WHAT COULD BE THREATENING HIS LUNG FUNCTION?

ABOUT ROBERT

- 68-year-old banker

INITIAL EVALUATION

- Worsening dyspnea
- Unexplained weight loss
- Inspiratory crackles on lung auscultation
- Visible mold in basement identified as potential exposure

INITIAL TESTING

- Broncho-alveolar lavage lymphocytes >50%
- PFTs reveal a restrictive pattern
- Positive HP panels detected precipitating antibodies against mold

HP, hypersensitivity pneumonitis;
PFTs, pulmonary function tests.

Not an actual patient.

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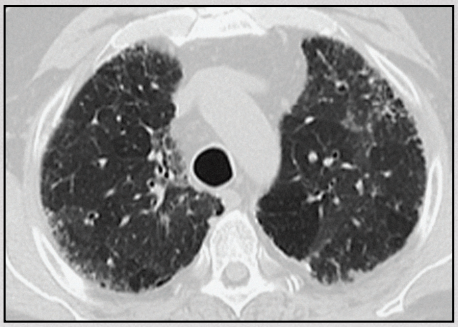
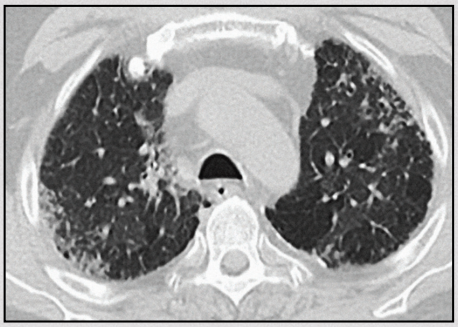
PULMONARY FIBROSIS IS CONFIRMED WITH HRCT

Robert's pulmonologist detects air trapping on HRCT scan



Inspiratory



Expiratory

- Areas of mosaic lung attenuation on inspiratory image are confirmed to be air trapping on expiratory images
- Peripheral reticulation; no honeycombing
- Unconfirmed ground glass opacity
- Upper and lower lobe involvement

Diagnosis: Chronic hypersensitivity pneumonitis (cHP)

TYPICAL MANAGEMENT PLAN FOR cHP INCLUDES AVOIDANCE OR REMOVAL OF EXPOSURE AND IMMUNOSUPPRESANTS[1]

WORSENING RESPIRATORY SYMPTOMS NECESSITATES FURTHER EVALUATION

At a 3-month follow up, Robert's pulmonologist evaluates for disease progression

- Dyspnea continues to worsen despite removal of inciting exposure and treatment with prednisone for 3 months
- PFTs have declined since diagnosis

PFTs	Baseline	3 months
FVC*	75%	71%
FEV_1*	73%	67%
FEV_1/FVC ratio	0.72	0.71
TLC*	72%	69%
DL_{CO}*	46%	43%

*% predicted.

DL_{CO}, diffusing capacity for carbon monoxide; FEV_1, forced expiratory volume in 1 second; FVC, forced vital capacity; HRCT, high-resolution computed tomography; TLC, total lung capacity.

WORSENING RESPIRATORY SYMPTOMS MAY INDICATE PROGRESSIVE DISEASE[2]

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SUSPECT PULMONARY FIBROSIS

PULMONARY FIBROSIS IS A COMMON THREAT ACROSS A WIDE RANGE OF ILDs, INCLUDING:[2-5]

- 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

cHP CAN BE CAUSED BY MANY DIFFERENT ENVIRONMENTAL ANTIGENS, MAKING A THOROUGH MEDICAL HISTORY CRUCIAL FOR PROMPT IDENTIFICATION[6]

Patients with cHP can experience ≥10% decrease in predicted FVC over 6 to 12 months[6]

4 to 7 years median survival of patients with cHP[1,6]

21% of patients with cHP are at risk of developing a progressive fibrosing phenotype and may have worse outcomes[7]*

cHP, chronic hypersensitivity pneumonitis; ILD, interstitial lung disease.
*According to an online survey of physicians.[7]

References: 1. Vourlekis JS, Schwarz MI, Cherniack RM, et al. The effect of pulmonary fibrosis on survival in patients with hypersensitivity pneumonitis. *Am J Med*. 2004;116(10): 662-668. **2.** 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. **3.** Demedts M, Wells AU, Antó JM, et al. Interstitial lung diseases: an epidemiological overview. *Eur Respir J Suppl*. 2001;32:2s-16s. **4.** 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. **5.** 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. **6.** Gimenez A, Storrer K, Kuranishi L, et al. Change in FVC and survival in chronic fibrotic hypersensitivity pneumonitis. *Thorax*. 2018;73(4):391-392. **7.** Wijsenbeek MS, Kreuter M, Fischer A, et al. Non-IPF Progressive Fibrosing Interstitial Lung Disease (PF-ILD): The Patient Journey. *Am J Respir Crit Care Med*. 2018;197:A1678.

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