WHEN PULMONARY FIBROSIS IS SUSPECTED,

WHAT'S NEXT FOR DAVID?

MEDICAL HISTORY:

- 52-year-old Caucasian male[1-3]
- Former smoker, 30 pack-years[1,3]
- Presented with dyspnea and fatigue in hospital; consolidation found on chest X-ray[3]
- Treated for pneumonia with broad-spectrum antibiotics[3]
  - —Cultures consistently negative

INITIAL CLINICAL EVALUATION:

- Exertional dyspnea, mild fatigue[3]
- Dry inspiratory crackles at the lung bases[3]
- Reduced PFTs with restriction[1]

69% FVC % predicted

48% DL_{co} % predicted

- Rheumatologic exam was negative[3]
- Rheumatoid factor and anticitrullinated C-peptide positive, slightly increased creatinine kinase and CRP levels[3]

Treated with steroids and an alkylating agent and showed some symptomatic improvement[3]

1-YEAR FOLLOW-UP:

- Respiratory symptoms have worsened[3]
- Underlying etiology unclear[3]
- Stable PFTs[4]
- Extensive fibrosis on HRCT[3]
  - —Ground glass opacities, traction bronchiectasis, mosaic attenuation, and reticulation

After review of clinical, radiographic, and histologic findings by an MDT did not produce a conclusive diagnosis, ILD was considered unclassifiable[3]

CRP, C-reactive protein; DL_{co}, diffusing capacity of the lungs for carbon monoxide; FVC, forced vital capacity; HRCT, high-resolution computed tomography; ILD, interstitial lung disease; MDT, multidisciplinary team; PFTs, pulmonary function tests.

FVC is not declining, but symptoms and fibrosis are worsening, which are indicators of progressive disease.[4]

WHAT IS THE NEXT STEP IN DAVID'S DISEASE MANAGEMENT PLAN?

Not an actual patient.

HRCT scan courtesy of Leung SC et al. *Respirol Case Rep*. 2015;3(3):85-88. Published by John Wiley & Sons Ltd.

~1 IN 4 PATIENTS WITH ILD MAY DEVELOP A PROGRESSIVE PHENOTYPE[5]*

THE INSIDIOUS THREAT OF PULMONARY FIBROSIS CROSSES DIVERSE ILDs[6-8]

- Idiopathic pulmonary fibrosis (IPF)
- Systemic sclerosis-associated ILD
- Rheumatoid arthritis-associated ILD
- Other connective tissue disease-associated ILDs
- Hypersensitivity pneumonitis
- Occupational exposure-related ILDs
- Idiopathic nonspecific interstitial pneumonia
- Unclassifiable ILD
- Sarcoidosis

SIMILAR TO IPF, SOME ILDs CAN DEVELOP A PROGRESSIVE FIBROSING PHENOTYPE CHARACTERIZED BY[7,8]:

- Worsening respiratory symptoms
- Accelerated decline in lung function
- Worsening quality of life
- Early mortality

EARLY IDENTIFICATION OF ILD IS CRITICAL

Observe for respiratory symptoms[9]:

- Cough
- Dyspnea

Listen for[10,11]:

- Dry inspiratory crackles, typically at the lung bases

Order baseline PFTs and monitor regularly[9]:

- Restrictive PFT
- Reduced DL_{co}

Order HRCT if ILD is suspected[9,12]:

- Presence of fibrotic ILD

EARLY IDENTIFICATION OF PROGRESSIVE PULMONARY FIBROSIS CAN HELP ENSURE PATIENTS RECEIVE APPROPRIATE INTERVENTION[7]

*Data from a global, online survey of physicians (N=486).[5]

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