WHEN PULMONARY FIBROSIS IS SUSPECTED,

WHAT'S NEXT FOR SANDRA?

MEDICAL HISTORY:

- 64-year-old African American female[1]
- Presented with dyspnea on exertion, worsening cough, and fatigue[2-4]
- PFTs revealed restriction with reduced FVC and DL_{co}[2,3]
- Presence of bilateral hilar lymphadenopathy with reticulation seen on HRCT and sarcoidosis confirmed by lung biopsy
- Sarcoidosis diagnosis confirmed[3,4]
- Medications: corticosteroid and immunosuppressant to inhibit inflammatory pathways[1,2]

FOLLOW-UP AT 1 YEAR:

- Respiratory symptoms continue to worsen[3]
- Serial PFTs reveal decline from diagnosis[3]

6% decrease in FVC % predicted[3]
4% decrease in DL_{co} % predicted[3]

Worsening symptoms and lung function indicate progressive pulmonary fibrosis[5]

FOLLOW-UP RADIOLOGIC FINDINGS:

- Fibrotic changes consistent with advanced stage sarcoidosis[2]:
  —Honeycombing —Reticulation —Traction bronchiectasis —Ground glass opacity

Patients with sarcoidosis who have progressive pulmonary fibrosis face a worse prognosis[2,6]

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

Despite treatment with a corticosteroid and an immunosuppressant, lung function has declined and fibrotic ILD is detected.

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

Not an actual patient.

HRCT scan courtesy of and used with permission from Steven D. Nathan, MD.

~1 IN 4 PATIENTS WITH ILD MAY DEVELOP A PROGRESSIVE PHENOTYPE[7]*

THE INSIDIOUS THREAT OF PULMONARY FIBROSIS CROSSES DIVERSE ILDs[6,8,9]

- 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[6,9]:

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

EARLY IDENTIFICATION OF ILD IS CRITICAL

Observe for respiratory symptoms[10]:

- Cough
- Dyspnea

Listen for[11,12]:

- Dry inspiratory crackles, typically at the lung bases

Order baseline PFTs and monitor regularly[10]:

- Restrictive PFT
- Reduced DL_{co}

Order HRCT if ILD is suspected[10,13]:

- Presence of fibrotic ILD

EARLY IDENTIFICATION OF PROGRESSIVE PULMONARY FIBROSIS CAN HELP ENSURE PATIENTS RECEIVE APPROPRIATE INTERVENTION[6]

*Data from a global, online survey of physicians (N=486).[7]

IPF, idiopathic pulmonary fibrosis.

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