Dyspnea Associated With Dermatomyositis

Kevin P Collins MSc RRT RPFT AE-C, Harsh Babbar MD, and Said H Soubra MD

Introduction

Dermatomyositis is an inflammatory muscle disease characterized by weakness of the skeletal muscles, in particular, the proximal muscles of the arms and thighs.[1] If untreated, it is associated with progressive deterioration, and the severity ranges from mild to severe and can include paralysis. The onset is slow, over months in the majority of patients, but some people have rapid onset and progression. Dermatomyositis pulmonary disease occurs in 5–30% of cases, depending on the diagnostic method, and typically involves inflammation of the lung tissue.[2]

Case Summary

A 39-year-old man presented to the emergency department with chief complaints of dyspnea, weakness, weight gain, and generalized edema. He had seen several physicians over several months previously but had no conclusive diagnosis. He had a 4-month history of declining health, which began with eye irritation, followed by swelling of his hands and joints over a period a several weeks, which progressed to involve his lower extremities and knees and gradually affected his thighs. He also developed progressively worsening dyspnea and joint pain during that period. During the initial assessment in the emergency department he presented with a cough but denied any hemoptysis. He had gained about 14 kg over the past 2–3 months.

We found no history of rheumatologic disease, heart disease, diabetes, hypertension, or hyperlipidemia. On the day of presentation in the emergency department he was not in acute distress, and was awake, alert, and cooperative. His temperature was 37.9°C, his heart rate was 102 beats/min, his respiratory rate was 24 breaths/min, his blood pressure was 152/85 mm Hg, and his S_{pO_2} on room air was 96%. His neck was supple and there was no jugular vein distention. His eyes had some redness and periorbital swelling, his lungs were clear to auscultation, and his heart sounds had no murmurs or gallops. His abdomen was soft and nontender, but there was some swelling and edema of the anterior wall. His hands and feet were edematous and there was joint swelling and inflammation. His skin had no rashes or lesions. He was admitted to the progressive care unit with telemetry and started on intravenous fluid. The initial diagnostic tests included hepatitis profile, electrocardiogram, liver panel, and rheumatology workup for inflammatory arthritis.

The laboratory results included an admission creatine phosphokinase of 9,508 U/L (reference range 21–232 U/L), which is consistent with rhabdomyolysis. JO-1 antibody was < 1.0 negative (reference range < 1.0 negative antibody index). Troponin was < 0.1 ng/mL (reference range < 0.4 ng/mL), and creatine kinase MB was 294.1 ng/mL (reference range 0.0–3.6 ng/mL). Liver function studies showed an elevated aspartate aminotransferase (serum glutamic-oxalocetic transaminase) of 433 U/L (reference range 0–35 U/L) and alanine aminotransferase (serum glutamic-pyruvic transaminase) of 313 U/L (reference range 4–36 U/L). He admitted to drinking alcohol daily and to smoking 2.5 packs of cigarettes per day. Rheumatoid factor was negative (reference range < 1.0 antibody index). Aldolase was elevated at 129 U/L (reference range ≤ 8.1 U/L).

Chest radiograph (Fig. 1) and computed tomogram (CT) (Fig. 2) both revealed diffuse bilateral interstitial infiltrates. The bilateral infiltrates were worrisome for pneumonia, but he had a normal white-cell count of 10×10^3 cells/L (reference range $4.1–11.2 \times 10^3$ cells/L) and no fever. We obtained a pulmonary consult. Bronchoscopy, lung biopsy, and muscle biopsy were planned. Bronchoscopy revealed mild chronic inflammation, with predominantly lymphocytic infiltration.

His FEV_1 was 2.48 L (55% of predicted), forced vital capacity was 3.44 L (57% of predicted), diffusion capacity

Mr Collins is affiliated with the Department of Respiratory Care, Texas State University, San Marcos, Texas. Drs Babbar and Soubra are affiliated with the Division of Pulmonary and Critical Care Medicine, Seton Medical Center, Round Rock, Texas.

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Correspondence: Kevin P Collins MSc RRT RPFT AE-C, Department of Respiratory Care, Texas State University, 601 University Drive, San Marcos TX 78666. E-mail: kc35@txstate.edu.

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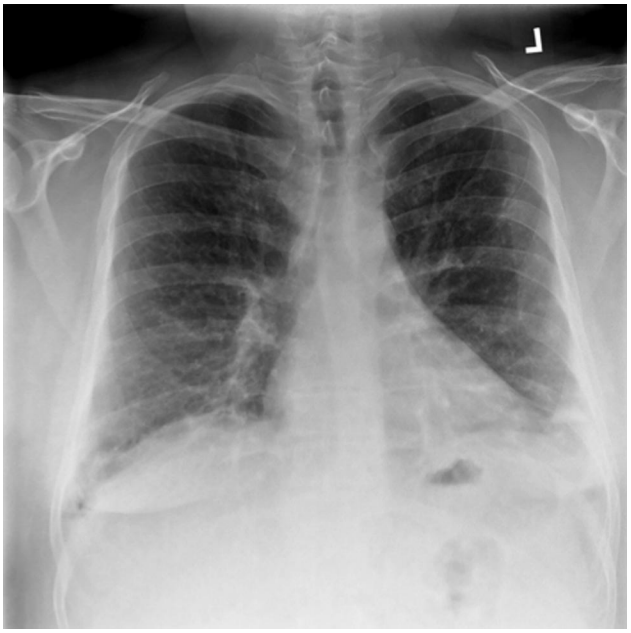

Fig. 1. Chest radiograph shows diffuse bilateral interstitial infiltrates.

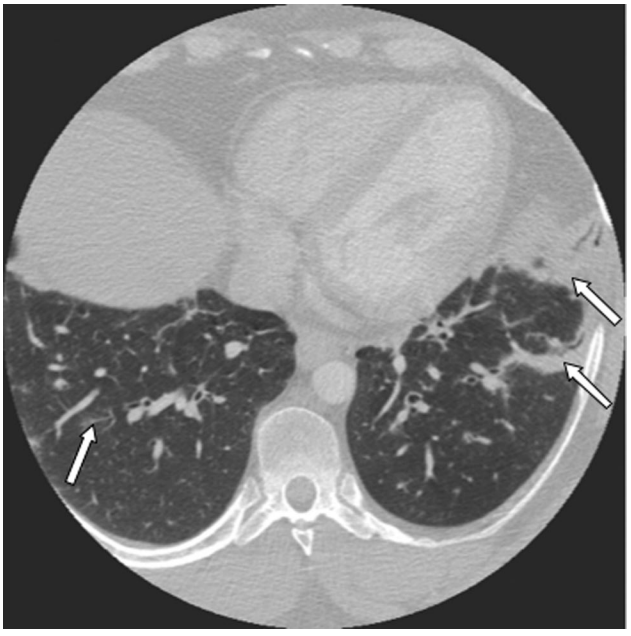

Fig. 2. Computed tomogram shows diffuse bilateral interstitial infiltrates (arrows).

of the lung for carbon monoxide was 32.9 mL/mm Hg/min (76% of predicted), total lung capacity was 5.49 L (65% of predicted), residual volume was 2.06 L (73% of predicted), and functional residual capacity was 2.43 L (74% of predicted). His maximum inspiratory and expiratory pressures were normal. Electrocardiogram was normal. Echocardiogram revealed a normal ejection fraction and no segmental wall motion.

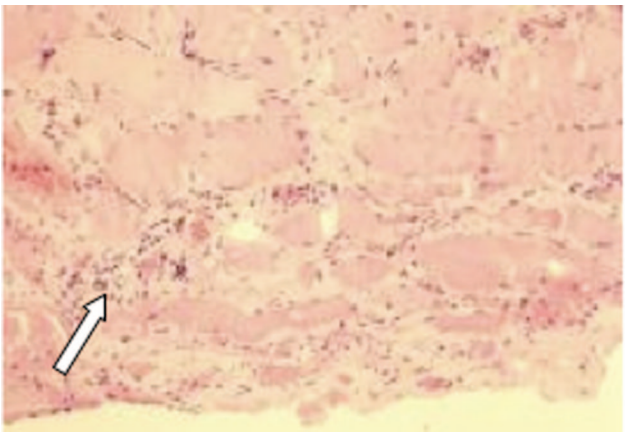

Fig. 3. Representative muscle biopsy micrograph (from another patient) shows inflammatory myopathic changes seen in dermatomyositis. (From Reference 3, with permission.)

He was also seen by a neurologist and the consensus was to perform muscle biopsy to obtain a definitive diagnosis. The muscle biopsy confirmed severe dermatomyositis, but because a muscle biopsy image was not available, Figure 3 shows a representative example of inflammatory myopathic changes seen in dermatomyositis. He was started on high-dose corticosteroids and had marked improvement of his respiratory symptoms and some improvement of his muscle weakness. He was discharged from the hospital with improving symptoms.

Discussion

Our patient presented to the emergency department with dyspnea, proximal muscle weakness, generalized edema, decreased exercise capacity, positive skin changes, a restrictive pulmonary defect, and bilateral interstitial infiltrates, which suggested dermatomyositis. The clinical presentation of dermatomyositis and polymyositis have similarities (eg, proximal muscle weakness and elevated serum muscle enzymes), but dermatomyositis also presents with a typical reddish-purple rash (heliotrope rash, Fig. 4).[4] Our patient presented with periorbital redness and swelling. Cutaneous manifestations may also include Gottron papules (Fig. 5), which are red-purple keratotic lesions on the extensor surfaces of the finger joints.[6] Although our patient did not have heliotrope rash or Gottron papules, they are important symptoms of dermatomyositis. Other symptoms include fever (45%), dyspnea (30–80%), cough (45–100%), musculoskeletal symptoms (35%), rash (11%), Raynaud phenomenon (20%), and dysphagia (10%).[4] Dermatomyositis presents acutely in about a third of patients, and 6% have rapidly progressive pneumonitis.[2]

Although dermatomyositis and polymyositis are part of a group of conditions known to cause muscle weakness, the occurrence is rare: 1–10 in 1,000,000.[2] Dermatomyo-

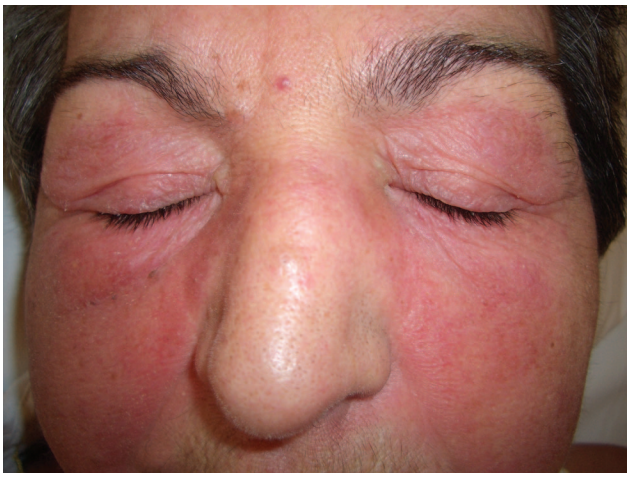

Fig. 4. Heliotrope rash in a woman with dermatomyositis. (Courtesy of Eleni Theodorakopoulou, Dermatlas, http://www.dermatlas.org.)

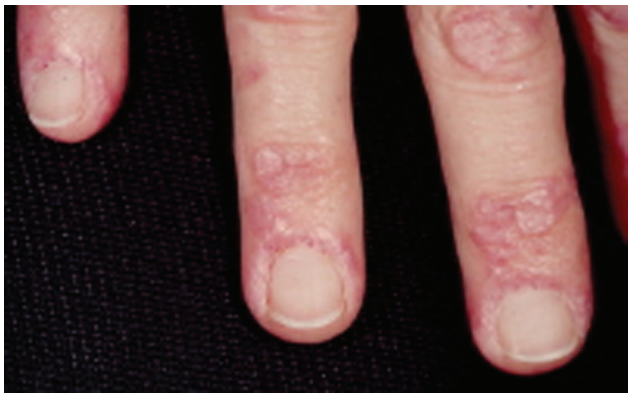

Fig. 5. Gottron papules on a representative patient with dermatomyositis. (From Reference 5, with permission.)

sitis is an autoimmune disease characterized by the presence of auto-antibodies and tissue inflammation, primarily involving the muscles.[2] Various auto-antibodies to nuclear and cytoplasmic antigens are characteristic of polymyositis and dermatomyositis.[7] The antigens at which these auto-antibodies are directed include aminoacyl-*t*RNA (*t* ribonucleic acid) synthetase (histidyl-tRNA synthetase, JO-1 antibody).[7] The JO-1 antibody is found only in a small percentage of the general population with myositis, but is much more common in polymyositis than in dermatomyositis.[7]

The diagnosis of dermatomyositis is made on the following findings:

- Proximal muscle weakness
- Muscle biopsy findings consistent with myositis
- Elevation of serum muscle enzymes
- Myopathic changes on electromyogram
- Heliotrope rash[2]

In our patient, muscle biopsy confirmed the diagnosis of severe dermatomyositis. Extramuscular involvement is common and includes systemic and vascular symptoms, dysphagia, cardiac disturbances, subcutaneous calcifications, and pulmonary dysfunction.[2]

Pulmonary involvement in patients with dermatomyositis can increase morbidity and mortality. In our patient the pulmonary involvement manifested as dyspnea and decreased exercise capacity. Pulmonary function testing revealed a restrictive lung defect, and there were bilateral interstitial infiltrates on CT. Several studies have described CT findings of interstitial lung disease in patients with dermatomyositis/polymyositis.[3,8] Bonnefoy et al reported that, in 18 patients with dermatomyositis/polymyositis-associated interstitial lung disease, high-resolution CT showed ground-glass opacities (85%), consolidation (55%), septal lines (55%), traction bronchiectasis (50%), reticulation (40%), subpleural lines (20%), parenchymal bands (15%), honeycombing (15%), micronodules (5%), and cyst (5%).[3,8]

The pulmonary problems may be divided into primary complications (the interstitial lung diseases) and secondary complications (aspiration pneumonitis, infection, and drug-induced disease).[2] Interstitial lung disease complicates dermatomyositis in 5–30% of the cases.[2] The mean age at presentation is 50 years.[2] In patients with dermatomyositis/polymyositis and interstitial lung disease, the interstitial lung disease may precede, coincide with, or follow the muscular manifestations.[4] Another finding is that the extent or severity of the inflammatory involvement of the muscles does not correlate with the occurrence of intrinsic lung disease.[2] Other pulmonary manifestations of dermatomyositis are classified by histopathology and include bronchiolitis obliterans organizing pneumonia, pulmonary capillaritis, alveolar hemorrhage, and diffuse alveolar damage.[2] Additional types of pulmonary involvement in polymyositis/dermatomyositis include respiratory muscle myositis, diaphragm dysfunction, pleural effusions, and pneumomediastinum.[2]

Due to a lack of adequately powered randomized controlled trials, the optimal therapy has not been determined.[3,8] Corticosteroids have been the mainstay of therapy in treating idiopathic inflammatory myositis, but their efficacy has not been tested fully in randomized controlled trials.[9] Most physicians agree that corticosteroids improve the symptoms in the majority of patients with inflammatory myositis.[1] In one study the initial treatment in 67 of the 70 patients included corticosteroids, usually in the form of oral prednisone (most commonly 40–60 mg/d), with occasional use of intravenous hydrocortisone.[10]

A treatment regimen that has been advocated includes oral high-dose prednisolone (60 mg/d) initially.[1] Over a 3-year treatment period with high-dose corticosteroids the majority of patients had improvement in disability, but one study found that the mortality and morbidity associated with dermatomyositis and polymyositis remained high despite corticosteroids.[9] Long-term corticosteroids have had adverse effects in 32–41% of patients with dermatomyositis and polymyositis.[9]

Other treatments include immunosuppressive agents, methotrexate, and cyclosporine, which are used in patients with refractory dermatomyositis or polymyositis.[9] Two reviews suggested that cutaneous dermatomyositis responds well to methotrexate: there was improvement in all the patients, which allowed a reduction in the corticosteroid dose.[11,12] Our patient responded well to high-dose corticosteroids, which markedly improved his symptoms.

Teaching Points

- Dermatomyositis is an inflammatory muscle disease characterized by proximal muscle weakness, elevated serum muscle enzymes, and a reddish-purple rash (heliotrope rash).
- Onset is slow, over months, in the majority of patients, but some people have rapid onset and progression.
- Elevated creatine phosphokinase increases the suspicion of an inflammatory muscle disease, and muscle biopsy is the definitive diagnostic tool for inflammatory myopathies.
- Pulmonary complications of dermatomyositis can include interstitial lung disease, and CT may show ground-glass opacities, consolidation, septal lines, traction bronchiectasis, reticulation, subpleural lines, parenchymal bands, honeycombing, micronodules, and/or cyst. Pulmonary function tests may show a restrictive lung defect.
- Optimal treatment of dermatomyositis has not been determined. Corticosteroids have been the mainstay of therapy. Without treatment, dermatomyositis may progress from mild to severe, and can cause paralysis.

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