WHAT COULD BE THREATENING HIS LUNG FUNCTION?

ABOUT BEN

- 64 years old
- Truck driver
- Referred to pulmonologist by GPfor pulmonary evaluation

MEDICAL HISTORY

- Presenting symptoms
  - — Worsening dyspnea
  - — Chronic cough
- Smoker (10 pack-years)
- Previously diagnosed with COPD, but unresponsive to therapy
- Cardiac evaluation revealed no cardiac involvement

PHYSICAL EXAM

- Fine Velcro®-like crackles on lung auscultation
- Evidence of finger clubbing

Not an actual patient.

COPD, chronic obstructive pulmonary disease; GP, general practitioner.

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PRESENTING CLINICAL PICTURE RAISES SUSPICION OF IPF

Ben's pulmonologist rules out other known causes for ILD, including:

- Environmental exposures
- Drug toxicity
- Connective tissue disease

Pulmonary function monitoring shows a restrictive pattern inconsistent with COPD:

PFTs	
FVC*	73%
FEV_1*	85%
FEV_1/FVC	0.86
TLC*	68%
DL_{CO}*	66%

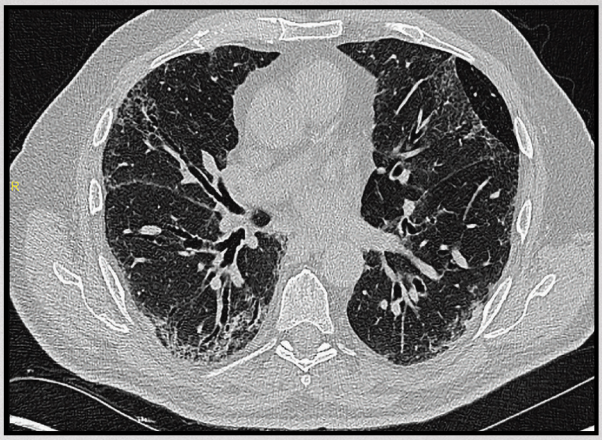
*% predicted.

DL_{CO}, diffusing capacity of the lungs for carbon monoxide; FEV_1, forced expiratory volume in 1 second; FVC, forced vital capacity; HRCT, high-resolution computed tomography; ILD, interstitial lung disease; IPF, idiopathic pulmonary fibrosis; MDT, multidisciplinary team; PFT, pulmonary function test; TLC, total lung capacity; UIP, usual interstitial pneumonia.

IN THE ABSENCE OF OTHER KNOWN CAUSES, HRCT, IN DISCUSSION WITH AN MDT, IS SUFFICIENT TO CONFIRM AN IPF DIAGNOSIS[1,2]

RADIOLOGIC EVIDENCE CONFIRMS SUSPICION OF PULMONARY FIBROSIS

Ben's pulmonologist detected abnormalities on HRCT scan



- Subpleural reticulation
- Traction bronchiectasis
- No definite evidence of honeycombing
- **Further investigation required to confirm UIP pattern**

Subsequent MDT decision to obtain a surgical lung biopsy enabled confirmation of UIP

Diagnosis: IPF

IDENTIFY PULMONARY FIBROSIS IN PATIENTS AS EARLY AS POSSIBLE[2,3]

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SUSPECT PULMONARY FIBROSIS

PULMONARY FIBROSIS IS A COMMON THREAT ACROSS A WIDE RANGE OF ILDs, INCLUDING:[4-7]

- **Idiopathic pulmonary fibrosis**
- Systemic sclerosis-associated ILD
- Rheumatoid arthritis-associated ILD
- Other connective tissue disease-associated ILD
- Hypersensitivity pneumonitis
- Exposure-related ILDs
- Idiopathic non-specific interstitial pneumonia
- Unclassifiable idiopathic interstitial pneumonia
- Sarcoidosis

WHILE ALL PATIENTS WITH IPF HAVE PROGRESSIVE PULMONARY FIBROSIS, THERE EXISTS A VARIETY OF OTHER ILDS THAT MAY ALSO DEVELOP A PROGRESSIVE FIBROSING PHENOTYPE[4,8]

Lung function decline[4]

Worsening quality of life[4]

Worsening respiratory symptoms[4]

2 to 5 years median survival[2,3]

Acute IPF exacerbations can occur at any time and cause sudden and severe lung function worsening[2,6]

References: 1. Raghu G, Remy-Jardin M, Myers JL, et al; on behalf of the American Thoracic Society, European Respiratory Society, Japanese Respiratory Society, and Latin American Thoracic Society. Diagnosis of Idiopathic Pulmonary Fibrosis. An Official ATS/ERS/JRS/ALAT Clinical Practice Guideline. *Am J Respir Crit Care Med*. 2018;198(5):e44-e68. **2.** Raghu G, Collard HR, Egan JJ, et al; on behalf of the ATS/ERS/JRS/ALAT Committee on Idiopathic Pulmonary Fibrosis. An official ATS/ERS/JRS/ALAT statement: idiopathic pulmonary fibrosis: evidence-based guidelines for diagnosis and management. *Am J Respir Crit Care Med*. 2011;183(6):788-824. **3.** Molina-Molina M, Aburto M, Acosta O, et al. Importance of early diagnosis and treatment in idiopathic pulmonary fibrosis. *Expert Rev Respir Med*. 2018;12(7):537-539. **4.** Cottin V, Hirani NA, Hotchkin DL, et al. Presentation, diagnosis and clinical course of the spectrum of progressive-fibrosing interstitial lung diseases. *Eur Respir Rev*. 2018;27(150):pii:180076. **5.** Demedts M, Wells AU, Antó JM, et al. Interstitial lung diseases: an epidemiological overview. *Eur Respir J Suppl*. 2001;32:2s-16s. **6.** Ley B, Collard HR, King TE Jr. Clinical course and prediction of survival in idiopathic pulmonary fibrosis. *Am J Respir Crit Care Med*. 2011;183(4):431-440. **7.** Wells AU, Brown KK, Flaherty KR, et al. What's in a name? That which we call IPF, by any other name would act the same. *Eur Respir J*. 2018;51(5):1800692. **8.** Flaherty KR, Brown KK, Wells AU, et al. Design of the PF-ILD trial: a double-blind, randomised, placebo-controlled phase III trial of nintedanib in patients with progressive fibrosing interstitial lung disease. *BMJ Open Resp Res*. 2017;4(1):e000212.

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