TABLE 17-1 MANIFESTATIONS OF INTERSTITIAL LUNG DISEASE

DISEASE	PHYSICAL EXAMINATION	RADIOGRAPHS	LABORATORY FINDINGS	HISTOLOGIC FINDINGS
Pneumoconioses				
Silicosis	Various findings	Large nodules Eggshell calcification of hilar nodes PMF, upper lobes Emphysema, nodules	Restrictive PFTs	Silica: inflammation, birefringent crystals, alveolar proteinosis
Coal worker's pneumoconiosis	Normal	PMF	FEV1 and FVC may be decreased	Coal: pigmented macules, anthracotic pigment
Asbestosis	Crackles	Pleural plaques, lower lobe fibrosis	Restrictive PFTs	Asbestos: UIP pattern, asbestos bodies, mesothelioma
Beryllium exposure	Nonspecific findings	Lymphadenopathy, lung nodules	Nonspecific except beryllium: lymphocyte transformation test	Beryllium: noncaseating granulomas
Hypersensitivity pneumonitis	Fever, cough, crackles	Centrilobular nodules, air trapping, fibrosis	Serum precipitins to specific proteins CD8>CD4 cells Lymphocytic BAL fluid Obstructive and/or restrictive PFTs	Chronic airway centered inflammation, poorly formed granulomas
Idiopathic interstitial pneumonias				
DIP, RB-ILD	Various findings	Centrilobular nodules, ground-glass infiltrates	Nonspecific	DIP: Intra-alveolar accumulation of pigmented macrophages
IPF	Crackles, clubbing	Basilar predominant fibrosis, honeycombing	Restrictive PFTs	IPF: UIP pattern with heterogeneous areas of fibrosis, fibroblast foci
AIP	Tachypnea, respiratory distress	Bilateral alveolar infiltrates	Nonspecific	AIP: Diffuse alveolar damage
NSIP	Crackles, possible clubbing	Ground-glass, subpleural reticulations	Restrictive PFTs	NSIP: Uniform thickening of interstitium with inflammatory cells and fibrosis
Collagen vascular	Collagen vascular disease, crackles, pleural rub	Pleural effusions Diffuse interstitial infiltrates, nodular infiltrates Occasional cavities	Serologic findings for specific disease Occasionally obstructive, usually restrictive PFTs	Interstitial inflammation Vasculitis Bronchiolar obstruction Organizing pneumonia Fibrosis: UIP, NSIP, LIP
Drug-induced ILD	Fever, crackles, pleural rub	Fibrosis Migratory infiltrates, diffuse interstitial infiltrates Pulmonary edema	Restrictive PFTs	Alveolar macrophages with lamellar bodies in amiodarone Interstitial inflammation Fibrosis Eosinophilic infiltration
Sarcoidosis	Fever, malaise, weight loss Erythema nodosum, lupus pernio, and skin plaques Salivary and lacrimal gland enlargement Arthritis Iritis, uveitis, chorioretinitis; keratoconjunctivitis Cranial nerve palsies Occasional rales or wheezes	Reticulonodular infiltrates Nodules Hilar adenopathy Mediastinal adenopathy Fibrosis	Lymphocytic BAL, T4 > T8 cell subsets Obstructive and/or restrictive PFTs Elevated transaminases with liver involvement Occasional hypercalcemia	Noncaseating granuloma with giant cells and negative acid-fast bacilli and fungal staining Fibrosis
Radiation exposure	Crackles, fever	Focal interstitial infiltrates corresponding to radiation port Occasional diffuse infiltrates Fibrosis	Restrictive PFTs	Acute: endothelial and alveolar lining cell damage Chronic: fibrosis
Pulmonary Langerhans cell histiocytosis	None to cough, dyspnea, chest pain Fatigue, weight loss, occasional fever	Spontaneous pneumothorax Nodules Reticulonodular infiltrates Middle and upper lobe predominance Honeycombing Sparing of costophrenic angle Cysts and nodules on HRCT	Normal lung volumes with decreased DLCO	OKT-6 (CD1) and S100-positive immunostaining Few eosinophils Peribronchiolar inflammation Macrophages filling lumen of bronchioles and intraluminal fibrosis

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