DERMATOMYOSITIS AND THE LUNG


Take home points:
1. Dermatomyositis usually presents as proximal muscle weakness and characteristic skin changes
2. Know the skin changes associated with dermatomyositis as this is the key to diagnosis.
3. Approximately 30-40% of patients with dermatomyositis will have ILD; these patients are more likely to have anti-Jo-1 antibodies and may have a worse prognosis.

What do I need to know about dermatomyositis?
• Inflammatory myopathy (proximal muscle weakness) + characteristic skin changes; 25% will have joint involvement
• Characteristic rashes:
  − Heliotrope rash: periorbital violaceous/erythematous rash, sometimes assoc. with periorbital edema
  − Gottron’s papules: erythema over bony prominences, usually over MCP, PIP, DIP joints
  − Shawl sign: poikiloderma (atrophy, dyspigmentation, telangectasias); a photosensitivity rash usually in a V-shaped distribution around the neck.
  − Mechanic hands: rough, dry exfoliating skin on the palms and fingers
• Can be associated with lung involvement (see below) and cardiac involvement (rare)
• Diagnosis is based primarily on clinical findings; muscle biopsy can be helpful as well.
• Anti-Jo-1 antibody is neither sensitive nor specific. Patients with this antibody, however, may have a worse prognosis and are more likely to have lung disease.
• CK can be elevated as can serum aldolase but these can be normal in the case of Dermatomyositis-sine myositis (dermatomyositis without muscle weakness or pain).
• Risk of malignancy: approximately 25-40%, increases with age. If your patient has dermatomyositis, look for malignancy based on age and risk factors. There is a higher incidence of ovarian cancer in women with dermatomyositis. However, in a young male, testicular cancer is your best bet, whereas in older patients, think prostate cancer, lung cancer, and breast cancer.
• Treatment: bedrest can help; otherwise glucocorticoids are the mainstay of therapy for this disorder.

What about dermatomyositis and the lung?
• Approximately 30-40% of patients with dermatomyositis will have ILD.
• Patients with anti-Jo-1 antibodies are more likely to have lung disease associated with their dermatomyositis.
• Lung disease with dermatomyositis confers a worse prognosis as does an initial DLCO < 45%.
• Typically presents as an acute, antibiotic-resistant pneumonia; however, most common finding on pathology is NSIP. This may be due to a sampling bias because the sicker patients get biopsies. Therefore, BOOP may be the #1 finding in these patients.
• All patients with dermatomyositis should get screen with HRCT and PFT’s.
• Treatment: corticosteroids ± methotrexate or azathioprine or similar drugs for immunosuppression.

Search strategies on PubMed for this coversheet:
1. For dermatomyositis general reviews: dermatomyositis [ti] AND review [pt] AND jsubsetaim
2. For dermatomyositis + ILD: dermatomyositis [ti] AND interstitial lung disease AND jsubsetaim

Key: [ti] = title, [pt] = publication type, jsubsetaim = core clinical journals subset