

RHEUMATIC LUNG DISEASES -HRCT & REPORTING GUIDE

1. INTERSTITIAL LUNG DISEASES (ILD) & PATTERN DIFFERENTIAL DIAGNOSIS

The most common HRCT patterns in rheumatic diseases are NSIP (Nonspecific Interstitial Pneumonia) and UIP (Usual Interstitial Pneumonia); less frequently, OP (Organizing Pneumonia) and LIP are observed.

Rheumatoid Arthritis (RA)

1. Parenchymal Involvement (Interstitial Lung Disease - RA-ILD)

It is one of the most common causes of death in RA. HRCT pathologies:

UIP Pattern (40-60%): Most specific to RA and prognostically the most severe. Subpleural, lower lobe-predominant honeycombing, traction bronchiectasis.

NSIP Pattern (30-40%): Ground-glass opacities (GGO) and symmetric reticular increase in lower lobes; honeycombing is absent or minimal.

OP (Organizing Pneumonia) Pattern: Peribronchovascular or subpleural consolidations and reverse halo (atoll) sign.

AIP/DAD (Diffuse Alveolar Damage): Widespread ground-glass opacities and consolidation seen during acute exacerbations (high mortality).

2. Airway Diseases (Airway Involvement)

Airway involvement is observed in HRCT in up to 50-70% of RA patients (even if not clinically manifested):

Bronchiectasis: Cylindrical bronchiectasis especially in lower lobes (can develop as primary airway involvement, independent of fibrosis).

Follicular Bronchiolitis: Centrilobular small nodules (2-3 mm) and tree-in-bud appearance.

Constrictive (Obliterative) Bronchiolitis: Mosaic perfusion and air trapping accentuated on expiratory HRCT. It is an important cause of progressive dyspnea.

3. Pleural Involvement (Pleural RA)

Involvement seen most frequently at autopsy (70%), and clinically/radiologically at a rate of 20%:

Pleural Effusion: Usually unilateral, exudative in nature (characteristically glucose is very low).

Pleural Thickening and Plaques: Focal/diffuse pleural thickening at apices or basals secondary to chronic pleuritis.

Pneumothorax / Pyopneumothorax: Rarely can develop as a result of necrobiotic nodules opening into the pleura.

4. Parenchymal Nodules (Rheumatoid Nodules / Necrobiotic Nodules)

Nodules with smooth or regular margins, 0.5-5 cm in size, frequently located in subpleural areas or upper/middle lobes.

Cavitation: Nodules can undergo central necrosis forming thin/thick-walled cavities. Confused with infection or vasculitis.

Caplan Syndrome: Rapidly developing large necrotic nodules in the lungs in the presence of RA + Coal worker's pneumoconiosis.

5. Pulmonary Vascular and Other Involvements

Pulmonary Arterial Hypertension (PAH): Secondary to parenchymal fibrosis (Group 3) or rarely secondary to refractory vasculitis.

Rheumatoid Vasculitis: Pulmonary vasculitis and diffuse alveolar hemorrhage (DAH) are very rare but can be fatal.

Drug-Induced Lung Injury (Iatrogenic): Drug-induced pneumonitis/hypersensitivity and reactivation infections (TB, Opportunistic) developing secondary to Methotrexate (MTX), Leflunomide, TNF-alpha blockers, or Rituximab used in RA treatment.

QUICK "RA LUNG" SUMMARY CARD TO BE ADDED TO GUIDE (TRICK)

Trick / Golden Rule (RA Lung Spectrum):

1. **Male + Smoking + Fibrosis:** UIP pattern (honeycombing) is prominent.
2. **Ground-Glass + Mosaic Attenuation:** Consider bronchiolitis or drug-induced (Methotrexate) pneumonitis.
3. **Subpleural Cavitory Nodule:** Rheumatoid nodule (cavitating) / Caplan syndrome.
4. **Persistent Minimal Effusion + Thick Pleura:** RA Pleuritis.

Systemic Sclerosis (SSc / Scleroderma)

1. Parenchymal Involvement (SSc-ILD)

80-90% of lung parenchymal involvement in SSc is in the NSIP (Nonspecific Interstitial Pneumonia) pattern.

NSIP Pattern (Most Common):

- o Symmetric ground-glass opacities (GGO) and fine reticular thickenings in lower lobe basals and peripheral/subpleural areas.
- o **Subpleural Sparing:** Preservation of a 1-2 mm parenchymal strip immediately beneath the outermost pleural line is very characteristic for SSc-NSIP.
- o Accompanied by traction bronchiectasis/bronchiolectasis (indicator of fibrosis).

UIP Pattern (10-15%): Much less common compared to RA. Honeycombing cysts, if seen, are usually small in size.

- **OP (Organizing Pneumonia) & DAD:** Rare, can be seen in scleroderma crises or acute exacerbations.

2. Vascular Involvement and Pulmonary Hypertension (SSc-PAH)

SSc is the disease with the highest risk of Pulmonary Arterial Hypertension (PAH) among connective tissue diseases (develops in 10-15% of cases).

Isolated PAH (Group 1 PAH): Hypertension directly due to pulmonary vasculopathy developing without severe parenchymal fibrosis (especially common in limited cutaneous / CREST form).

Secondary PAH (Group 3 PAH): Develops secondary to severe fibrosis/hypoxia.

HRCT Findings:

- o Main Pulmonary Artery (PA) diameter > 29 mm (or PA / Ascending Aorta diameter ratio > 1.0).
- o Right ventricular hypertrophy/dilation, flattening (compression) of the interventricular septum toward the left ventricle.

3. Esophageal and Aspiration/Inspiration Findings ("Esophago-Lung" Relationship)

Esophageal smooth muscle atrophy and dilation are seen in more than 80% of SSc patients. When looking at the lung parenchyma on HRCT, examining the esophagus is of vital importance:

Esophageal Dilation (Wide-open Esophagus): Lumen widening, fluid/air levels in the middle and lower thoracic esophagus.

Aspiration Pneumonia and Bronchiolitis: Chronic microaspirations secondary to esophageal reflux and dysmotility; lead to patchy consolidations, centrilobular nodules, and worsening bronchiectasis in lower lobes.

4. Pleural and Mediastinal Involvement

Pleural Effusion / Thickening: Compared to RA and SLE, prominent pleural effusion is relatively rare in SSc. If seen, pleural thickening or mild effusion is observed.

Mediastinal Lymphadenopathy: Reactive mediastinal/hilar LAP can be seen in up to 60% of cases secondary to active fibrosis or aspiration (generally non-calcified lymph nodes not exceeding 1-1.5 cm).

5. Other / Extra-Pulmonary and Iatrogenic Findings

Chest Wall Soft Tissue Calcinosis (Calcinosis Cutis): Punctate/coarse calcinations in chest wall soft tissues or around shoulders on HRCT cuts (the 'C' of CREST syndrome).

Malignancy Risk: Increased risk of scirrhous/primary lung cancer (especially adenocarcinoma) on the background of chronic fibrosis.

Drug/Immunosuppressive Side Effects: Opportunistic infections (Pneumocystis jirovecii - PCP, CMV, Fungal) in those receiving antirheumatic/antifibrotic treatment such as Cyclophosphamide or nintedanib/pirfenidone.

QUICK "SSc LUNG" SUMMARY CARD TO BE ADDED TO GUIDE (TRICK)

Trick / Golden Rule (SSc Lung Spectrum):

1. **NSIP Pattern + Subpleural Sparing:** Symmetric ground-glass and traction bronchiectasis in lower lobes.
2. **Dilated Esophagus (Atonic Lumen):** When you see an air/fluid-filled esophagus appearance, the diagnosis behind it is 90% Systemic Sclerosis.
3. **Pulmonary Artery Enlargement Without Fibrosis:** Primary PAH development should be considered on the background of CREST / SSc.
4. **Skin/Soft Tissue Calcinosis:** Calcifications seen in the chest wall reinforce the diagnosis.

IDIOPATHIC INFLAMMATORY MYOPATHIES (Polymyositis / Dermatomyositis / Anti-Synthetase Syndrome)

In inflammatory myopathies, lung involvement is one of the most important determinants of morbidity and mortality. Involvement can be rapid-course (acute/subacute) or chronic.

1. Parenchymal Involvement (Myopathy-ILD)

Parenchymal involvement in inflammatory myopathies frequently presents in the form of overlap patterns:

NSIP + OP Overlap Pattern (50-60%): Most characteristic HRCT appearance for myopathies. Subpleural and peribronchovascular ground-glass opacities (NSIP) are observed simultaneously with areas of consolidation (OP).

NSIP Pattern (20-30%): Symmetric ground-glass opacities, reticular increases, and traction bronchiectasis/bronchiolectasis in lower lobes; honeycombing appearance is rare or minimal.

AIP/DAD (Diffuse Alveolar Damage / RP-ILD): Courses as Rapidly Progressive Interstitial Lung Disease. Characterized by bilateral widespread ground-glass opacities, geographic alveolar consolidations, and rapidly developing fibrotic bands (mortality is extremely high).

Pure OP Pattern: Subpleural or peribronchovascular consolidations, reverse halo (atoll) sign, and parenchymal mass-like appearance.

2. Respiratory Muscles and Diaphragm Involvement (Extra-Parenchymal)

Respiratory mechanics are significantly affected due to striated muscle inflammation and weakness:

Diaphragmatic Weakness and Elevation: Bilateral or unilateral elevation of diaphragmatic domes.

Lung Volume Loss: Significant volume loss and platelike (discoid) atelectasis in both lung bases secondary to restricted diaphragmatic movement.

Chest Wall Muscle Atrophy: Decreased volume and signs of fatty regeneration/atrophy in paravertebral, intercostal, and shoulder girdle muscle groups on HRCT cuts.

3. Aspiration Complications (Swallowing Dysfunction)

Dysphagia develops secondary to involvement of oropharyngeal and upper esophageal striated muscles:

Aspiration Pneumonia / Bronchiolitis: Patchy alveolar consolidations, centrilobular tree-in-bud nodules, and worsening of established bronchiectasis in lower lobe basals and posterior segments.

4. Specific Involvement Spectrums According to Serological Subtypes

Anti-Synthetase Syndrome (Anti-Jo-1, etc.): Myositis + ILD + "Mechanic's hands" + Raynaud phenomenon + Arthritis clinical picture. Prominent basal volume loss, prominent traction bronchiectasis, and NSIP/OP overlap appearance are paramount on HRCT.

Anti-MDA5 Positive Dermatomyositis: Fulminant RP-ILD (DAD) picture developing within weeks while muscle involvement is mild or absent (amyopathic/hypomyopathic dermatomyositis). Subpleural bands in lower lobe basals, rapidly spreading ground-glass, and consolidations are observed on HRCT (may be accompanied by Pneumomediastinum/subcutaneous emphysema).

5. Pleural and Vascular Involvements

Pleural Effusion: Rarer compared to RA or SLE (10-15%); frequently develops secondary to lower lobe atelectasis or heart failure.

Pneumomediastinum / Subcutaneous Emphysema: Can develop spontaneously as a result of rupture of parenchymal/subpleural cysts and cavities (Macklin effect) or severe vasculopathy (poor prognostic sign in Anti-MDA5).

Secondary Pulmonary Hypertension: Can develop secondary to severe and long-standing parenchymal fibrosis.

QUICK "MYOPATHY LUNG" SUMMARY CARD TO BE ADDED TO GUIDE (TRICK)

Trick / Golden Rule (Myopathy Lung Spectrum):

Ground-Glass + Consolidation + Diaphragmatic Elevation: NSIP/OP overlap pattern and basal volume loss due to muscle weakness (Inflammatory myopathy / Anti-synthetase syndrome).

1. **No / Minimal Muscle Findings + Rapidly Progressive Widespread Ground-Glass:** Anti-MDA5 positive RP-ILD (DAD) alarm; urgent intervention required.
2. **Consolidation in Posterior Segments + Centrilobular Nodule:** Chronic microaspiration pneumonia/bronchiolitis secondary to oropharyngeal muscle involvement.
3. **Spontaneous Pneumomediastinum + Subpleural Cyst:** Sign of severe parenchymal vasculopathy / tissue necrosis (poor prognosis).

SJÖGREN SYNDROME

Lung involvement in Sjögren syndrome is characterized by lymphocytic infiltration of exocrine glands and parenchyma. The disease frequently affects small airways (bronchiolitis) and parenchyma (cystic and lymphoproliferative diseases).

1. Parenchymal and Lymphoproliferative Involvement (Sjögren-ILD)

The parenchymal HRCT appearances most specific to Sjögren syndrome are cystic and lymphoid infiltrative processes:

LIP (Lymphoid Interstitial Pneumonia) Pattern (15-25%): Most specific HRCT appearance for Sjögren syndrome. Characterized by thin-walled cysts (usually 2-30 mm in size, peribronchovascular or parenchymal in distribution) located on a background of patchy ground-glass opacities (GGO) and centrilobular nodules.

NSIP Pattern (45-50% - Most Common): Symmetric ground-glass opacities, fine reticular increases, and traction bronchiectasis in lower lobe basals and subpleural area.

OP (Organizing Pneumonia) Pattern: Peribronchovascular or subpleural areas of consolidation, reverse halo (atoll) sign, and patchy parenchymal involvements.

UIP Pattern (5-10%): Rare. Courses with honeycombing appearance and traction bronchiectasis in lower lobes.

2. Airway Diseases (Small and Large Airway Involvement)

Prominent airway involvement is observed in 50-60% of Sjögren syndrome patients (even if clinically asymptomatic):

Follicular Bronchiolitis (Small Airway Involvement): Mosaic perfusion, air trapping accentuated on expiratory HRCT, centrilobular nodules, and tree-in-bud appearance secondary to lymphoid hyperplasia around airways.

Bronchiectasis and Bronchiolitis: Prominent thickening of bronchial walls, cylindrical bronchiectasis, and mucus plugs.

Xerotrachea (Dry Airway): Chronic cough secondary to tracheobronchial mucosal dryness, recurrent lower respiratory tract infections, and peribronchial fibrosis.

3. Malignancy Risk and Lymphoproliferative Complications

Sjögren syndrome is the connective tissue disease with the highest risk of lymphoma (especially MALT Lymphoma and Diffuse Large B-Cell Lymphoma):

- o **MALT Lymphoma / Nodular Lymphoid Hyperplasia (NLH):** Slowly growing, irregular-margined solitary or multiple nodules and consolidation masses on HRCT.
- o Air bronchograms and small cystic areas may be present within the mass or nodule.

Lymphadenopathy (LAP): Persistent, enlarging, or non-calcified mediastinal and hilar lymph node enlargements (should raise suspicion of lymphoma).

4. Pleural and Vascular Involvements

Pleural Effusion / Thickening: Quite rare compared to SLE or RA (5-10%); when seen, secondary infections, lymphoma, or serositis should be considered.

Pulmonary Arterial Hypertension (PAH): Rare, can develop secondary to vasculitis or chronic parenchymal disease.

Amyloidosis (Co-existing): Parenchymal amyloidosis can develop on the background of Sjögren; observed as parenchymal nodules that can calcify, cysts, and interlobular septal thickenings on HRCT.

5. Inflammatory and Iatrogenic Complications

Secondary Infections: Risk of staphylococcal, pseudomonal, and fungal infections increases as a result of impaired mucociliary clearance secondary to exocrine gland dysfunction.

Drug Side Effects: Hypersensitivity pneumonitis or opportunistic infections due to immunosuppressive therapy.

QUICK "SJÖGREN LUNG" SUMMARY CARD TO BE ADDED TO GUIDE (TRICK)

Trick / Golden Rule (Sjögren Lung Spectrum):

1. **Thin-Walled Cysts on Ground-Glass Background:** LIP (Lymphoid Interstitial Pneumonia) pattern, which is the most typical appearance for Sjögren syndrome.
2. **Air Trapping on Expiration + Centrilobular Nodules:** Small airway involvement (Follicular bronchiolitis / Mosaic perfusion).
3. **Newly Developing Consolidation Mass / Nodule with Air Bronchogram:** Biopsy warning should be made regarding MALT Lymphoma development!
4. **Dry Mouth/Eyes + Lower Lobe Fibrosis:** NSIP pattern is paramount (honeycombing is rare).

SYSTEMIC LUPUS ERYTHEMATOSUS (SLE)

Primary parenchymal fibrosis (ILD) is quite rare in SLE; clinically and radiologically, the disease presents predominantly as serositis (pleura/pericardium), vascular/acute parenchymal injury, and respiratory muscle dysfunction.

1. Pleural and Pericardial Involvement (Serositis - Most Common)

Serositis is the most characteristic and most frequently observed thoracic involvement pattern of SLE (30-50%):

Pleural Effusion and Pleuritis: Usually bilateral or unilateral, small-to-moderate amount, exudative pleural fluid. Presents with chest pain (pleuritis).

Pericardial Effusion: Frequently accompanies pleural fluid (Polyserositis). Pericardial thickening and enhancement can be observed.

Pleural Thickening and Adhesions: Focal/diffuse pleural thickenings developing secondary to chronic recurrent pleuritis attacks.

2. Acute and Life-Threatening Parenchymal Involvements

Acute parenchymal involvements in SLE carry a high mortality risk:

Diffuse Alveolar Hemorrhage (DAH) (2-5%): Most lethal emergency picture of SLE.

- o **HRCT Findings:** Bilateral widespread, geographically distributed ground-glass opacities (GGO) and alveolar consolidations. Lobular sparing and intralobular septal thickenings (crazy-paving) may accompany.
- o Manifests clinically with sudden hemoglobin drop and respiratory failure.

Acute Lupus Pneumonitis (1-4%): Rare but severe picture.

- o **HRCT Findings:** Widespread patchy ground-glass, consolidations, and accompanying pleural effusion in lower lobe basals and subpleural area.
- OP (Organizing Pneumonia) Pattern:** Peribronchovascular or subpleural patchy consolidations and reverse halo (atoll) sign.

3. Diaphragm and Respiratory Muscle Involvement (Extra-Parenchymal)

Shrinking Lung Syndrome: Specific to SLE, develops without parenchymal fibrosis or parenchymal disease.

- o **Etiology:** Myositis/neuropathy of diaphragm and intercostal muscles, and respiratory muscle restriction.
- o **HRCT Findings:** Parenchymal fibrosis is not observed; however, symmetric significant reduction in both lung volumes, elevation of both diaphragmatic domes, and secondary platelike (discoid) atelectasis in basals are observed.

4. Chronic Parenchymal Involvement (SLE-ILD - Rare)

Chronic fibrosis in SLE is quite rare compared to other connective tissue diseases (3-8%):

NSIP Pattern: Symmetric ground-glass opacities, reticular thickenings, and mild traction bronchiectasis in lower lobe basals.

UIP Pattern: Very rare; when honeycombing appearance is observed, another accompanying connective tissue disease (overlap) or RA should be considered.

5. Pulmonary Vascular Involvements and Thromboembolism

Pulmonary Arterial Hypertension (PAH): Frequently develops secondary to accompanying Antiphospholipid Syndrome (APS) or vasculitis. Main pulmonary artery diameter > 29 mm and right ventricular enlargement are observed on HRCT.

Pulmonary Thromboembolism (PTE): Risk is significantly increased in SLE patients with antiphospholipid antibody positivity. Filling defects in pulmonary arteries, infarct areas (wedge-shaped consolidations and 'reverse halo' appearance) are observed on contrast-enhanced CT angiography.

QUICK "SLE LUNG" SUMMARY CARD TO BE ADDED TO GUIDE (TRICK)

Trick / Golden Rule (SLE Lung Spectrum):

1. **Young Female + Bilateral Pleural Effusion + Pericardial Fluid + NO Fibrosis:**
Classic SLE Serositis / Polyserositis appearance.
2. **Acute Respiratory Failure + Hemoglobin Drop + Widespread Ground-Glass:**
Diffuse Alveolar Hemorrhage (DAH) emergency alert must be given!
3. **Reduction in Lung Volumes Without Fibrosis + Diaphragmatic Elevation:**
Shrinking Lung Syndrome.
4. **Sudden Onset Chest Pain / Dyspnea + Wedge-Shaped Consolidation:**
Pulmonary Thromboembolism / Infarction secondary to antiphospholipid syndrome should be investigated.

MIXED CONNECTIVE TISSUE DISEASE (MCTD)

MCTD is a systemic disease presenting with high-titer anti-U1-RNP antibody positivity, in which clinical and laboratory features of Systemic Sclerosis (SSc), Systemic Lupus Erythematosus (SLE), and Polymyositis (PM) overlap. Lung involvement is observed in 50-85% of patients and is among the main causes of morbidity/mortality.

1. Parenchymal Involvement (MCTD-ILD - SSc/PM-like)

Parenchymal involvement in MCTD mainly carries HRCT characteristics of Systemic Sclerosis and Idiopathic Inflammatory Myopathies:

NSIP Pattern (50-60% - Most Common): As in SSc, symmetric ground-glass opacities (GGO), fine reticular increases, and traction bronchiectasis/bronchiolectasis in lower lobe basals and subpleural/peripheral areas.

NSIP + OP Overlap Pattern (20-30%): Secondary to myositis component, subpleural/peribronkovascular ground-glass along with patchy areas of alveolar consolidation.

UIP Pattern (5-10%): Honeycombing appearance and prominent architectural distortion are observed in a small group.

2. Pulmonary Vascular Involvement (MCTD-PAH - SSc-like)

In MCTD, Pulmonary Arterial Hypertension (PAH) is one of the most important causes of death (develops in 10-20% of patients):

Isolated / Primary PAH: Develops secondary to direct vasculopathy without significant parenchymal fibrosis (similar to CREST form in SSc).

HRCT Findings: Main Pulmonary Artery (PA) diameter > 29 mm, PA / Ascending Aorta diameter ratio >1.0, right ventricular enlargement, and compression/flattening of the interventricular septum toward the left ventricle.

3. Esophageal and Aspiration Findings (SSc-like "Esophago-Lung" Relationship)

Esophageal smooth muscle atrophy and loss of motility are observed secondary to SSc component:

Esophageal Dilation (Wide-open Esophagus): Widening in thoracic esophageal lumen, air or fluid fullness inside.

Aspiration Pneumonia and Bronchiolitis: Chronic microaspirations secondary to esophageal reflux and dysmotility; lead to patchy consolidations, centrilobular nodules, and worsening bronchiectasis in lower lobes.

4. Pleural and Pericardial Involvement (SLE-like Serositis)

Serous membrane involvements are observed quite frequently depending on SLE component (30-50%):

Pleural Effusion and Pleuritis: Unilateral or bilateral, exudative pleural fluid.

Pericardial Effusion: Pericardial fluid and pericardial thickening accompanying pleural fluid (Polyserositis).

5. Respiratory Muscle and Diaphragm Involvement (Polymyositis-like)

Diaphragmatic Elevation and Volume Loss: Weakening in respiratory muscles secondary to myositis involvement, bilateral diaphragmatic dome elevation, and secondary platelike (discoid) atelectasis in lung bases.

QUICK "MCTD LUNG" SUMMARY CARD TO BE ADDED TO GUIDE (TRICK)

Trick / Golden Rule (MCTD Lung Spectrum):

- 1. NSIP Pattern (Ground-Glass) + Dilated Esophagus + Pleural Effusion:** Classic MCTD clinical picture presenting as a combination of three diseases (SSc+SLE+PM).
- 2. Pulmonary Artery Enlargement Without Fibrosis / with Minimal Fibrosis:**
Check for right heart strain findings due to high PAH risk.
- 3. Ground-Glass + Consolidation (NSIP/OP Overlap) + Diaphragmatic Elevation:**
Indicates accompanying inflammatory myositis component.

PULMONARY VASCULITIDES AND PARENCHYMAL NODULES

Pulmonary vasculitides are diseases with high mortality requiring rapid diagnosis and treatment, characterized by inflammation and necrosis of the pulmonary vascular bed (artery, vein, capillary). On HRCT, they present with findings fundamentally in the spectrum of parenchymal nodules/cavities, airway involvements, and acute alveolar hemorrhage (DAH).

1. Granulomatosis with Polyangiitis (GPA / Former name: Wegener's Granulomatosis)

GPA is the ANCA-associated (frequently PR3-ANCA / c-ANCA positive) vasculitis that most commonly forms mass and cavitory lesions in lung parenchyma.

Parenchymal Nodules and Cavities (70-90% - Most Common):

- o Randomly distributed nodules in both lungs, single or multiple in number, reaching sizes from a few millimeters to a few centimeters (mass).
- o **Cavitation:** Approximately 50% of nodules and masses undergo central necrosis forming irregular, thick-walled cavities (walls can thin with treatment).
- o **Halo Sign:** Faint ground-glass opacity surrounding the nodule or mass (indicates perilesional alveolar hemorrhage).

Airway and Tracheobronchial Involvement (15-25%): Subglottic stenosis, focal/diffuse thickenings narrowing the lumen in trachea and main bronchial walls, and secondary segmental atelectasis.

Pleural Involvement: Pleural thickening or reactive pleural effusion can accompany (10-20%).

2. Microscopic Polyangiitis (MPA)

MPA is a small vessel vasculitis (frequently MPO-ANCA / p-ANCA positive) that does not form granulomatous inflammation and prominent cavitory nodules; it primarily involves the capillary bed.

Diffuse Alveolar Hemorrhage (DAH) (50-70% - Most Common): Most characteristic and life-threatening emergency presentation of MPA.

- o **HRCT Findings:** Bilateral patchy or widespread geographically distributed ground-glass opacities (GGO), alveolar consolidations, lobular sparing, and intralobular septal thickenings (crazy-paving).

Chronic Parenchymal Fibrosis (MPA-ILD): In 20-30% of patients in the chronic period, NSIP or UIP-like (coursing with honeycombing) fibrotic changes develop in lower lobe basals.

3. Eosinophilic Granulomatosis with Polyangiitis (EGPA / Former name: Churg-Strauss Syndrome)

Necrotizing vasculitis characterized by asthma, marked eosinophilia in peripheral blood, and tissue eosinophilia (p-ANCA positivity frequently 30-40%).

Fleeting (Fluctuant) Consolidations: Focal, subpleural, or peribronchovascular-located, patchy consolidations and ground-glass areas on HRCT shifting spontaneously or with treatment.

Centrilobular Nodules and Airway Involvement: Prominent centrilobular ground-glass nodules, bronchial wall thickenings secondary to asthma, bronchiolitis, and mucus plugs.

Cavitation is Rare: Unlike GPA, cavitation of nodules in EGPA is extremely rare.

4. Diffuse Alveolar Hemorrhage (DAH) Emergency Management Note

Emergency picture that can develop in all vasculitides (MPA, GPA, EGPA) and connective tissue diseases (SLE, Goodpasture/Anti-GBM).

Clinical/Radiological Triad: Rapidly developing dyspnea + Sudden hemoglobin drop (hemoptysis may not be present in 30% of patients!) + Acute bilateral widespread geographic ground-glass/consolidation on HRCT.

Quick HRCT Sign: Reticular increases and irregularities appear in ground-glass areas within a few days with regression of bleeding, can completely clear in 1-2 weeks with appropriate treatment.

5. Rheumatoid Nodules (Necrobiotic Nodules)

Leading nodular parenchymal pathology seen in non-vasculitic rheumatic diseases.

Smooth / Regular-Margined Nodules: Nodules 0.5-5 cm in size, frequently located in subpleural area or upper/middle lobes in RA patients (especially males with subcutaneous nodules and RF/ACPA positive).

Cavitation: Nodules can undergo central necrosis forming smooth, thin-walled cavities. If opened to the pleura, can lead to pneumothorax or pyopneumothorax.

Caplan Syndrome: Rapidly developing large necrotic nodules in the lung periphery in the presence of RA + Coal worker's pneumoconiosis.

QUICK "VASCULITIS & NODULE" SUMMARY CARD TO BE ADDED TO GUIDE (TRICK)

Trick / Golden Rule (Vasculitis and Nodule Spectrum):

1. **Irregular Thick-Walled Cavitory Nodule/Mass + Sinusitis/Kidney:**
Granulomatosis with Polyangiitis (GPA / c-ANCA) is prioritized.
2. **Acute Widespread Ground-Glass Without Cavity + Sudden Anemia:** Diffuse Alveolar Hemorrhage (DAH/MPA/MPO-ANCA) emergency alert must be given!
3. **Asthma History + Eosinophilia + Fleeting (Fluctuant) Consolidation:**
Eosinophilic Granulomatosis with Polyangiitis (EGPA).
4. **Subpleural Smooth Thin-Walled Cavitory Nodule + Deformity in Joints:**
Rheumatoid Nodule (RA).

ANKYLOSING SPONDYLITIS (AS)

Lung involvement in ankylosing spondylitis is observed less frequently compared to other connective tissue diseases (1-2%); however, it is extremely specific and characteristic in terms of showing upper lobe-predominant apicobasal fibrobullous involvement.

1. Parenchymal Involvement (Apicobasal Fibrobullous Disease)

Unlike other rheumatic diseases, involvement in AS begins not from lower lobe basals, but from upper lobe apical sections of lungs:

Early Period (Apical Pleural Thickening and Parenchymal Infiltrate): Focal/diffuse pleural thickenings, subpleural reticular increases, and patchy apical ground-glass opacities at upper lobe apices.

Late / Advanced Period (Apical Fibrosis and Bullae):

- o Prominent symmetric or asymmetric fibrotic retraction, traction bronchiectasis, and architectural distortion in upper lobes.
- o Thin/thick-walled cysts, bullae, and paraseptal emphysema areas developing within fibrotic tissue.

Cavitation: Large, thick-walled cavities can form as a result of central necrosis of apical fibrotic areas.

2. Secondary Complications and Opportunistic Infections

Apical fibrobullous and cavitary lesions create a favorable background for secondary infections:

Fungal Ball (Mycetoma / Aspergilloma):

- o Most common complication developing inside apical cavities secondary to AS (in rates up to 50%).
- o **HRCT Findings:** Round/oval soft tissue mass located within the cavity moving with gravity, and air-crescent sign around it.

Atypical Mycobacterial Infections: Tuberculosis and Non-Tuberculous Mycobacteria (NTM) infections can easily settle in apical cavities.

3. Chest Wall and Skeletal System Involvement (Extra-Parenchymal)

Axial skeletal involvement directly restricts thoracic mechanics:

Joint Ankylosis: Ankylosis (fusion) and calcifications in costovertebral, costotransverse, and manubriosternal joints.

Thoracic Expansion Restriction: Significant reduction in rib cage expansion due to joint ankylosis (restrictive type respiratory limitation).

Spinal Deformities: Increased thoracic kyphosis, "bamboo spine" appearance in spine, syndesmophyte formations, and fractures in ankylosed spine (Andersson lesion).

4. Pleural and Vascular Involvements

Pleural Thickening: Chronic apical pleural thickening and pleural plaques, prominent at apices.

Pleural Effusion: Asymptomatic or mild level, extremely rare; when seen, secondary infections should be investigated.

Aortitis and Aortic Insufficiency: Ascending aortic wall thickening and dilated ascending aorta (observed in mediastinal window rather than lung window).

QUICK "ANKYLOSING SPONDYLITIS" SUMMARY CARD TO BE ADDED TO GUIDE (TRICK)

Trick / Golden Rule (Ankylosing Spondylitis Lung Spectrum):

1. **Fibrosis in Upper Lobe Apices + Bulla/Cavity + Fungal Ball (Aspergilloma):**
Classic Apicobasal Fibrobullous Disease specific to AS.
2. **Costovertebral Joint Ankylosis + Thoracic Kyphosis:** Skeletal involvement clues restricting thoracic expansion.
3. **Air-Crescent Sign Inside Apical Cavity:** Secondary *Aspergillus* colonization (mycetoma) warning must be given!

DRUG-INDUCED LUNG DISEASES AND INFECTION COMPLICATIONS (IATROGENIC)

Immunosuppressive, biological, and antifibrotic treatments used in rheumatic diseases can directly cause lung parenchymal toxicity (drug-induced pneumonitis), as well as prepare a background for opportunistic infections due to immunosuppression.

1. Drug-Induced Lung Injury (Drug Pneumonitis)

Picture that can appear days, weeks, or months after treatment onset, confusing clinically and radiologically with infection or rheumatic disease flare.

Methotrexate (MTX) Pneumonitis

Frequency & Course: Among the most common drug pneumonitides; usually develops subacutely within the first 1 year of treatment.

HRCT Findings:

- o Bilateral widespread or patchy ground-glass opacities (GGO) and alveolar consolidations.
- o Centrilobular ground-glass nodules and mild interlobular septal thickenings similar to hypersensitivity pneumonitis (HP).
- o Pleural effusion (accompanies in 30-50% of cases, more frequent compared to rheumatic ILD).

Leflunomide and Anti-TNF Agents (Infliximab, Etanercept, Adalimumab)

Leflunomide: Can lead to rapidly progressive interstitial pneumonia or sudden exacerbation (acute exacerbation) in pre-existing RA-ILD picture. Forms DAD (Diffuse Alveolar Damage) or NSIP/OP-like appearances on HRCT.

Anti-TNF Agents: Can trigger existing fibrosis or cause newly developing sarcoid-like granulomatous reactions (mediastinal/hilar LAP and lymphangitic distribution nodules).

Rituximab and Other Biological / Synthetic Target Therapies

Rituximab: Can form subacute hypersensitivity pneumonitis or organizing pneumonia (OP) pattern (subpleural/peribronchovascular consolidations) after treatment cycles.

JAK Inhibitors (Tofacitinib, Baricitinib) / Antifibrotics: Rarely lead to drug-induced ground-glass or eosinophilic pneumonia pictures.

2. Opportunistic Infections Secondary to Immunosuppression

Infection picture developing in rheumatic patients receiving intense immunosuppressive or biological agents, mimicking rheumatic involvement on HRCT.

Pneumocystis Jirovecii Pneumonia (PCP / PJP)

Risk Group: Especially patients using high-dose steroid, Methotrexate, or anti-TNF.

HRCT Findings:

- o Bilateral, symmetric, upper lobe and central-predominant mosaic ground-glass opacities (lobular sparing is prominent).
- o Thin-walled cysts (pneumatocele) and interlobular septal thickenings (crazy-paving) on ground-glass background.
- o Pleural effusion and lymphadenopathy are usually absent (absence is in favor of PCP).

Tuberculosis (TB) Reactivation and Atypical Mycobacteria (NTM)

Risk Group: Reactivation risk is significantly increased in patients receiving Anti-TNF treatment.

HRCT Findings:

- o Cavitary lesions, consolidations, and apical indurations in upper lobe apical and posterior segments.
- o **Miliary Tuberculosis:** Homogeneous miliary nodules 1-3 mm in size, randomly distributed in both lung parenchyma.
- o Centrilobular nodules and tree-in-bud appearances.

Fungal Infections (Invasive Aspergillosis / Candidiasis)

HRCT Findings:

- o **Halo Sign:** Ground-glass halo ring secondary to hemorrhage around fungal nodule (Early-stage invasive aspergillosis).
- o **Air-Crescent Sign:** Necrosis and air space formation within nodule as a result of neutrophil recovery with treatment (Late stage).

Viral Infections (CMV / HSV / VZV)

HRCT Findings: Bilateral patchy ground-glass opacities, centrilobular small nodules, ground-glass nodules, and interlobular septal thickenings.

3. Differential Diagnosis Criteria (Drug Pneumonitis vs. Infection vs. Disease Flare)

Feature	Drug-Induced Injury (Drug Pneumonitis)	Opportunistic Infection (e.g., PCP/TB)	Rheumatic Disease Flare
Onset	Subacute (Weeks/Months)	Acute / Subacute (Days/Weeks)	Chronic / Subacute Flare
Fever & Leukocytosis	Variable (Eosinophilia may accompany) Common (e.g., 50% in MTX)	High Fever, Prominent Leukocytosis/Leukopenia ABSENT in PCP, Unilateral present in TB	Compatible with disease activity COMMON in RA/SLE, Rare in SSc
Pleural Effusion	Widespread, patchy	PCP: Central GGO + Cyst, TB: Apical Cavity	Lower lobe basal-predominant (UIP/NSIP)
Distribution	GGO, Centrilobular RAPID improvement with drug withdrawal and steroid	Drug withdrawal not enough, Antimicrobial required	Improves when immunosuppression is increased
Response to Drug Withdrawal			

QUICK "IATROGENIC & INFECTION" SUMMARY CARD TO BE ADDED TO GUIDE (TRICK)

Trick / Golden Rule (Iatrogenic Spectrum):

- 1. Ground-Glass + Pleural Effusion + Methotrexate History:** MTX Pneumonitis (Drug-secondary hypersensitivity should be considered rather than rheumatic fibrosis).
- 2. Bilateral Central Mosaic Ground-Glass + Thin-Walled Cyst + NO Effusion:** Pneumocystis Jirovecii Pneumonia (PCP) emergency alert in patient receiving Steroid/Immunosuppressive!
- 3. Apical Cavity / Tree-In-Bud in Patient Receiving Anti-TNF Treatment:** Sputum AFB and PCR correlation should be requested regarding Tuberculosis Reactivation.
- 4. Ground-Glass Halo Appearance Around Nodule:** Sign of Invasive Fungal Infection (Aspergillosis) in immunocompromised patient.

1. Basic Rheumatic Diseases and HRCT Findings

Disease	Basic HRCT Pattern	Key HRCT and Radiological Findings	Important Additional/Extra-Pulmonary Clues
Rheumatoid Arthritis (RA)	UIP (40-60%), predominant NSIP (30-40%), honeycombing, traction OP, DAD/AIP	Subpleural and lower lobe- bronchiectasis, architectural distortion.	Male patient, smoking history, pleural thickening/effusion, air trapping.

Disease	Basic HRCT Pattern	Key HRCT and Radiological Findings	Important Additional/Extra-Pulmonary Clues
Systemic Sclerosis (SSc / Scleroderma)	NSIP (80-90%), UIP (10-15%)	Symmetric ground-glass opacities (GGO), reticulation, traction bronchiectasis in lower lobes.	Subpleural sparing, dilated atonic esophagus, calcinosis cutis.
Idiopathic Inflammatory Myopathies (PM/DM)	NSIP + OP Overlap (50-60%), RP-ILD (DAD)	Peribronchovascular ground-glass opacities along with simultaneous consolidation areas.	Diaphragmatic elevation, basal discoid atelectasis, muscle atrophy, fulminant DAD in presence of Anti-MDA5.
Sjögren Syndrome	LIP (15-25%), NSIP (45-50%)	Thin-walled peribronchovascular cysts on ground-glass background, centrilobular nodules.	Xerostomia/Xerophthalmia (dry mouth/eyes), follicular bronchiolitis, air trapping, MALT lymphoma risk.
Systemic Lupus Erythematosus (SLE)	Serositis (Primary fibrosis is very rare)	Bilateral/unilateral pleural effusion, peribronchovascular consolidations.	Shrinking Lung Syndrome (volume loss without fibrosis), Diffuse Alveolar Hemorrhage (DAH), pericardial effusion.
Mixed Connective Tissue Disease (MCTD)	NSIP (50-60%), NSIP+OP Overlap	Ground-glass, reticulation, traction bronchiectasis in lower lobe basals.	SSc (esophageal dilation) + SLE (serositis) + PM (diaphragmatic elevation) overlap findings.
Ankylosing Spondylitis (AS)	Apicobasal Fibrobullous Disease	Fibrosis, traction bronchiectasis, cysts and cavities in upper lobe apices.	Fungal Ball (Aspergilloma / Air crescent) inside apical cavity, costovertebral joint ankylosis.

2. Airway, Pleural, Vascular, and Parenchymal Nodule Pathologies

Pathology / Clinical Condition	Related Diseases	Key HRCT Findings	Radiological & Clinical Trick
Rheumatoid / Necrobiotic Nodules	Rheumatoid Arthritis (RA)	Smooth-margined nodules 0.5-5 cm in subpleural/upper-	Forms pneumothorax if opened to pleura; forms

Pathology / Clinical Condition	Related Diseases	Key HRCT Findings	Radiological & Clinical Trick
		middle lobes, central cavitation.	Caplan Syndrome in coal worker.
Granulomatosis with Polyangiitis (GPA / Wegener)	ANCA Vasculitis (c-ANCA / PR3)	Randomly distributed, irregular thick-walled cavitory nodules/masses.	Halo Sign secondary to bleeding around nodule, sinusitis and kidney involvement accompany.
Microscopic Polyangiitis (MPA)	ANCA Vasculitis (p-ANCA / MPO)	Alveolar capillaritis, NSIP/UIP-like fibrosis in chronic period.	Does not form cavity; primarily presents with emergency Diffuse Alveolar Hemorrhage (DAH) picture.
Eosinophilic Granulomatosis with Polyangiitis (EGPA)	ANCA Vasculitis (Churg-Strauss)	Patchy, fleeting (fluctuant) consolidations, centrilobular ground-glass nodules.	History of asthma and prominent peripheral eosinophilia is present; cavitation is extremely rare.
Diffuse Alveolar Hemorrhage (DAH)	MPA, GPA, SLE, Anti-GBM	Bilateral widespread/patchy geographic ground-glass opacities, alveolar consolidations.	Emergency Picture: Sudden hemoglobin drop + Respiratory failure (Hemoptysis may not always be present).
Small Airway Disease (Bronchiolitis)	RA, Sjögren	Air trapping on expiratory HRCT, mosaic perfusion, centrilobular nodules.	Follicular and obliterative (constrictive) bronchiolitis; xerotrachea and mucus plugs.
Pleural Involvement (Pleuritis / Effusion)	SLE, RA, MCTD	Single or bilateral pleural effusion, pleural thickening and adhesions.	Glucose is extremely low in RA effusion; pericardial effusion accompanies in SLE (polyserositis).
Pulmonary Arterial Hypertension (PAH)	SSc (CREST), MCTD, SLE	Main Pulmonary Artery diameter > 29 mm (or PA/Aorta ratio > 1.0), right ventricular enlargement.	Isolated PAH without fibrosis (Group 1) is a common cause of death in SSc and MCTD.

3. Iatrogenic Conditions and Drug/Infection Differential Diagnosis

Condition / Agent	Related Drug or Clinical Background	Key HRCT Findings	Differential Diagnosis Criterion
Methotrexate (MTX) Pneumonitis	Methotrexate use	Bilateral widespread/patchy ground-glass opacities, centrilobular nodules.	Accompanied by pleural effusion (30-50%); rapidly regresses with drug withdrawal and steroid.
Pneumocystis Jirovecii (PCP / PJP)	Intense immunosuppression / Steroid	Bilateral symmetric, upper lobe/central mosaic ground-glass, thin-walled cysts.	Pleural effusion and LAP ARE ABSENT; absence supports PCP diagnosis.
Tuberculosis (TB) Reactivation	Anti-TNF therapies	Cavities, miliary nodules, tree-in-bud in upper lobe apical/posterior segments.	Reactivation risk is high in those receiving Anti-TNF; AFB/PCR correlation required.
Invasive Fungal Infection	Severely immunocompromised patient	Halo Sign around nodule (early), Air-Crescent Sign (late).	Represents <i>Aspergillus</i> invasion and alveolar hemorrhage area.

SUMMARY PRACTICAL DIFFERENTIAL DIAGNOSIS TREE AND QUICK CHECK CARDS (MASTER CHECKLIST)

The cards below are detailed clinical and radiological guidance references providing decision-making within seconds at the reporting desk while examining HRCT.

CARD 1: LOWER LOBE / BASAL-PREDOMINANT PARENCHYMAL INVOLVEMENTS

HRCT Basic Appearance	Related Disease / Pattern	Key HRCT and Radiological Findings	Differential Diagnosis & Trick Findings
Honeycombing Present	RA-UIP	Lower lobe basal-predominant honeycombing, traction bronchiectasis, architectural distortion.	Male patient, smoking history, pleural thickening/plaque, accompanying bronchiolitis.

HRCT Basic Appearance	Related Disease / Pattern	Key HRCT and Radiological Findings	Differential Diagnosis & Trick Findings
Honeycombing Present	SSc-UIP	Honeycombing appearance in lower lobe basals (cysts are usually small).	Presence of atonic dilated esophagus (wide-open esophagus).
Ground-Glass (GGO) Predominant	SSc-NSIP	Symmetric ground-glass opacities, reticulation, traction bronchiectasis in lower lobes.	Subpleural sparing, dilated esophagus, calcinosis cutis.
Ground-Glass (GGO) Predominant	PM / DM (Anti-Synthetase)	Peribronchovascular/subpleural ground-glass + consolidation (NSIP/OP Overlap).	Diaphragmatic elevation, basal discoid atelectasis, chest wall muscle atrophy.
Ground-Glass (GGO) Predominant	Sjögren (LIP)	Patchy ground-glass opacities and centrilobular small nodules.	Thin-walled cysts on ground-glass background, xerostomi/xerophthalmia.

- **If Honeycombing + Traction Bronchiectasis is Prominent:**
 - **First to Consider:** RA-UIP (Male patient, smoking history, pleural thickening, pleuritis, presence of accompanying bronchiolitis/mosaic perfusion supports diagnosis).
 - **Second to Consider:** SSc-UIP (Honeycombing cysts are small, esophageal dilation accompanies).
- **If Ground-Glass Opacity (GGO) + Reticulation is Prominent (Honeycombing Absent / Minimal):**
 - **Subpleural Sparing + Atonic Esophagus:** Systemic Sclerosis (SSc-NSIP).
 - **Ground-Glass + Consolidation (NSIP/OP Overlap) + Diaphragmatic Elevation:** Idiopathic Inflammatory Myopathies (PM/DM / Anti-Synthetase Syndrome).
 - **Thin-Walled Cysts on Ground-Glass Background + Dryness:** Sjögren Syndrome (LIP Pattern).

CARD 2: ACUTE/SUBACUTE DEVELOPING GROUND-GLASS AND CONSOLIDATION (EMERGENCY ALARM CARD)

Clinical / Serological Picture	Related Emergency / Agent	Key HRCT Findings	Emergency Management & Trick Warning
No / Minimal Muscle Involvement	Anti-MDA5 DM (RP-ILD/DAD)	Subpleural bands in lower lobes, rapidly spreading ground-glass and consolidations.	RP-ILD emergency alarm coursing fatal within days/weeks!
Sudden Hemoglobin Drop	Diffuse Alveolar Hemorrhage (DAH)	Bilateral widespread, geographically distributed ground-glass opacities, alveolar consolidations.	Complication of ANCA vasculitides (MPA/GPA) or SLE; urgent intervention required.
Immunocompromised Patient / Steroid	Pneumocystis Jirovecii (PCP/PJP)	Bilateral symmetric, upper lobe/central-predominant mosaic ground-glass, thin-walled cysts.	Pleural effusion and LAP ARE ABSENT (absence is in favor of PCP).
Drug Use (Methotrexate)	Methotrexate (MTX) Pneumonitis	Bilateral widespread/patchy ground-glass opacities, centrilobular nodules.	Pleural effusion accompanies (30-50%); rapidly improves with drug withdrawal and steroid.

□ No/Minimal Muscle Involvement + Respiratory Failure Progressing Within Days:

Anti-MDA5 Positive Dermatomyositis (RP-ILD / DAD) - High mortality emergency alarm.

Sudden Hemoglobin Drop + Widespread Geographic Ground-Glass: Diffuse Alveolar Hemorrhage (DAH) complication of ANCA Vasculitides (MPA/GPA) or SLE.

Bilateral Central Mosaic Ground-Glass + Thin-Walled Cyst + NO Pleural Effusion: Pneumocystis Jirovecii Pneumonia (PCP) - Immunocompromised patient.

Patchy Ground-Glass + Centrilobular Nodules + Pleural Effusion PRESENT (50%): Methotrexate (MTX) Pneumonitis.

- Hypersensitivity reaction.

CARD 3: NODULE, CAVITY, AND CYSTIC PARENCHYMAL PATHOLOGIES

Lesion Character	Related Disease / Picture	Key HRCT Findings	Radiological Sign & Trick
Irregular / Thick-Walled Cavity	Granulomatosis with Polyangiitis (GPA / Wegener)	Randomly distributed, irregular thick-walled cavitory nodules and masses.	Halo Sign secondary to bleeding around nodule; sinusitis and kidney involvement accompany.
Smooth-Margined Subpleural Nodule	Rheumatoid / Necrobiotic Nodule (RA)	Subpleural/upper-middle lobe smooth nodules 0.5-5 cm (can cavitate centrally).	Forms pneumothorax if opened to pleura; forms Caplan Syndrome in coal worker.
Thin-Walled Multiple Cysts	Sjögren Syndrome (LIP)	Thin-walled cysts with peribronchovascular or parenchymal distribution.	Cysts are located on background of ground-glass opacities.
Upper Lobe Apical Cyst / Bulla / Cavity	Ankylosing Spondylitis (AS)	Symmetric fibrosis, cyst, bulla, and cavity formations at apices.	Fungal Ball (Aspergilloma / Air-Crescent Sign) can develop inside cavity.

Irregular, Thick-Walled Cavitory Nodules/Masses + Halo Sign: Granulomatosis with Polyangiitis (GPA / c-ANCA).

Smooth/Regular-Margined Subpleural Nodules (Can Cavitate Centrally): Rheumatoid / Necrobiotic Nodule (RA).

Peribronchovascular Thin-Walled Cysts on Ground-Glass Background: Sjögren Syndrome (LIP).

Cyst/Bulla/Cavity in Upper Lobe Apices + Fungal Ball (Air Crescent): Ankylosing Spondylitis (Apicobasal Fibrobullous Disease).

CARD 4: EXTRA-PARENCHYMAL AND CLUE (TRICK) FINDINGS

Extra-Parenchymal Finding	Related Disease / Condition	Key HRCT & Thorax Finding	Clinical / Radiological Trick
Thoracic Esophageal Dilation	Systemic Sclerosis (SSc) / MCTD	Prominent widening in thoracic esophageal lumen, air or fluid fullness inside.	"Esophago-lung" relationship; leads to chronic microaspiration bronchiolitis and consolidation.

Extra-Parenchymal Finding	Related Disease / Condition	Key HRCT & Thorax Finding	Clinical / Radiological Trick
Pleuro-Pericardial Serositis	Systemic Lupus Erythematosus (SLE)	Bilateral/unilateral pleural effusion and pericardial fluid (Polyserositis).	Parenchymal fibrosis is ABSENT or minimal.
Diaphragmatic Elevation & Muscle Atrophy	Inflammatory Myopathies (PM/DM)	Bilateral diaphragmatic dome elevation, discoid atelectasis in basals.	Accompanying parenchymal fibrosis (NSIP/OP overlap) is present.
Diaphragmatic Elevation & Muscle Atrophy	Shrinking Lung Syndrome (SLE)	Symmetric significant reduction in lung volumes, diaphragmatic elevation.	Parenchymal fibrosis is ABSENT; secondary to respiratory muscle weakness.
Main Pulmonary Artery > 29 mm	SSc (CREST) / MCTD	PA diameter > 29 mm (or PA/Aorta > 1.0), right ventricular enlargement, septal flattening.	Isolated Pulmonary Arterial Hypertension without fibrosis or with minimal fibrosis (Group 1 PAH).

Thoracic Esophageal Widening (Air/Fluid-Filled Lumen): Systemic Sclerosis (SSc) or Mixed Connective Tissue Disease (MCTD).

Bilateral Pleural Fluid + Pericardial Fluid + NO Fibrosis: SLE Serositis (Polyserositis).

Diaphragmatic Elevation + Volume Loss in Lung Bases (Discoid Atelectasis):

o If Fibrosis is Present → Inflammatory Myopathies (PM/DM).

o If Fibrosis is Absent → Shrinking Lung Syndrome (SLE).

□ **Main Pulmonary Artery > 29 mm (Without Fibrosis / with Minimal Fibrosis):**

Isolated Pulmonary Arterial Hypertension associated with SSc (CREST) or MCTD (Group 1 PAH).