

Acute Unilateral Pneumonitis in a Patient With Polymyositis: A Unique Presentation of Polymyositis-Associated Pneumonitis

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We describe a unique presentation of polymyositis-associated pneumonitis. A 45-year-old man with a history of polymyositis presented with an episode of fever, cough, dyspnea, rapidly progressive respiratory failure, and unilateral pulmonary infiltrates. Although bacterial pneumonia was initially suspected, all cultures, including bronchoalveolar cultures, remained negative, and the patient's condition worsened despite wide-spectrum antibiotics. Lung biopsy showed organizing pneumonia. The patient was treated with systemic corticosteroids and had complete resolution of respiratory failure and pulmonary infiltrates. We discuss polymyositis/dermatomyositis-associated pneumonitis. Key words: pneumonitis, polymyositis, respiratory failure. [Respir Care 2007;52(12):1774–1778. © 2007 Daedalus Enterprises]

Introduction

Polymyositis is a systemic inflammatory disease characterized by involvement of skeletal muscles. Pulmonary disease is common in patients with polymyositis/dermatomyositis. Pulmonary disease in polymyositis/dermatomyositis may be due to interstitial lung disease (ILD), infection, aspiration pneumonia, drug-induced lung disease, or respiratory muscle weakness. Polymyositis/dermatomyositis-associated pneumonitis may present as rapidly progressive respiratory failure, nonresolving pneumonia, or recurrent episodes of pneumonitis.^{1,2} Unilateral pneumonitis, as described in this case, is an unusual manifestation of this disease.

Case Summary

A 45-year-old man presented to the emergency department with 1 week of fever, progressive dyspnea, and cough.

At the time of this study Bobbak Vahid MD was, and Enrique Machare-Delgado MD and Paul E Marik MD currently are affiliated with the Department of Pulmonary and Critical Care Medicine, Thomas Jefferson University Hospital, Philadelphia, Pennsylvania.

The authors report no conflicts of interest related to the content of this paper.

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Three days before this presentation he had been evaluated in the emergency department for the same symptoms, and a chest radiograph showed clear lung fields. He was sent home with a diagnosis of acute viral illness. On the day of this presentation he was in moderate respiratory distress. Physical examination showed a temperature of 38.3°C, heart rate of 118 beats/min, respiratory rate of 26 breaths/min, and blood pressure of 130/77 mm Hg. The oxygen saturation while breathing ambient air was 88%. No jugular neck distention was noted. Lung auscultation revealed crackles at the right base. Skin, cardiac, and abdominal examinations were unremarkable. Cyanosis, clubbing, and peripheral edema were absent.

The chest radiograph showed right-sided pulmonary infiltrates (Fig. 1). Laboratory results included a white-blood-cell count of $14.2 \times 10^3/\text{mL}$, normal serum creatinine, erythrocyte sedimentation rate of 50 mm/h, serum creatine phosphokinase of 1,340 U/L (normal range 32–250 U/L), antinuclear antibody titer of 1:1,280, serum aldolase of 16.5 U/L (normal range 1.0–7.5 U/L), and positive anti-Jo-1 antibody.

The patient's medical history was notable for polymyositis, which was diagnosed 14 years prior, via muscle biopsy and clinical and electromyogram findings. The patient was on prednisone 4 mg/d long-term. His muscle strength, aldolase, and creatine phosphokinase had been stable for several years. He had a pulmonary function test 1 year before this presentation, and his forced expiratory volume in the first second (FEV₁) was 2.38 L (73% of predicted), forced vital capacity was 3.78 L (73% of pre-



Fig. 1. Chest radiograph showing extensive right-lung infiltrate.

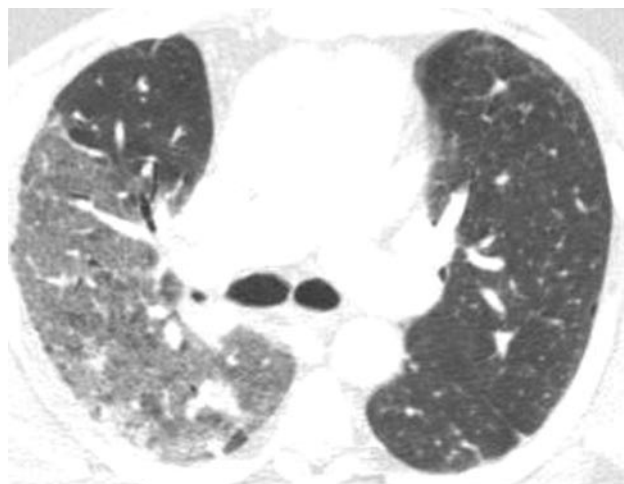


Fig. 2. Computed tomogram showing right-lung infiltrate.

dicted), and diffusion capacity of the lung for carbon monoxide was 26.4 mL/min/mm Hg (83% of predicted). He also was being treated for hypertension (metoprolol, hydrochlorothiazide, and losartan), diabetes mellitus (metformin), and hypercholesterolemia (lovastatin). The patient was a former smoker but had no other history of exposure to chemicals. He had a negative human immunodeficiency virus screening test 2 months before this presentation.

The patient was started on moxifloxacin and ceftriaxone for treatment of presumed community-acquired pneumonia. The initial sputum culture grew normal flora. Two sets of blood cultures were negative. Urine antigen test for *Legionella* was also negative. Shortly thereafter the patient developed respiratory failure that required intubation and mechanical ventilation. Computed tomography (CT) showed no evidence of pulmonary embolism, but extensive infiltrates in the right lung (Fig. 2). Bronchoscopy found no evidence of alveolar hemorrhage or endobronchial lesion. The bronchoalveolar lavage fluid (BALF) differential cell count showed 65% lymphocytes, 25% neutrophils, and 10% macrophages. The BALF cultures for bacterial, viral, *Legionella*, fungal, and mycobacterial pathogens were negative. Vancomycin and piperacillin/tazobactam were added to the treatment. Five days after intubation, the patient was febrile and hypoxemic (P_{aO_2}/F_{IO_2} 240 mm Hg). Lung biopsy of the right lower, right middle, and right upper lobes were obtained via video-assisted thoracic surgery. Tissue cultures were negative for microbial pathogens. The lung biopsy showed organizing pneumonia and interstitial pneumonitis (Fig. 3). Stains for mycobacteria, fungi, and *Pneumocystis jirovecii* (formerly known as *Pneumocystis carinii*) were negative. No viral inclusion bodies were found. Prednisone 60 mg/d was started. Two days after starting the prednisone, the P_{aO_2}/F_{IO_2} improved to 360 mm Hg, and the patient was subsequently extubated.

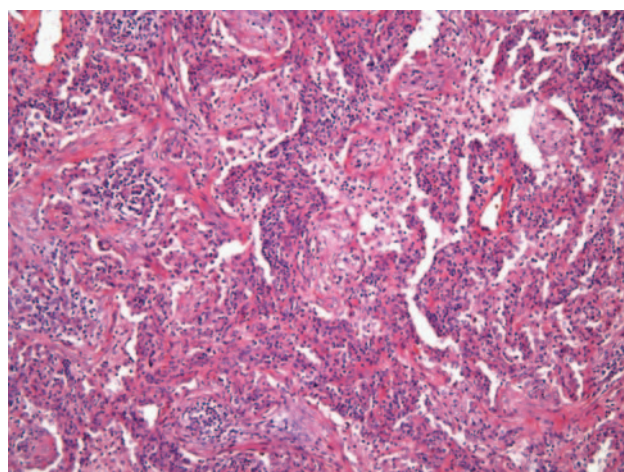


Fig. 3. Open lung biopsy specimen showing organizing pneumonia and interstitial pneumonitis (hematoxylin and eosin stain, magnification 200).

A barium swallow study after extubation showed normal esophageal motility, with no evidence of barium aspiration. One week after starting prednisone therapy the patient had no dyspnea and normal oxygen saturation while breathing ambient air. A 6-min walk test showed no exercise-induced hypoxemia. Follow-up chest radiograph 2 weeks after starting prednisone therapy showed marked improvement in the right-lower-lobe infiltrates (Fig. 4). Prednisone at 60 mg/d was continued for 6 weeks, then tapered over 3 months to 4 mg/d.

Discussion

This case describes a unique presentation of polymyositis-associated pneumonitis. The presentation of a unilateral right-lower-lobe pulmonary infiltrate accompanied



Fig. 4. Chest radiograph showing resolution of pulmonary infiltrates after steroid therapy.

by cough and fever suggested a diagnosis of bacterial community-acquired pneumonia or aspiration pneumonia. Although the patient was treated with appropriate antibiotics, he did not improve. Since the BALF and lung tissue cultures were negative, we excluded infectious pneumonia as a cause of respiratory failure. The patient had no history of dysphagia, choking on food, or previous aspiration. The barium swallow study and clinical swallow evaluation were normal after extubation. These findings argue against aspiration in our patient. The marked improvement with corticosteroid therapy in a patient with known polymyositis and positive anti-Jo-1 antibody strongly suggests polymyositis-associated pneumonitis as the cause of respiratory disease in this patient.

ILD is a common manifestation of polymyositis/dermatomyositis. The reported prevalence of pulmonary involvement in polymyositis/dermatomyositis ranges from 5% to 65%, depending on whether clinical, radiological, functional, or pathology criteria were used to diagnose the pulmonary involvement.^{3–5} Infection, aspiration pneumonitis, and drug-induced lung disease should be considered in the differential diagnosis. Severe opportunistic infections (eg, pneumocystis pneumonia, nocardiosis, cytomegalovirus pneumonia, and nontuberculous mycobacterial infection) have been described in patients with corticosteroid-refractory polymyositis/dermatomyositis on immunosuppressive therapy.^{6–9}

ILD in patients with polymyositis/dermatomyositis may precede, coincide with, or follow the muscular and/or cutaneous manifestations.¹⁰ Based on radiological and functional findings, Fathi et al found evidence of ILD in 65% of patients with newly diagnosed polymyositis/dermatomyositis.³ There are 3 main types of clinical presentation of polymyositis/dermatomyositis-associated ILD:

(1) bilateral pneumonitis with a clinical picture that mimics community-acquired pneumonia that persists despite appropriate antibiotic therapy; (2) chronic ILD, with gradual onset of cough, dyspnea, and pulmonary infiltrates; and (3) subclinical ILD, with abnormal chest images or pulmonary function test results but no respiratory symptoms.^{1,5,11} A rapidly progressive pneumonitis that results in respiratory failure and acute respiratory distress syndrome is a less common presentation. This form of fulminant pneumonitis can be the first clinical manifestation of polymyositis/dermatomyositis-associated ILD or may present as accelerated disease in a patient with chronic ILD.^{12–15}

Polymyositis/dermatomyositis-associated ILD also may present with recurrent pneumonitis and migratory pulmonary infiltrates that mimic hypersensitivity pneumonitis.² Although unilateral pneumonitis has not been previously mentioned in the literature, unilateral pneumonitis has been reported in systemic lupus erythematosus, rheumatoid arthritis, and drug-induced pneumonitis.^{16–20} Other types of pulmonary involvement in polymyositis/dermatomyositis include respiratory muscle myositis and diaphragmatic dysfunction, pleural effusions, and pneumomediastinum. Respiratory muscle myositis and pleural effusions are usually seen in more chronic disease; on the other hand, pneumomediastinum has been mostly described in patients with rapidly progressive pneumonitis.^{15,21–24}

Clinical symptoms include fever (45%), dyspnea (30–80%), cough (45–100%), musculoskeletal symptoms (35%), rash (11%), Raynaud's phenomenon (20%), and dysphagia (10%).^{25,26} About one third of patients have acute presentation²⁶ and 6% present have rapidly progressive (fulminant) pneumonitis.¹⁵ Acute presentation may be the initial manifestation of the ILD or superimposed on well-established chronic ILD pneumonitis. Synchronous malignancy (eg, breast cancer, renal cell cancer) has been reported in 5.7% of patients with polymyositis/dermatomyositis-associated ILD. Laboratory findings are elevated lactic dehydrogenase (85%), aspartate aminotransferase (84%), aldolase (80%), creatine phosphokinase (18%), and erythrocyte sedimentation rate > 30 mm/h (45%). Antinuclear antibodies are positive in only 40% of the patients.²⁵ Autoantibodies directed against the aminoacyl tRNA (t ribonucleic acid) synthetase (eg, anti-Jo-1, anti-Zo, anti-PL-7, anti-PL-12, anti-OJ, anti-EJ, anti-KS) are associated with myositis, arthritis, Raynaud's phenomenon, fever, and ILD, which are together clinically known as anti-synthetase syndrome.²⁷ Anti-Jo-1 antibodies are present in 19–75% of cases of ILD in patients with polymyositis/dermatomyositis, compared to 2.6–3% in patients without ILD.^{25,26,28}

Subtle differences have been described in patients with rapidly progressive pneumonitis and chronic ILD. Ito et al compared 7 patients with dermatomyositis and rapidly pro-

gressive pneumonitis to 9 patients with dermatomyositis and chronic ILD, and the degree of muscle weakness in the extremities and the frequency of positive autoantibodies (antinuclear autoantibody, anti-Jo1) were significantly lower in patients who had rapidly progressive pneumonitis than in those with chronic ILD.¹⁴ Although polymyositis-associated ILD and dermatomyositis-associated ILD are very similar, there are some differences. Polymyositis is primarily mediated by autoreactive cytotoxic T cells, whereas dermatomyositis is mainly caused by a humoral immune response, complement activation, and destruction of the muscular capillaries. It is possible that these differences in immunopathology may cause differences in the clinical pictures of polymyositis and dermatomyositis.²⁹ Fujisawa et al compared 16 patients with polymyositis-associated ILD to 12 patients with dermatomyositis-associated ILD, and found 2 differences; namely, the serum creatine phosphokinase concentrations were significantly higher in polymyositis-associated ILD than in dermatomyositis-associated ILD, and the bronchoalveolar lavage cell count showed significantly higher percentages of lymphocytes and eosinophils in dermatomyositis-associated ILD than in polymyositis-associated ILD. Based on response to corticosteroids in the 28 patients reported, the authors concluded that dermatomyositis-associated ILD may be more refractory to corticosteroids and may result in poorer prognosis.²⁹

There are 4 major histopathologic patterns reported in polymyositis/dermatomyositis-associated pneumonitis: (1) nonspecific interstitial pneumonitis, (2) usual interstitial pneumonia, (3) bronchiolitis obliterans organizing pneumonia, and (4) diffuse alveolar damage.^{1,15,30} In more recent studies, nonspecific interstitial pneumonitis has been reported in over 80% of the patients with polymyositis/dermatomyositis-associated ILD. Bronchiolitis obliterans organizing pneumonia and diffuse alveolar damage seem to correlate more with acute presentation, whereas nonspecific interstitial pneumonitis and usual interstitial pneumonia are associated with a more chronic course. Diffuse alveolar damage is associated with poor prognosis.^{1,15,30} Acute fibrinous and organizing pneumonia was recently described as a histologic pattern of acute lung injury. It is distinct from diffuse alveolar damage because this pattern lacks the classic hyaline membranes of diffuse alveolar damage, and it involves the lung in a more patchy distribution. The dominant pathology finding is the presence of intra-alveolar fibrin, in the form of fibrin "balls" within the alveolar spaces.³¹ Acute fibrinous and organizing pneumonia has been described in juvenile dermatomyositis.³² Although pulmonary capillaritis and diffuse alveolar hemorrhage are commonly associated with collagen-vascular diseases, it is rare in patients with polymyositis/dermatomyositis.³³ Polymyositis/dermatomyositis should be differentiated from mixed connective-tissue disease that

has polymyositis features, which has also been associated with diffuse alveolar hemorrhage.³⁴

Several studies have described chest CT findings of polymyositis/dermatomyositis-associated ILD.⁵ Mino et al followed 17 patients with polymyositis or dermatomyositis for a period of 2–61 months with sequential chest CTs. The common findings were pleural irregularities, prominent interlobular septa, ground-glass opacities, patchy consolidations, parenchymal bands, subpleural lines, and irregular peribronchovascular thickening.³⁵

Bonnefoy et al described the findings from high-resolution chest CT in 18 patients with polymyositis/dermatomyositis-associated ILD: 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%).⁵ Patchy peripheral consolidations, ground-glass opacities, parenchymal bands, and irregular peribronchovascular thickening tend to resolve with treatment, but honeycombing tends to progress.^{5,35} High-resolution CT patterns may correlate with the pathology findings. Diffuse alveolar damage lesions show extensive bilateral consolidation or ground-glass opacities; bronchiolitis obliterans organizing pneumonia shows patchy subpleural and peribronchovascular consolidations and subpleural band-like opacities. Usual interstitial pneumonia shows honeycombing.^{36,37}

Corticosteroids are the mainstay of therapy. A better response to corticosteroid therapy has been associated with younger patients, organizing pneumonia on lung biopsy, and consolidation on chest CT, instead of evidence of fibrosis. Azathioprine, cyclosporine, and pulse cyclophosphamide have been used in patients with severe or corticosteroid-refractory ILD.^{38–40}

Polymyositis-associated pneumonitis may present as a rapidly progressive unilateral pneumonitis that mimics severe community-acquired pneumonia. In selected cases, lung biopsy may provide important information that can lead to a change in therapy and outcome.

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