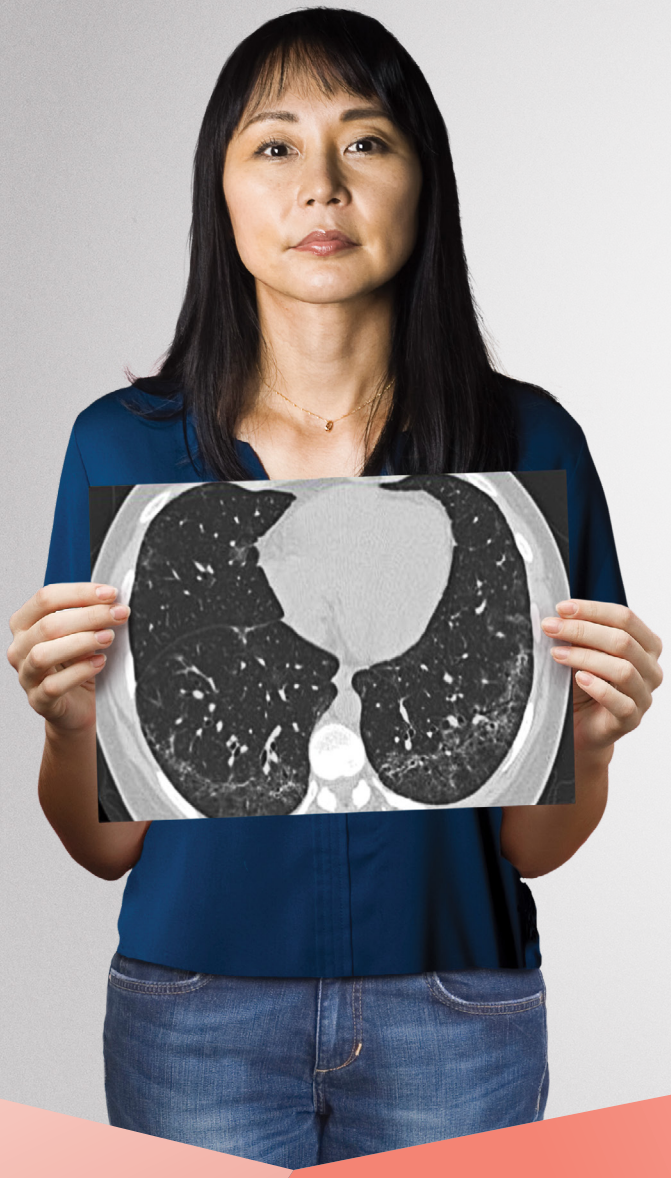
WHEN PULMONARY FIBROSIS IS SUSPECTED,

WHAT'S NEXT FOR ANN?



MEDICAL HISTORY:

- 40-year-old Asian woman[1-3]
- Diagnosed with mixed connective tissue disease 15 years ago[1,4]
- Current smoker[3]
- History of tachycardia[1]
- Current treatments: beta blocker, NSAID, immunosuppressant[5]

CLINICAL EVALUATION:

- Dyspnea[1]
- Dry inspiratory crackles at the lung bases[1]
- Reduced PFTs with restriction[1]

RADIOLOGIC FINDINGS:

- Fibrotic ILD confirmed by features consistent with an NSIP pattern[1]:
  - —Subpleural sparing
  - —Bilateral ground glass opacity with reticulation

Diagnosis: mixed connective tissue disease-associated ILD[1]

MONITORING:

- Lung function declined over 6 months[3]
- Respiratory symptoms worsening[3]

8% decrease in FVC % predicted[3]
2% decrease in DL_{co} % predicted[3]

Declining lung function and worsening symptoms are signs of progressive disease[3]

DL_{co}, diffusing capacity of the lungs for carbon monoxide; FVC, forced vital capacity; HRCT, high-resolution computed tomography; ILD, interstitial lung disease; MCTD, mixed connective tissue disease; NSAID, nonsteroidal anti-inflammatory drug; NSIP, nonspecific interstitial pneumonia; PFT, pulmonary function test.

Fibrotic ILD is detected and symptoms and lung function are worsening despite immunomodulatory therapy.[5]

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

Not an actual patient.

HRCT scan courtesy of and used with permission from Robert Suh, MD.

~1 IN 4 PATIENTS WITH ILD MAY DEVELOP A PROGRESSIVE PHENOTYPE[6*]

THE INSIDIOUS THREAT OF PULMONARY FIBROSIS CROSSES DIVERSE ILDs[7-9]

- 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[8,9]:


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[8]

*Data from a global, online survey of physicians (N=486).[6]

HRCT, high-resolution computed tomography; IPF, idiopathic pulmonary fibrosis.

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