SC-VN-02147

WHAT COULD BE THREATENING HER LUNG FUNCTION?

ABOUT STEPHANIE

- 43 years old
- Accountant and single mother
- Diagnosed with limited cutaneous systemic sclerosis (lcSSc) 3 years ago
- At diagnosis she had no respiratory symptoms and a baseline chest HRCT did not show evidence of ILD. Furthermore, PFTs did not show evidence of impairment
- Has recently noticed that she is becoming short of breath during regular activities

DIAGNOSTIC HISTORY

- Presenting symptoms leading to her original lcSSc diagnosis:
  - A long history of Raynaud's phenomenon
  - Skin thickening on fingers and face
  - Calcinosis
  - Digital ulceration
  - Puffy fingers
- Anti-nuclear antibody positive
- Organ involvement: esophageal dysfunction
- Current medications: methotrexate, proton pump inhibitor

HRCT, high-resolution computed tomography;
PFT, pulmonary function test;
ILD, interstitial lung disease.

Not an actual patient.

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SCREEN EARLY AND REGULARLY TO DETECT SSc-ILD FROM ITS OUTSET[1-4]

3 years after her lcSSc diagnosis, Stephanie presents with the following signs and symptoms:

- Dry cough
- Dyspnea on exertion (apparent over the last 5 months)
- Mild inspiratory bibasilar fine crackles on auscultation

Pulmonary function testing shows decreased FVC and DL_{CO}:

PFTs	
FVC*	82%
FEV_1*	81%
FEV_1/FVC	0.82
TLC*	81%
DL_{CO}*	65%

A new HRCT scan was performed

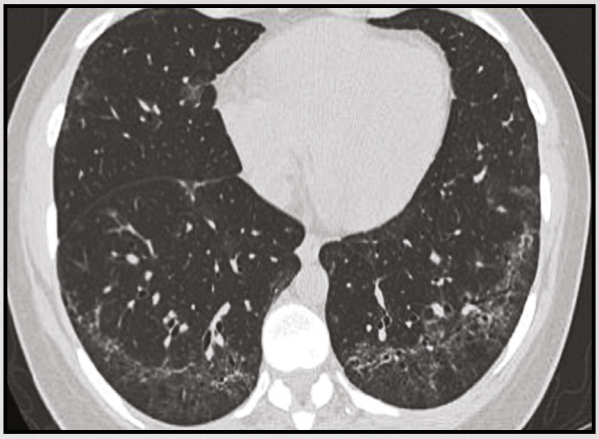
RESPIRATORY SYMPTOMS AND RESTRICTIVE PFTs IN PATIENTS WITH SSc CAN INDICATE THE PRESENCE OF PULMONARY FIBROSIS, THAT SHOULD BE CONFIRMED BY HRCT[1]

*% 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; NSIP, non-specific interstitial pneumonia; SSc, systemic sclerosis; PFT, pulmonary function test; SSc-ILD, systemic sclerosis-associated interstitial lung disease; TLC, total lung capacity.

RADIOLOGIC EVIDENCE CONFIRMED SUSPICION OF PULMONARY FIBROSIS

Abnormalities were detected on Stephanie's latest HRCT scan



- Subpleural sparing
- Bilateral ground glass opacity with reticulation
- **These features are consistent with a non-specific interstitial pneumonia (NSIP) HRCT pattern. This pattern is common in patients with SSc-ILD.[1,4]**

Diagnosis: SSc-ILD

SCREENING WITH HRCT FOR THE PRESENCE OF ILD IS RECOMMENDED AT BASELINE FOR ALL PATIENTS WITH A DIAGNOSIS OF SSc[2,4,5]

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SUSPECT PULMONARY FIBROSIS

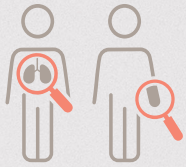
PULMONARY FIBROSIS IS A COMMON THREAT ACROSS A WIDE RANGE OF ILDs, INCLUDING:[6-9]

- 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

IN SSc, ILD IS A COMMON, EARLY, AND POTENTIALLY FATAL MANIFESTATION[10-12]

53% of those with
DIFFUSE CUTANEOUS SSc
(n=1349)

35% of patients with
LIMITED CUTANEOUS SSc
(n=2101)



Patients are at the highest risk of ILD within the first 3 years from SSc onset[11]

35%

of SSc-related deaths are due to ILD[12]

ILD, interstitial lung disease; SSc, systemic sclerosis.

References: 1. Silver KC, Silver RM. Management of Systemic-Sclerosis-Associated Interstitial Lung Disease. *Rheum Dis Clin North Am*. 2015;41(3):439-457. **2.** Cottin V, Brown KK. Interstitial lung disease associated with systemic sclerosis (SSc-ILD). *Respir Res*. 2019;20(1):13. **3.** Roofeh D, Jaafar S, Vummidi D, Khanna D. Management of systemic sclerosis associated interstitial lung disease. *Curr Opin Rheumatol*. 2019;31(3):241-249. **4.** Chowaniec M et al. *Reumatologia*. 2018;56(4):249-254. **5.** Molberg O, Hoffmann-Vold A-M. Interstitial lung disease in systemic sclerosis: progress in screening and early diagnosis. *Curr Opin Rheumatol*. 2016;28(6):613–618. **6.** 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. **7.** Demedts M, Wells AU, Antó JM, et al. Interstitial lung diseases: an epidemiological overview. *Eur Respir J Suppl*. 2001;32:2s-16s. **8.** 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. **9.** 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. **10.** Walker UA, Tyndall A, Czirják L, et al. Clinical risk assessment of organ manifestations in systemic sclerosis: a report from the EULAR Scleroderma Trials And Research group database. *Ann Rheum Dis*. 2007;66(6):754-763. **11.** Steen V. Predictors of end stage lung disease in systemic sclerosis. *Ann Rheum Dis*. 2003;62(2):97-99. **12.** Tyndall AJ, Bannert B, Vonk M, et al. Causes and risk factors for death in systemic sclerosis: a study from the EULAR Scleroderma Trials and Research (EUSTAR) database. *Ann Rheum Dis*. 2010;69(10):1809-15.

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