Case Study Review*

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Examining case studies from patients with interstitial lung diseases (ILDs) is important in order to evaluate current diagnosis and treatment options. Two cases will be discussed; the first case examines a patient with usual interstitial pneumonia, and the second case examines a patient with subacute ILD and elements to suggest a *forme fruste* presentation of an unclassifiable connective tissue disease. Each case highlights components of the differential diagnosis, as well as reviews the treatments and prognoses of these patients. The cases provide clinical pearls that are designed to enhance the reader's understanding of ILDs. ***(CHEST 2005; 128:540S–546S)***

Key words: case study reviews; idiopathic interstitial pneumonia; interstitial lung disease; idiopathic pulmonary fibrosis

Abbreviations: ANA = antinuclear antibody; anti-Jo-1 = histidyl-t-RNA synthetase; DLCO = diffusing capacity of the lung for carbon monoxide; HRCT = high-resolution CT; ILD = interstitial lung disease; IPF = idiopathic pulmonary fibrosis; NSIP = nonspecific interstitial pneumonia; RNP = ribonucleoprotein; SCL = scleroderma; SSA = anti-Ro antibody; SSB = anti-La antibody; TLC = total lung capacity; UIP = usual interstitial pneumonia

Case 1: 68-Year-Old Man With Exertional Breathlessness

A 68-year-old radiologist presented to the pulmonologist with worsening exertional breathlessness that had become more apparent over the preceding 3 to 6 months. He reported being fairly active, including exercising regularly at a local gym. If he were to run up a flight of stairs or walk fast, he would become significantly short of breath. He reported an intermittent nonproductive cough, which had developed over the preceding several months. He had no history of respiratory disease, and was a lifelong nonsmoker. He described poor ventilation in the radiology reading room, in which he has worked for the past 30 to 35 years, and he reported a concern that this exposure contributed to the development of some type of lung disease. On review of systems, the patient reported occasional arthralgias, which are worse in the morning. He denied Raynaud symptoms, has experienced significant gastroesophageal reflux disease, but denied aspiration.

Physical Examination

Auscultation of the lungs revealed a few fine late inspiratory crackles at both bases with no wheezes, rhonchi, or other adventitious sounds. Pulmonary function studies revealed a mild restrictive ventilatory defect with the following results: FEV_1, 75% predicted; FVC, 77% predicted; total lung capacity (TLC), 75%; and diffusing capacity of the lung for carbon monoxide (DLCO), 68% (uncorrected for alveolar volume). Oxygen saturation declined from 95 to 92% during a "modified" 6-min walk test that included five flights of stairs. Digital clubbing was absent.

Laboratory Findings

The erythrocyte sedimentation rate was 20 mm/h, the test for antinuclear antibodies (ANAs) was positive at a titer of 1:40, the rheumatoid factor level was normal, the tests for anti-Ro antibody (SSA) and anti-La antibody (SSB) were negative, the tests for ribonucleoprotein (RNP) and scleroderma (SCL)-70 antibodies were negative, the aldolase level was normal at 5.6 U/L, and the test for the histidyl-t-RNA synthetase (anti-Jo-1) antibody was negative.

High-Resolution CT Scan Findings

A high-resolution CT scan (HRCT) of the patient's upper lung zones depicted minimal peripheral reticular opacities. Mid-lung zones exhibited fine reticular changes at the pleural surface with minimal changes in the peripheral areas and intermittent reticular abnormalities. Lower lung zones depicted asymmetric reticular changes predominantly on the right, with some cystic changes that occurred primarily in the middle of the lung rather than on the periphery. Honeycomb changes were present but not extensive (Figs 1-3).

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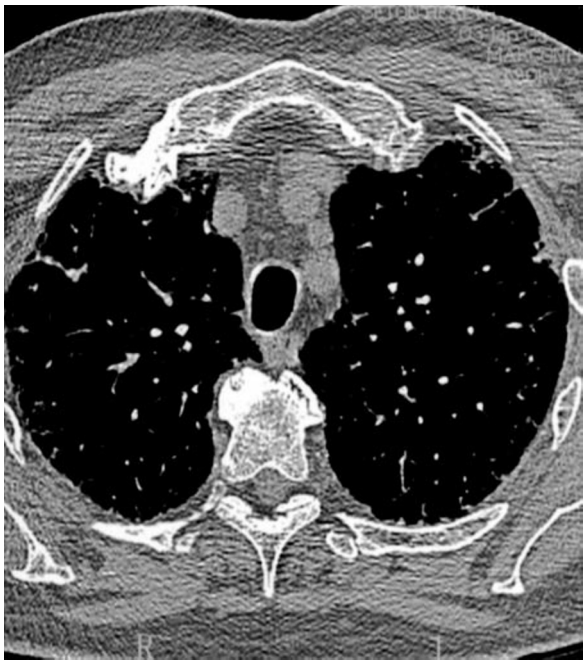

FIGURE 1. HRCT scan of upper lung zones reveals peripheral reticular infiltrates.

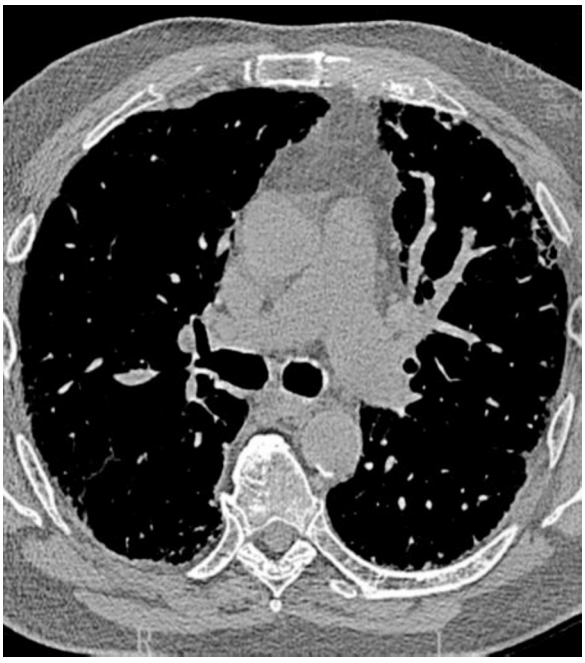

FIGURE 2. HRCT scan of mid-lung zones reveals additional peripheral opacities.

Pathology Findings

There was evidence of fibrosis with mild chronic inflammation adjacent to areas of fibrosis, and microscopic honeycombing was present. Temporal and spatial heterogeneity were also shown, with fibrosis at the periphery and a transition to areas of normal lung. Fibroblastic foci were present. The biopsy specimen was typical for usual interstitial pneumonia (UIP) [Figs 4–7].

Clinical Course

UIP was diagnosed. The patient was treated with prednisone and azathioprine for 6 months, according to the American Thoracic Society recommendations.[1] However, his lung disease continued to progress, and at 6 months his FVC was 67% predicted, TLC was 65%, and DLCO was 59%.

DISCUSSION

This case illustrates the value of obtaining surgical lung biopsy specimens from more than one lobe of the lung. Since this patient had areas resembling both nonspecific interstitial pneumonia (NSIP) and UIP on the biopsy specimen, NSIP may have been diagnosed if a single biopsy specimen had shown NSIP. When areas of NSIP and UIP are both present on a biopsy specimen, the clinical course appears to be more typical of UIP than NSIP. Patients with elements of both fibrotic NSIP and UIP have a worse prognosis compared to patients with fibrotic NSIP alone.[2] Furthermore, the importance of identifying areas of normal lung in the pathology of UIP is demonstrated.

The usefulness of a 6-min walk test goes beyond assessing diagnosis and requirements for supplemental oxygen with exertion. Exercise-induced hypoxia is also an index of the severity of interstitial lung

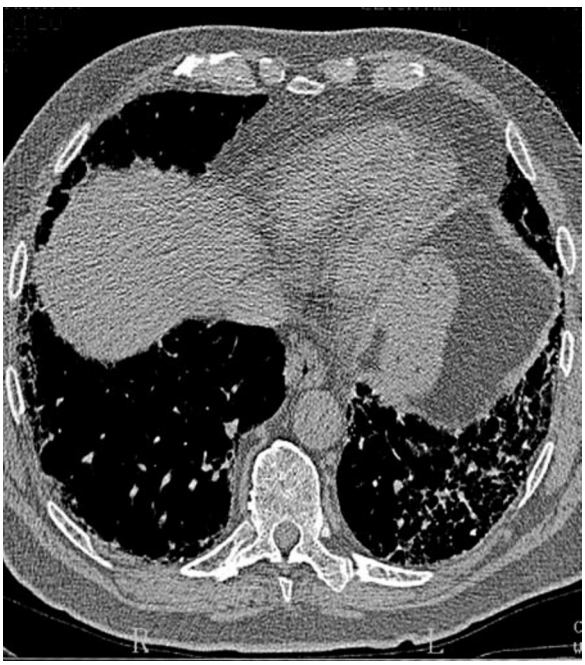

FIGURE 3. HRCT scan of lower lung zones reveals asymmetric basilar reticular opacities with some honeycomb changes.

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