DIAGNOSIS AND LONGITUDINAL MANAGEMENT OF SYSTEMIC SCLEROSIS INTERSTITIAL LUNG DISEASE (SSc-ILD)

Pre-existing ILD with suspected CTD

Pre-existing SSc ILD not yet diagnosed

Dyspnea with suspected ILD

Obtain PFT's and HRCT with inspiratory, expiratory and prone imaging (Routine chest CT is inadequate).

Refer to Rheumatologist

Refer to Pulmonologist

Interdisciplinary Evaluation

DIAGNOSTIC EVALUATION

- HRCT
- PFT
- ECHO
- Serologies
- Assessment of Oxygen Needs
- Modified Rodnan Skin Score
- Swallowing/Aspiration Evaluation
- Esophageal Motility/GERD
- Age-appropriate Cancer Screening

ASSESS RISK FOR PROGRESSIVE ILD PHENOTYPE

Antibody status rather than the extent of scleroderma is most informative related to the risk for ILD.

R

Patients with nucleolar pattern ANA, anti-Th/To antibodies or anti-Scl-70 antibodies

I

Patients with SSc Phenotypes:
Limited Cutaneous SSc | Diffuse Cutaneous SSc
Scleroderma Sine Scleroderma | Overlap Syndrome

S

K

Patients with anti-centromere or anti-polymerase III antibodies are at lower risk for progressive ILD and greater risk for PHN (ACA) and renal crisis (RNAP)

CONSENSUS CLINICAL SUMMARY DIAGNOSIS = SSc-ILD

Initial Therapeutic Strategies

ILD Drug Therapy

- Cyclophosphamide (CYC)
- Mycophenolate Mofetil (MMF)
- Nintedanib
- Prednisone (low dose) - short term. Use caution in patients at risk for scleroderma renal crisis.
- Azathioprine and Rituximab are reasonable alternatives to CYC and MMF

Non-Pharmacologic Therapy

- Oxygen to maintain normoxia
- Pulmonary Rehab
- Vaccination
- Sleep with HOB elevated
- Avoid eating within 3 hours of lying down

Drug Therapy for Non-ILD manifestations of SSc
- Consult with appropriate specialist

Surveillance

Q 3-6 month assessment for therapeutic response and longitudinal management

- Spirometry
- DLCO
- 6MWT
- Annual Echo
- Annual HRCT
- Skin Exam
- Vascular Assessment
- Consider alternative therapies and organ transplantation for progressive unresponsive disease
- Consider age-appropriate cancer screening

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