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Images in Emergency Medicine: Dermatomyositis

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A 41-year-old Guatemalan woman with a history of remote uterine cancer presented to an urban community Emergency Department complaining of a pruritic, initially painful rash on her chest, eyelids, thighs, and elbows for three weeks. She also stated she had weakness in her thighs and shoulders with difficulty standing. She denied any new exposures (ex. foods, clothes, soaps, fragrances, medicines, or illicit drugs), recent travel, bites, or recent trauma. She denied fever, night sweats, weight loss, recent illnesses, or sick contacts. Her vital signs were normal and stable. On exam she was found to have circumferential, violaceous patches on her eyelids and proximal chest, and erythematous patches on the extensor surfaces of her elbows and in a malar distribution on her face. She was noted to have proximal thigh weakness and was unable to get up from a chair without assistance. The remainder of her exam, including sensation, cranial nerves, and distal motor function, was otherwise unremarkable. Laboratory studies were remarkable for an AST of 336, ALT of 158, LDH of 1948, CK of 9031, and ANA +. Her CBC and remaining metabolic panel were within normal limits. She was presumptively diagnosed

with dermatomyositis and referred to general medicine and rheumatology clinics where she was started on a course of Prednisone with marked resolution of her symptoms. She recovered without sequelae.

Dermatomyositis (DM) is part of spectrum of idiopathic inflammatory myopathies. It has a prevalence rate estimated at approximately one per 100,000 in the general population with a 2:1 female predominance, and peak incidence between the ages of 40 and 50. Diagnosis is largely clinical and includes symmetric, proximal muscle weakness, characteristic rash, and elevated muscle enzymes. Common skin findings include Gottron's sign (symmetric, roughened, erythematous skin changes over the extensor surfaces of the metacarpophalangeal and interphalangeal joints, elbows, or knees), heliotrope rash (a violaceous eruption on the upper eyelids sometimes accompanied by edema), and shawl sign (a diffuse flat erythema in a shawl-like distribution over the chest and shoulder, or a V-shaped pattern over the anterior neck and chest).

Both measurement of serum muscle enzyme levels (CK, LDH, aldolase, AST, ALT) and testing for the presence of



Figure 1. Gottron's Sign



Figure 2. Hiliotrope Rash and Erythroderm

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Figure 3. Shawl sign

myositis-specific autoantibodies play important diagnostic and prognostic roles in the assessment of patients with

suspected DM. Other diagnostic and prognostic modalities include electromyography, MRI, and tissue biopsy. Prognosis is related to the severity of symptoms. Significant associated complications are not uncommon and can include interstitial lung disease and malignancy, and to a lesser extent esophageal disease and myocarditis. Therapy should be guided primarily by the degree of motor involvement and consists primarily of glucocorticoids and supportive therapy.

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