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July 2021 Critical Care Case of the Month: When a Chronic Disease Becomes Acute

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History of Present Illness

A 32-year-old woman with no known past medical history presented with progressive shortness of breath for 2 weeks. She denied having a cough, fever, or chills, but she did have a one-month history of fatigue, weakness, and painful rashes on her hands.

PMH, SH, and FH

- No known past medical history
- Former tobacco user (quit 2 years prior to admission)
- No drug use
- Worked as an office assistant
- Has two pet dogs and four pet macaws
- No family history of lung disease
- Not taking any prescription medications

Physical Exam

- BP: 116/65, Pulse: 105, T: 37°C, RR: 28, SpO2: 89% on HHFNC (60L; 100%)
- Pulmonary: Tachypneic, in respiratory distress, crackles throughout
- Cardiovascular: Tachycardic but regular, no murmurs
- Extremities: No edema
- Skin: Palms with purplish discoloration and erythematous papules

Radiography

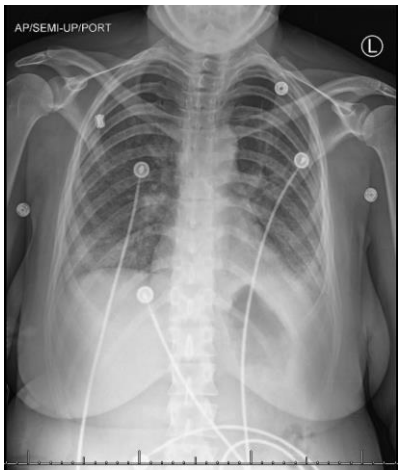

Figure 1. Initial portable chest x-ray.

Which of the following ***should be done*** next?

1. CT Chest
2. COVID-19 testing
3. Sputum gram stain and culture
4. 1 and 3
5. All of the above

Key words: pulmonary fibrosis, CT scan, NSIP, nonspecific interstitial pneumonitis, dermatomyositis, anti-MDA5, creatinine kinase, aldolase, antinuclear antibody, SSA

Correct!
4. All of the above

Her symptoms are nonspecific but could be suggestive of COVID-19. However, her COVID-19 antibody and DNA testing were negative. Her sputum Gram stain was unremarkable and she was treated with azithromycin for community-acquired pneumonia. Bronchoscopy is premature at this juncture. When she did not improve, she was transferred to the ICU for monitoring.

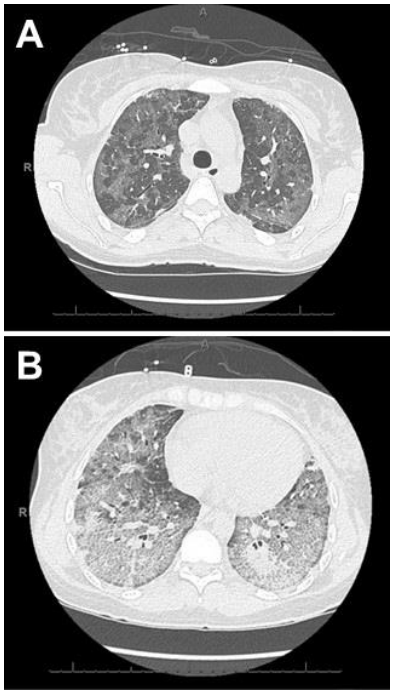

Figure 2. Representative lung windows from thoracic CT scan.

What ***radiographic pattern*** is seen in this CT scan?

1. IPF/UIP (Idiopathic pulmonary fibrosis/usual interstitial pneumonitis)
2. NSIP (nonspecific interstitial pneumonitis)
3. UIP (usual interstitial pneumonitis)
4. 1 and 3
5. All of the above

Correct!
2. NSIP

Nonspecific interstitial pneumonitis (NSIP) is temporally and spatially homogeneous, while UIP is typically heterogeneous, patchy, and irregular in size (1). Usual interstitial pneumonia (UIP) is the histopathologic and radiologic pattern of interstitial lung disease, which is the hallmark pattern for idiopathic pulmonary fibrosis (IPF). On imaging, usual interstitial pneumonia usually presents with a lung volume loss and a craniocaudal gradient of peripheral septal thickening, bronchiectasis, and honeycombing. The extent of honeycombing and traction bronchiectasis is greater in UIP than the extent of ground glass opacity or micronodules, which are more commonly associated with an NSIP pattern (1,2).

She stabilized in the ICU although she remained dyspneic. Her infectious workup was negative and she was diagnosed with interstitial lung disease (ILD).

What in her ***history is associated with developing an ILD***?

1. Painful skin rash
2. Working in an office
3. Pet birds
4. 1 and 3
5. All of the above

Correct!
4. 1 and 3

Exposure to birds might suggest hypersensitivity pneumonitis. Distinguishing between usual interstitial pneumonia and hypersensitivity pneumonitis can be difficult. Of course, old chest x-rays or CT scans can reveal a chronic process while normal prior radiography suggests an acute change. Thirty-six patients with idiopathic pulmonary fibrosis and 27 patients with hypersensitivity pneumonitis were studied. All diagnoses were confirmed or supported by open lung biopsy (3). A diagnosis of idiopathic pulmonary fibrosis was considered more likely in patients with honeycombing and peripheral or lower lung zone predominance of disease. In contrast, patients with micronodules but without honeycombing, were considered more likely to have chronic hypersensitivity pneumonitis (3). A CT diagnosis could be made with a high level of confidence in 39 (62%) of 63 patients. In these patients, the CT diagnosis was correct in 35 cases (90%): 23 of 26 patients with a CT diagnosis of idiopathic pulmonary fibrosis and 12 of 13 patients with a CT diagnosis of hypersensitivity pneumonitis.

A painful skin rash could suggest ILD especially disease secondary to dermatomyositis (4).

What laboratory evaluation might ***confirm a diagnosis of dermatomyositis-associated ILD***?

1. Liver function panel
2. Myositis panel (creatinine kinase, aldolase, antinuclear antibody, SSA)
3. BMP (brain naturetic peptide)
4. 1 and 3
5. None of the above

Correct!
2. Myositis panel

Her creatinine kinase (CK), aldolase, antinuclear antibody (ANA), and SSA were all positive highly suggestive of dermatomyositis.

What myositis-specific autoantibody is ***associated with rapidly progressive ILD***?

1. Anti-MDA5
2. Anti-SAE
3. Anti-Jo-1
4. All of the above
5. None of the above

Correct!
1. Anti-MDA5

Anti MDA5 is associated with a rapidly progressive ILD. It is typically diagnosed in patients with clinically amyopathic dermatomyositis (dermatomyositis without muscle weakness). Anti-MDA5 RP-ILD was originally reported in a study of Japanese patients with CADM and RP-ILD (5). Cutaneous findings typically include papules and ulcerations on the hands, alopecia, and oral ulcers.

Q: What is the ***treatment*** for anti-MDA5 RP-ILD?

1. Corticosteroids
2. Mycophenolate
3. Rituximab
4. All of the above
5. None of the above

Correct!
4. All of the above

Patients with anti-MDA-5 rapidly progressive ILD progress very quickly and may be refractory to conventional treatment such as corticosteroids. Experience is limited, but typically, a combination of immunosuppressive therapies is necessary (6). Because of this mortality with this condition remains very high.

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