presence of dermatomyositis may be the only presenting sign.[3]

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In cases of paraneoplastic dermatomyositis, relapse of skin and muscle disease may indicate recurrence of the tumor.[4]

Amyopathic dermatomyositis presents with skin findings identical to those of classic dermatomyositis. However, patients with amyopathic dermatomyositis lack clinically evident muscle disease. The term *hypomyopathic dermatomyositis* has been used for patients with laboratory or radiologic evidence but no clinical signs of muscle disease.[5] Amyopathic dermatomyositis also may occur as a paraneoplastic condition.[6]

Treatment with immunosuppressive drugs is successful in most patients with dermatomyositis and has greatly decreased the morbidity associated with this disease.[7] Although systemic corticosteroid therapy remains the treatment of choice for polymyositis and dermatomyositis, steroid complications are common, and skin disease may respond poorly to treatment with corticosteroids alone. Alternative therapeutic agents include methotrexate, cyclosporine, and intravenous immunoglobulin (IVIg). Although IVIg therapy is expensive, it can be effective in cases in which the disease is refractory to other agents.[8-10] The mechanism of action of IVIg in dermatomyositis deserves further study.[11] Recent evidence suggests that low-dose IVIg may be adequate for some cases of intractable cutaneous disease.[12] Mycophenolate mofetil is being used increasingly in dermatology and may prove to be a valuable agent in the treatment of dermatomyositis. Evidence suggests that more than half of patients with juvenile dermatomyositis initially treated with intravenous methylprednisolone pulse therapy respond within a few months and do not progress to chronic disease. Patients who fail to respond rapidly often progress to chronic disease and face a higher probability of disease and treatment-related complications.[13]

Pulmonary involvement, a particularly serious manifestation of dermatomyositis, may occur in 40% of patients and is associated with the presence of anti-Jo-1 antibody.[14] Interstitial pneumonitis may respond poorly to systemic corticosteroid therapy. Corticosteroid resistance is more common in patients who lack creatine phosphokinase elevations at the onset of pneumonitis. Cyclosporine can significantly prolong survival in patients with corticosteroid-resistant interstitial pneumonitis.[15] Low-dose methotrexate and systemic corticosteroids have been used effectively to treat cases of interstitial pneumonitis that are refractory to cyclosporine.[16] Cyclophosphamide pulse therapy also may be effective in treating severe refractory cases of dermatomyositis-associated interstitial pneumonitis.[17]

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