

# Recurrent Episodes of Dermatomyositis-Associated Pneumonitis Masquerading as Hypersensitivity Pneumonitis

BOBBAK VAHID, M.D., AND PAUL E. MARIK, M.D.

## Abstract

This case report describes a unique presentation of dermatomyositis-associated pneumonitis. A 44-year-old man presented with repeated episodes of fever, dyspnea, and hemoptysis accompanied with pulmonary infiltrates, on chest CT scan. Hypersensitivity pneumonitis was suspected. Further work-up showed clinical and serologic evidence of dermatomyositis-associated pneumonitis. The patient was treated with oral prednisone and azathioprine. The subject of dermatomyositis-associated pneumonitis is discussed.

**Key Words:** Hypersensitivity pneumonitis, dermatomyositis, interstitial lung disease.

## Introduction

DERMATOMYOSITIS is a systemic inflammatory disease characterized by involvement of skeletal muscles and skin rash. Pulmonary involvement has been reported in 5–30% of patients with dermatomyositis (DM) and polymyositis (PM), and pulmonary disease has contributed to the death of 10% of patients in some case series. The dermatomyositis-associated pneumonitis may present as non-resolving pneumonia or acute respiratory failure (1, 2). Recurrent episodes of such pneumonitis mimicking hypersensitivity pneumonitis, as described in this case, are an unusual manifestation of this disease.

## Case Report

A 44-year-old Caucasian male was referred to the pulmonary clinic for evaluation of his pulmonary disease. The patient had had three episodes of pneumonitis, hemoptysis, and respiratory failure in the preceding 6 months. The manifestations

of pneumonitis included hemoptysis, fever, hypoxemia, and non-productive cough. Each episode was treated with antibiotics and corticosteroids. The patient also reported dyspnea with exertion, fatigue, diffuse muscle pain, diffuse arthralgia, and back pain. Close review of the course of his pulmonary disease suggested that the recurrence of pneumonitis corresponded to rapid taper of corticosteroids. The patient also had a history of gastroesophageal reflux disease, intermittent asthma, hypertension, and chronic sinusitis. His medications on presentation included amlodipine, esomeprazole, and montelukast. The patient was a lifelong non-smoker who worked as a jeweler and had been exposed to polishing wax (brand name: ZAM). He was evaluated in another center before presentation; while he was there, his lung disease was diagnosed as hypersensitivity pneumonitis secondary to polishing wax exposure.

Physical examination revealed bilateral pulmonary crackles, with normal cardiac, abdominal and neurological findings. Skin evaluation showed a symmetric, scaly, violaceous or erythematous eruption over the extensor surfaces of the metacarpophalangeal and interphalangeal joints of the fingers. Laboratory results included white blood cell count of  $17.3 \times 10^3/\text{mL}$ , normal creatinine, erythrocyte sedimentation rate of 10 mm/hr, normal serum complement levels, normal serum creatine phosphokinase (CK) levels, normal C-reactive protein, negative anti-nuclear antibody, negative

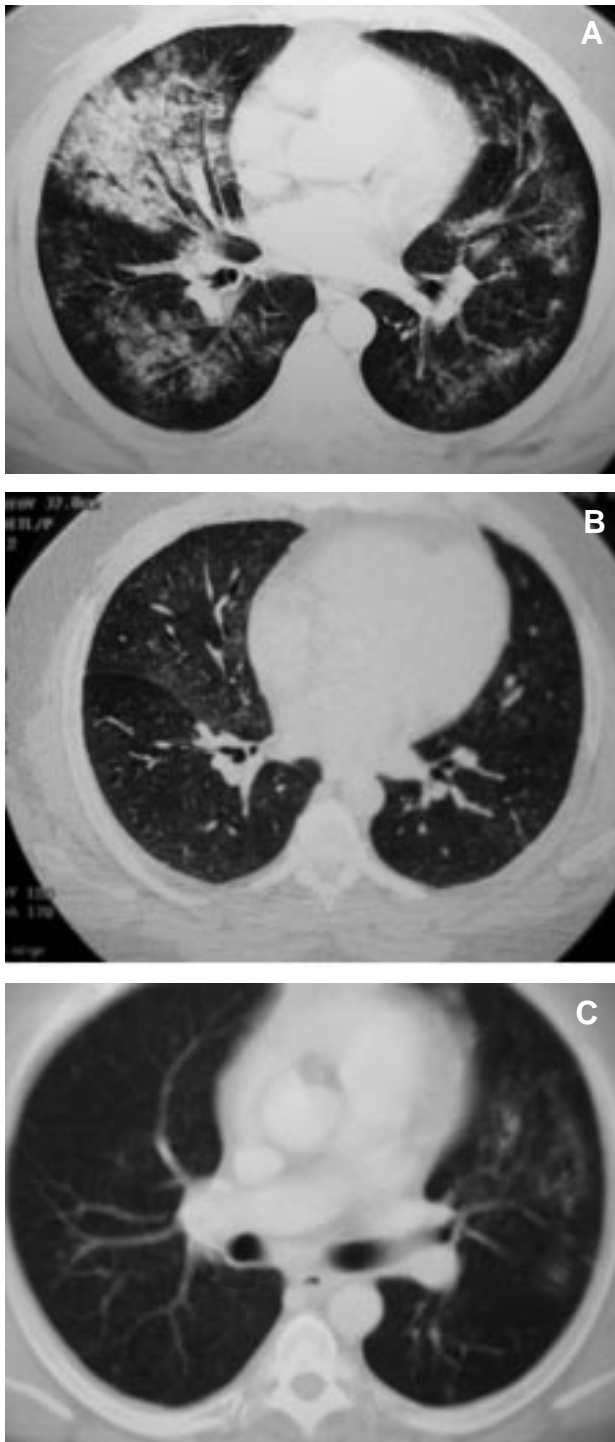
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Department of Pulmonary and Critical Care Medicine, Thomas Jefferson University, Philadelphia, PA.

Address all correspondence to Bobbak Vahid, M.D., 1015 Chestnut Street, Suite M-100, Philadelphia, PA 19107; email: Bobbak.Vahid@mail.tju.edu

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P-ANCA, negative C-ANCA, and normal AST and ALT. Further evaluation showed high serum aldolase level and positive anti Jo-1-antibody. Fig. 1 shows chest computed tomography (CT) scan find-



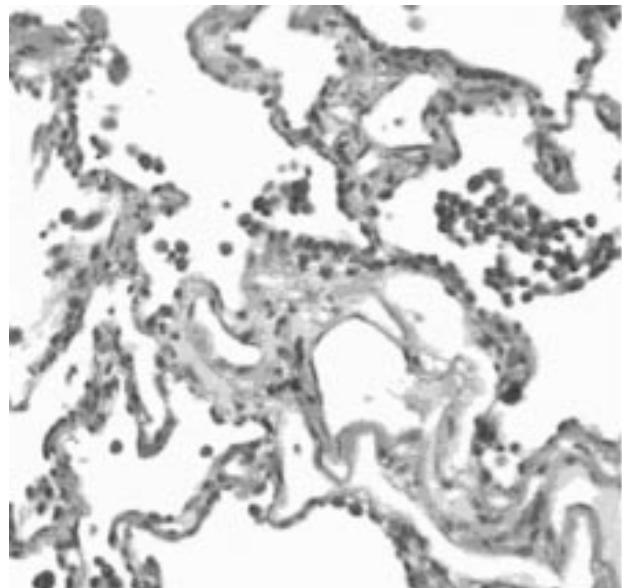
**Fig. 1.** (A) Chest CT scan showing bilateral patchy consolidations and ground-glass opacities 6 months before presentation. (B) Chest CT scan after a course of oral prednisone, showing complete resolution of infiltrates. (C) Chest CT scan 2 months before presentation, showing recurrence of pneumonitis.

ings and Fig. 2 shows the lung biopsy that was obtained via bronchoscopy. The lung biopsy showed nonspecific interstitial pneumonitis. The Table shows the patient's pulmonary function testing results during the 6 months before presentation. The decrease in lung volumes and single breath CO diffusion capacity (DLCO) suggests progressive interstitial lung disease.

### Discussion

The findings of typical skin rash (Gottron's sign), muscle pain and weakness, high aldolase, positive anti Jo-1-antibody, and nonspecific interstitial pneumonitis (NSIP) are consistent with a diagnosis of dermatomyositis-associated interstitial lung disease. NSIP can also be seen in hypersensitivity pneumonitis, but hypersensitivity pneumonitis was excluded by simply observing further flare-ups of pneumonitis without exposure to workplace or polishing wax.

Pulmonary involvement has been reported in 5–30% of patients with DM and PM, and pulmonary disease has contributed to the death of some 10% of patients in some series (1, 3). Differential diagnosis of pulmonary involvement in DM-PM includes interstitial lung disease (ILD), infection, aspiration pneumonia, respiratory muscle weakness, and drug-induced lung disease. ILD with myositis is rarely associated with malignancy. A majority of patients with PM-DM-associated



**Fig. 2.** Lung biopsy showing uniform involvement of lung parenchyma with prominent accumulation of alveolar macrophages, mild-to-moderate fibrosis of alveolar septae, and mild interstitial chronic inflammation consistent with nonspecific interstitial pneumonitis.

**TABLE**  
*Pulmonary Function Testing Showing Decrease in FVC and DLCO*

	<b>5 Months before Presentation</b>	<b>3 Months before Presentation</b>	<b>One Month before Presentation</b>
FVC L(% predicted)	4.33 (92%)	3.61 (77%)	3.26 (70%)
FEV1L(% predicted)	3.88 (101%)	3.17 (82%)	2.81 (73%)
%FEV1	89%	88%	86%
TLC L(% predicted)	5.58 (90%)	4.65 (75%)	4.21 (68%)
DLCO ml/min/mmHg(% predicted)	28.72 (80%)	23.33 (65%)	19.7 (55%)

FEV1 = forced expiratory volume in the first 1 second of expiration; FVC = forced vital capacity; TLC = total lung capacity; DLCO = diffusing capacity for carbon monoxide.

ILD present with pulmonary symptoms mimicking community-acquired pneumonia that is refractory to antibiotic therapy. A less common presentation is fulminant course with adult respiratory syndrome (ARDS) and respiratory failure (1, 3).

Pulmonary involvement can precede, coincide with, or follow muscular and/or cutaneous manifestations of PM-DM (3). Antibodies directed against the aminoacyl tRNA synthetase, including anti Jo-1-antibody, are most specific for inflammatory myopathies and ILD. The antisynthetase antibodies are associated with fever, Raynaud's phenomenon, polyarthritis, myositis, and ILD (4). Histopathology of PM-DM-associated ILD includes NSIP, usual interstitial pneumonia (UIP), bronchiolitis obliterans with organizing pneumonia (BOOP), and diffuse alveolar damage (DAD) (2). Bronchoscopy and bronchoalveolar lavage may be helpful to exclude infection, but don't provide specific diagnostic information (4). Treatment always includes corticosteroids. Better prognosis has been observed in patients with organizing pneumonia, those with a predominantly active cellular infiltration rather than fibrosis on CT or lung biopsy, younger patients, and those with an ele-

vated CK at the onset of ILD. Other reported treatments are azathioprine, methotrexate, pulse cyclophosphamide for initial remission, and hydroxychloroquine as a steroid-sparing agent (2, 5, 6).

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