
**Project ECHO for Interstitial Lung Disease
Case Form**

ECHO ID: ILDECHO-053

 New Case Follow-up Case

Current or suspected ILD diagnosis (if known):

How was this diagnosis made (select all that apply)?

- Radiology
- Pathology
- Multidisciplinary discussion

Main Question:

By presenting this case, I hope to obtain:

- Help making a diagnosis
- Help with lab/test interpretation
- Help with clinical management
- Other (please specify):

Patient Demographic Information:

Male Female Transgender Age (years): 73

State of primary residence:

Ethnicity: Hispanic or Latino Not Hispanic or Latino

Race (check all that apply): American Indian or Alaskan Native Asian

Black or African American Native Hawaiian/Pacific Islander White

Other Unknown

Case Summary:

Five years of dyspnea and cough, now with new hypoxia, chest CT scan showed ILD, thought to be IPF but she also has Raynaud's, severe esophageal disease and chest CT is not typical for UIP.

Exposure History:

- Previous or current bird ownership Down products in the home
- Woodworking Well water Hay exposure Hot tub/jacuzzi/sauna
- Humidifiers Dusty environments Water damage or mold at work or home
- Farmland/barns Amiodarone Chemotherapy past/present
- Chest radiation past/present Asbestos

Past Medical History:

Cervical disc disease; chronic GERD; dyslipidemia; past Hx of positive Tb Quant GOLD 2017; fibromyalgia; Hx of severe burns at age 10; Tb exposure at age 4, does not think she was ever treated

Medications:

Duloxetine, atorvastatin, famotidine, cyclosporine eye drops

Focused ROS:

- Rashes Skin thickening Arthralgias Myalgias Muscle weakness
- Dry mouth Dry eyes Red or painful eyes Raynaud's Oral ulcers
- Alopecia Dysphagia Heartburn/reflux Fevers Night sweats
- Palpitations Weight loss

Smoking History:

- Never smoked
 - Current smoker (packs per day)
 - Pack years: Quit date (if applicable):
 - Cocaine use (route)
 - Vaping/e-cigarettes (frequency)
 - Inhaled marijuana (quantity)
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Occupations, current and previous (if industrial or factory work, please provide specific details):

Tailor

Travel and Residential History:

No travel in over 2 years, lives alone, house with no mold or water damage.

Family History:

- Pulmonary fibrosis or interstitial lung disease
- RA, Lupus, or other autoimmune diseases
- Premature gray hair Cirrhosis of the liver Bone marrow disorders
- Leukemias

Comments:

Physical Exam:

Vital signs:

BP: 160/90 Height: 157 cm Wt: 83 kg BMI: 33.9

Oxygen saturation: 90% Ambulatory saturation: 84%

HEENT:

- Scleral injection Dry mucous membranes Poor dentition Ptosis

Pulmonary:

- Wheezes Rhonchi Crackles
- Squeaks Stridor Dullness to percussion Pleural Rub
- Bronchial breath sounds
- Other:

Cardiac:

- Murmur Gallop
- RV heave Pulmonary tap JVD Irregular
- Edema Other:

Abdomen:

- Distended
 Tender
 Tympanitic
 Pulsatile liver
 Fluid wave
 Other:

Skin/Nails:

- Rash (location, description)
 Clubbing
 Ragged cuticles
 Telangiectasias
 Abnormal nailfold capillaroscopy
 Digital swelling / Sclerodactyly
 Ulcerations
 Mechanics hands

Joints:

- Synovitis
 Deformity
 Tenderness
 Erythema

Neuro:

- Proximal muscle weakness
 Other:

Other pertinent findings:

Relevant Studies (please list key findings):

- PFTs:

	10/2023	06/2024	11/2024
FVC	1.86	1.74	1.38
FEV1	1.59	1.49	1.16
FEV1/FVC	0.86	0.86	0.84
DsbHb	14.5	14.6	12.4

TLC "reduced"

- CXR
 Uploaded to Ambra: Yes No
 CT chest
 Uploaded to Ambra: Yes No
 Echocardiogram
 Uploaded to Ambra: Yes No

Right heart catheterization:

Other relevant testing:

Echocardiogram: Normal LV, normal valves, normal RV size and function, SPAP 47, TAPSE 1.9, no pericardial effusion.

Lung Biopsy: Right upper, middle, and lower lobes, wedge biopsies. Chronic fibrosing interstitial pneumonia with a mixed pattern with superimposed chronic aspiration-related changes, favor connective tissue disease-related interstitial lung disease.

Relevant Labs:

Has the patient been evaluated by a Rheumatologist? Yes No

Date of last labs (month/year):

CBC/differential: Hb 13.2, WBC 8, Plts 374, Eos 0.4, normal differential otherwise

ANA: Neg Titer 1:1280 Pattern Speckled

ANA #2: Titer Pattern

ANA #3: Titer Pattern

Smith: Neg Titer

SSA/Ro60: Neg Titer

SSB/La: Neg Titer

dsDNA: Neg Titer

Scl-70: Neg Titer

Centromere: Neg Titer

RNA polymerase III: Neg Titer

RNP: Neg Titer

RF: Neg Titer

CCP: Neg Titer

ANCA: Neg Titer Pattern

MPO: Neg Titer

PR3: Neg Titer

PM1-Scl: Neg Titer Low positive

Ro52: Neg Titer

Jo-1: Neg Titer

EJ: Neg Titer

OJ: Neg Titer

PL-7: Neg Titer

PL-12: Neg Titer

MDA5: Neg Titer

KU: Neg Titer

MI-2: Neg Titer

P155/140 (TIF1y): Neg Titer

NXP-2: Neg Titer Low positive

SRP: Neg Titer

SAE-1: Neg Titer

HP panel: Neg Pos

C3/C4: WNL Low

ESR: 31

CRP: 0.3

CK: 83

Aldolase: 1.6

ACE: Not checked

Urinalysis: Normal

Additional Comments:

Questions:

- 1) Is this scleroderma ILD?
 - 2) How to decide treatment in "mixed" fibrosis: immune suppression or antifibrotic?
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