CHEST

A 41-Year-Old Woman With Shortness of Breath and History of Rash and Recurrent Laryngeal Edema

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A 41-year-old Hispanic woman with a 20 pack-year smoking history presented with worsening shortness of breath on exertion that gradually started 2 years ago, then significantly deteriorated over the last 4 months. She was diagnosed with COPD 2 months prior to her presentation and started on treatment with fluticasone propionate and albuterol. Her medical history was relevant for undifferentiated connective tissue disorder diagnosed 5 years prior due to a positive antinuclear antibody test, arthralgia, recurrent urticarial skin rash, peripheral neuropathy, abdominal pain, and diffuse body swelling. She was started on treatment with prednisone and azathioprine at the time and had substantial improvement in the occurrence of her urticaria. She also had a history of recurrent laryngeal edema of unclear etiology. She had no history of IV drug abuse, no exposure to animals, was not sexually active, and had no recent travel outside of Florida. There was no significant family history of lung diseases.

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Physical Examination Findings

On presentation, the patient was afebrile with a heart rate of 78 beats/min, BP of 132/92 mm Hg, and respiratory rate of 20 breaths/min and saturating 93% on 3 L oxygen. Her BMI was 22.6 kg/m^2. Her physical examination was remarkable for decreased air entry bilaterally and expiratory wheezing in all lung fields. There was no evidence of clubbing, her heart rate was regular with no added sounds, and there was no evidence of any active skin lesions.

Diagnostic Studies

Her CBC count and complete metabolic panel findings were normal except for a mild leukocytosis level of 14,000/mL. Her chest radiograph (Fig 1) and CT chest scan (Fig 2) are shown. Her erythrocyte sedimentation rate, C-reactive protein level, and autoimmune panel (antinuclear antibody [ANA], antineutrophil cytoplasmic antibody, double-stranded DNA [dsDNA], antitopoisomerase, Sjögren antibodies, rheumatoid factor, cyclic citrullinated peptide, smooth muscle,

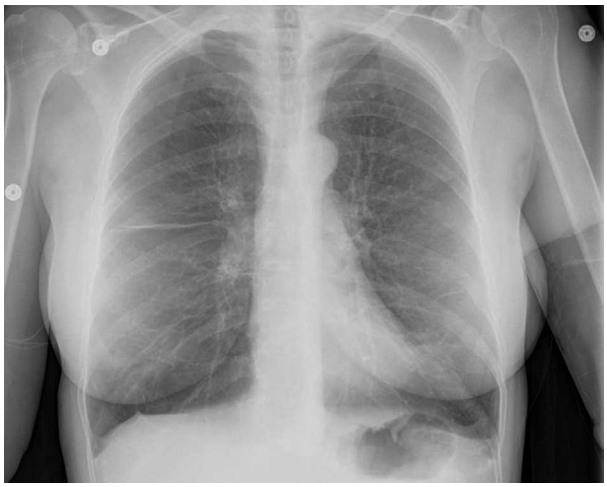

Figure 1 – *Lung volumes are hyperexpanded with attenuation of vascular markings in both lungs. There is evidence of pleural thickening or scar in the right minor fissure.*

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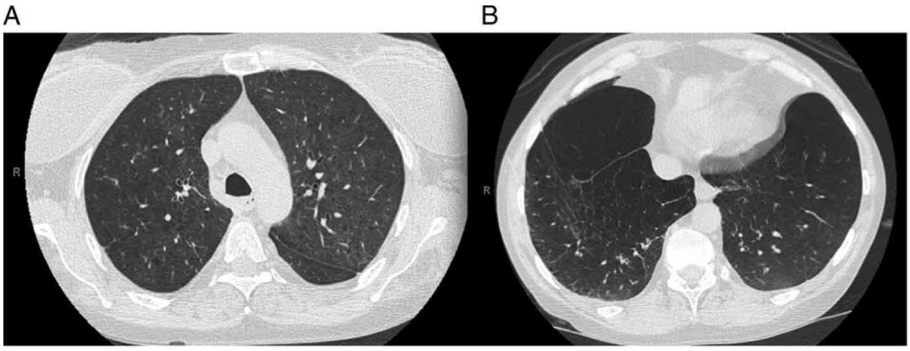

Figure 2 – *A, B, Hyperexpanded lungs with diffuse centrilobular emphysematous changes at the apices (A) that are more predominant and extensive at the bases (B).*

ribonucleoprotein, and histidyl tRNA synthetase) results were all normal. She tested negative for hepatitis and HIV. C1 esterase-inhibitor level was normal. Her α_1-antitrypsin genotype was normal. Her complement levels were low, with a C3 level of 42 mg/dL (normal, 90-180 mg/dL), C4 level of 9 mg/dL (normal, 15-46 mg/dL), and C1q level $<$ 3.6 mg/dL (normal, 5.0-8.6 mg/dL). Pulmonary function tests (PFTs) showed a very severe obstructive pattern (FEV_1, 29%) with significant reduction in diffusion capacity of lung for carbon monoxide (6% corrected for hemoglobin). Echocardiogram findings were unremarkable except for mildly elevated right ventricular systolic pressure (35-40 mm Hg).

What is the diagnosis?