CHEST

A 28-Year-Old Man Presenting With Intractable Dry Cough and a History of Ulcerative Colitis

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A 28-year-old man of Japanese descent presented to the ED with a 2-month history of dry cough, shortness of breath, and weakness. He did not complain of fever, chest pain, or abdominal symptoms, and had no history of smoking. The patient's medical history was significant for an episode of ulcerative colitis 6 years previously after presenting with bloody diarrhea, stomach pain, fever, weight loss, and bilateral episcleritis. He had been treated consecutively with mesalazine, azathioprine, infliximab, golimumab, and adalimumab. Concomitant respiratory symptoms had been present during 2 flare-ups of severe ulcerative colitis disease activity and were successfully treated with a course of oral prednisone.

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Physical Examination Findings

The patient was afebrile with stable vital signs, a heart rate of 75 beats/min, a respiratory rate of 15 breaths/min, and an oxygen saturation level of 98% on room air. His humeral blood pressure was 122/70 mm Hg on the left arm and 112/65 mm Hg on the right arm. There was no cyanosis, clubbing, or edema in his extremities. Proximal and distal pulses were not diminished, and no bruits were heard. Heart sounds were normal without murmur or rubs. Chest auscultation and abdominal status were unremarkable.

Diagnostic Studies

The patient's hemoglobin level was 13.8 g/dL, WBC count was 8,600/μL, and platelet count was 388,000/μL. His sodium level was 138 mM, the potassium level was 3.8 mM, and the creatinine level was 0.73 mg/d. C-reactive protein levels were increased to 88 mg/L, and the brain natriuretic peptide level was 5 ng/L. Total bilirubin was 0.71 mg/dL, aspartate aminotransferase was 25 U/L, alanine aminotransferase was 38 U/L, alkaline phosphatase was 135 U/L, and gamma-glutamyl transpeptidase was 132 U/L. Results of serologic testing for antineutrophil cytoplasmic antigen were negative. The ECG tracing was normal, and his chest radiograph showed clear lung fields and no pleural effusion. Results of complete pulmonary function testing demonstrated a restrictive ventilatory defect, with a total lung capacity of 71% of predicted and normal diffusing capacity of the lung for carbon monoxide.

Chest Imaging

The patient underwent an initial unenhanced chest CT scan to exclude an airway or parenchymal complication of ulcerative colitis. Only nonspecific perivascular and peribronchial thickening of the upper lobes, including nonspecific left hilar enlargement, was observed, indicating a suspicion of pulmonary artery disease.

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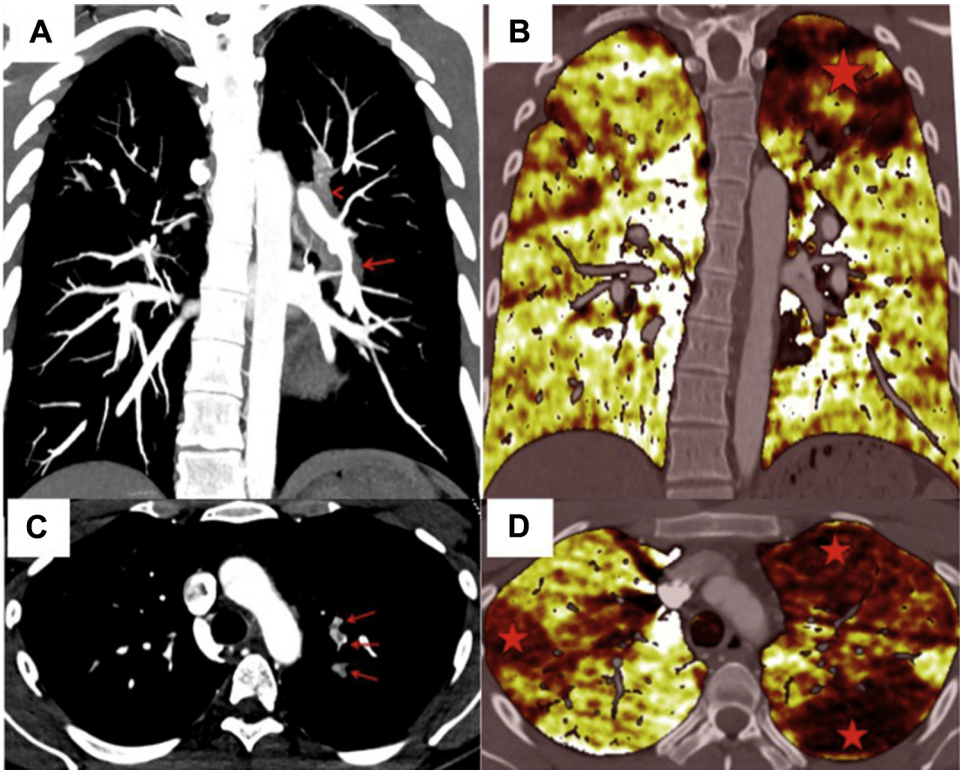

Figure 1 – *A-D, Dual-energy CT scans revealed thickening of the pulmonary arteries as well as segmental pulmonary artery stenosis and occlusions (arrow and arrowheads; A and C). Iodine maps showing segmental hypoperfusion of both upper lobes (stars; B and D).*

Supplemental imaging was thus required to precisely assess the pulmonary arteries, and an enhanced dual-energy CT (DECT) scan of the chest was performed (Somatom Definition Flash, Siemens Healthcare). Parenchymal and mediastinal reconstructions were performed for the morphologic analysis. Perfusion iodine maps were then generated (Syngo.via Multimodality Workplace; Siemens Medical Solutions). The mean dose-length product was 252 mGy × cm, corresponding to an effective dose of approximately 3.5 mSv, which is largely below the European recommended dose. Initial morphologic analysis demonstrated concentric thickening of the pulmonary arteries extending to the lobar and segmental arteries, as well as segmental pulmonary artery stenosis and occlusions (Figs 1A, C). There were no aneurysms or pulmonary infarcts. An additional combined functional evaluation revealed segmental hypoperfusion of both upper lobes, resulting in peripheral vascular defects (Figs 1B, 1D).

What is the diagnosis?