Thoracic Manifestations of Systemic Disease

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Thoracic Manifestations of Systemic Disease

- Neoplastic
- Thrombotic
- Infectious
- Immune
- Drug toxicity
- Metabolic
- Deposition
- Storage
- Hereditary

Systemic Inflammation

- Systemic Inflammatory Response Syndrome (SIRS)
- Autoimmune (collagen vascular)
- IRIS
- Autoimmune (IBD)
- Autoimmune (vasculitis)
- Drug toxicity

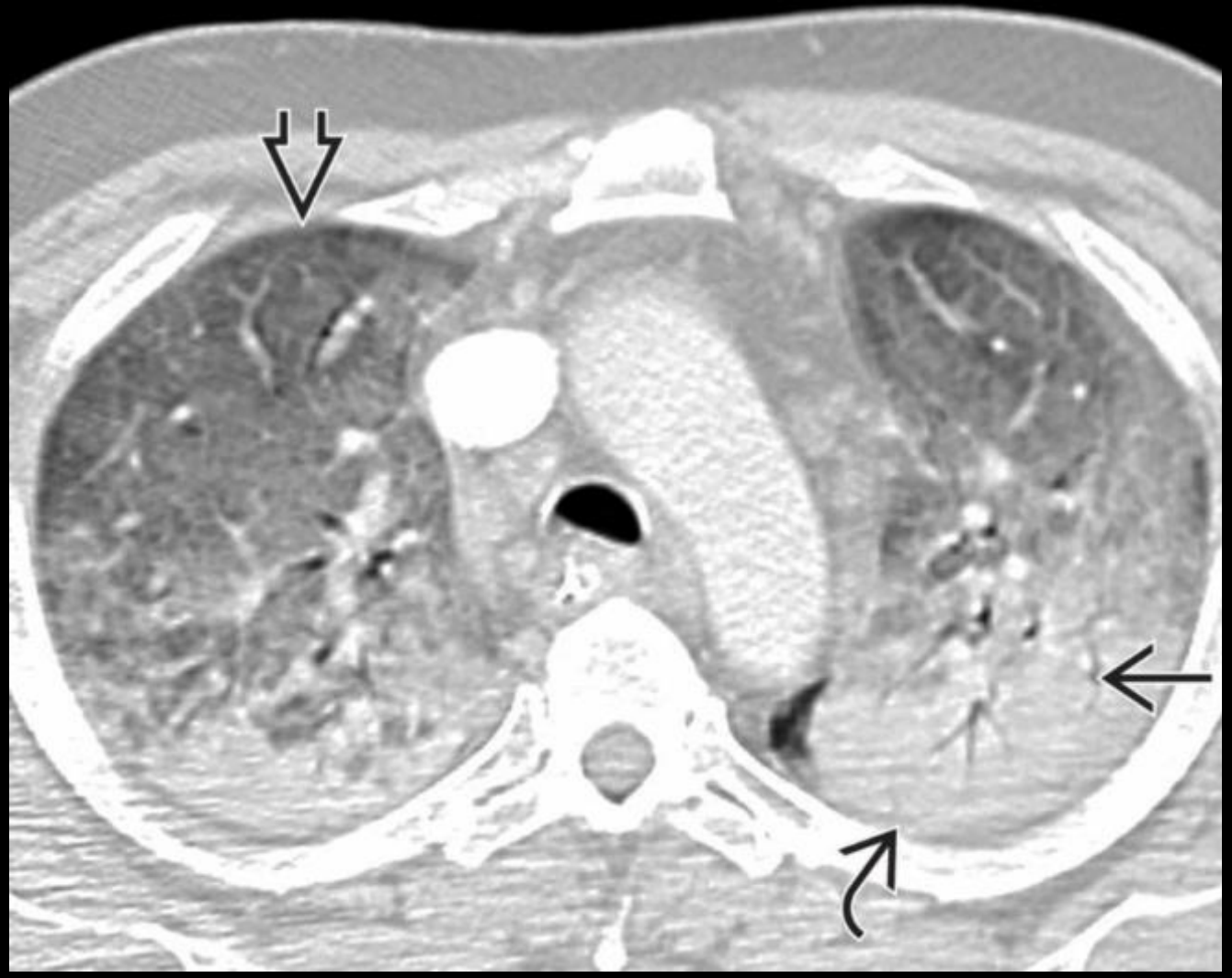
Systemic Inflammation

Systemic Inflammatory Response Syndrome

Acute hypoxemic respiratory failure in a critically ill patient due to effects of circulating inflammatory mediators

Diffuse alveolar damage

Patchy and/or confluent ground glass and consolidation, often worse dependently



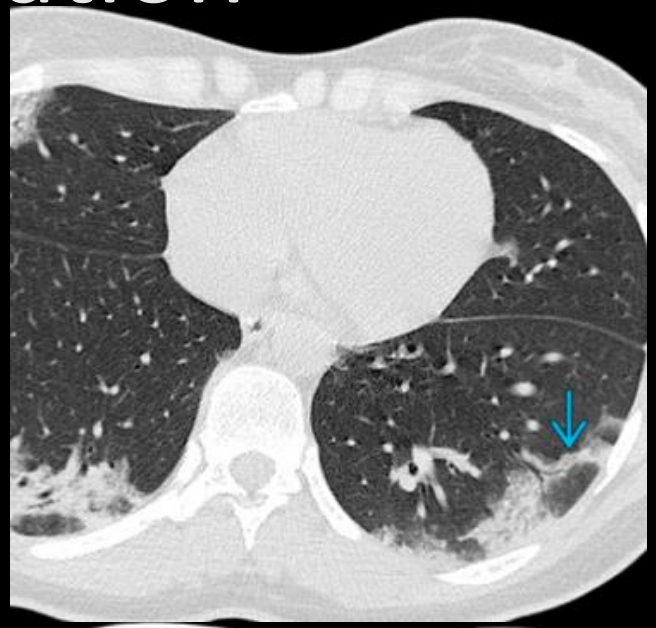
Systemic Inflammation

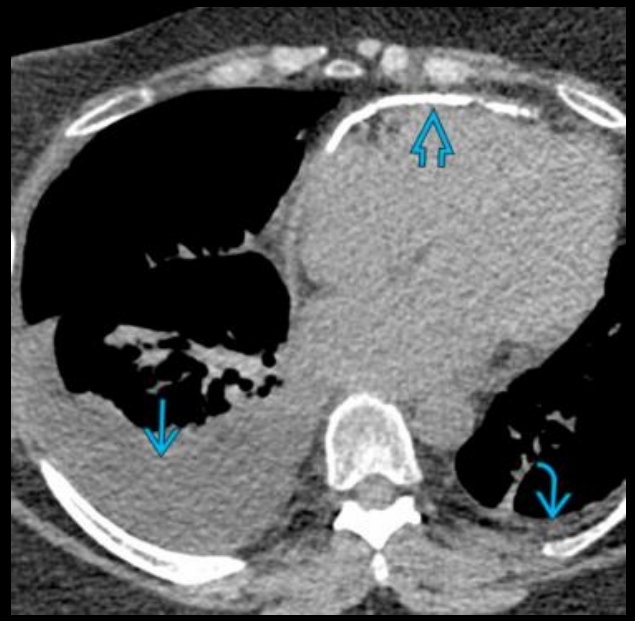
Systemic Lupus Erythematosis

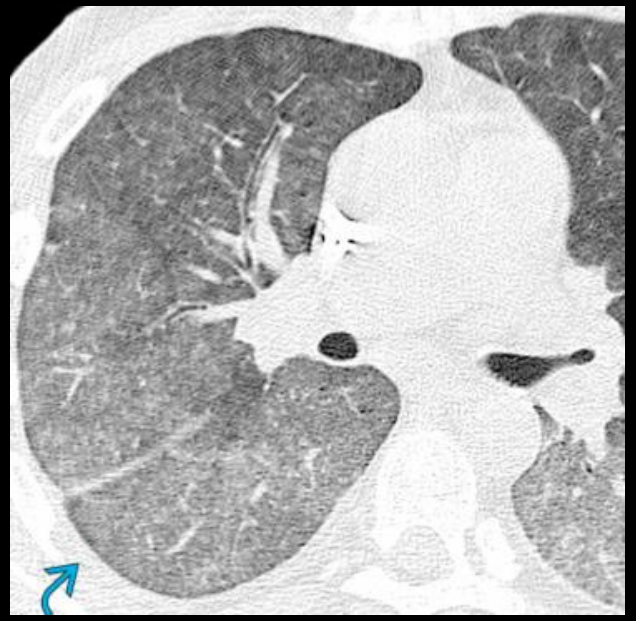
Multiorgan involvement. Most common presenting manifestations: Photosensitive rash, glomerulonephritis, arthritis.

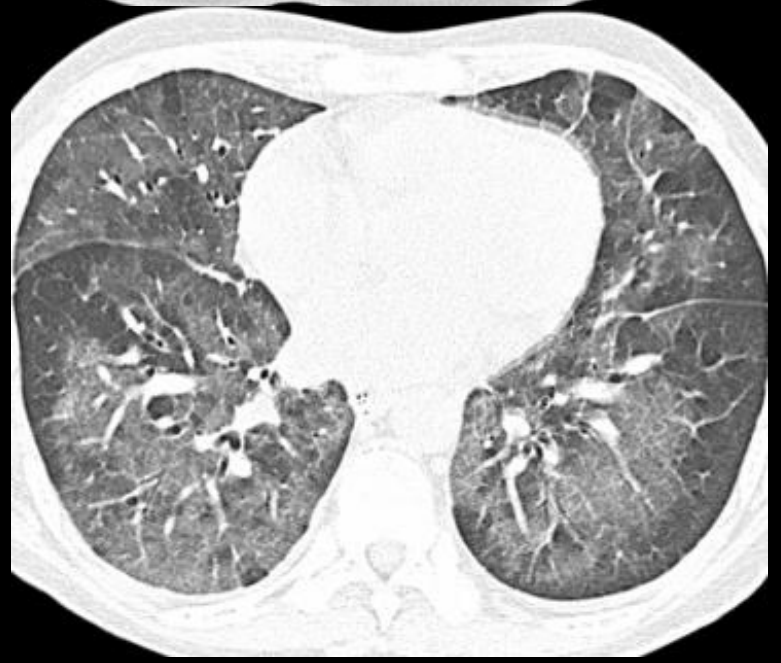
Pleural and lung involvement in 50-60% patients

NSIP, UIP, OP, DAD, vasculitis with DAH





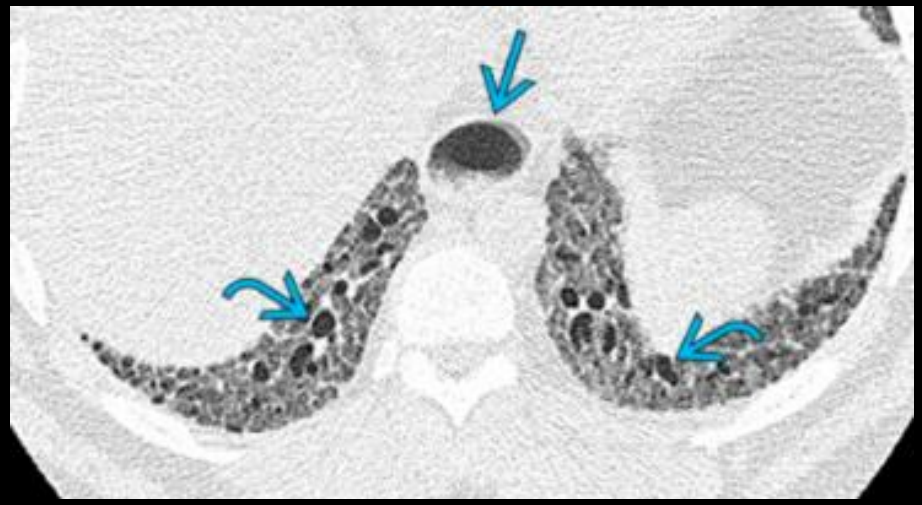




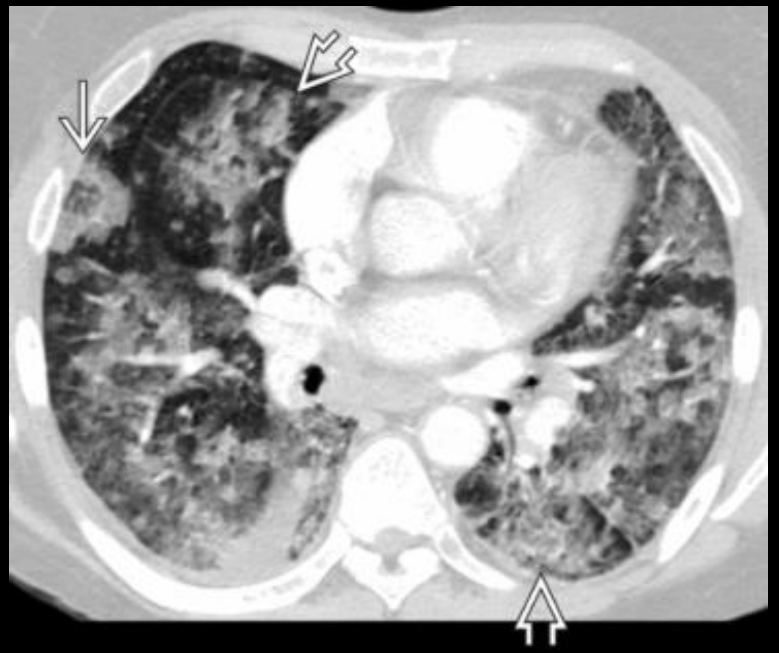
Systemic Inflammation

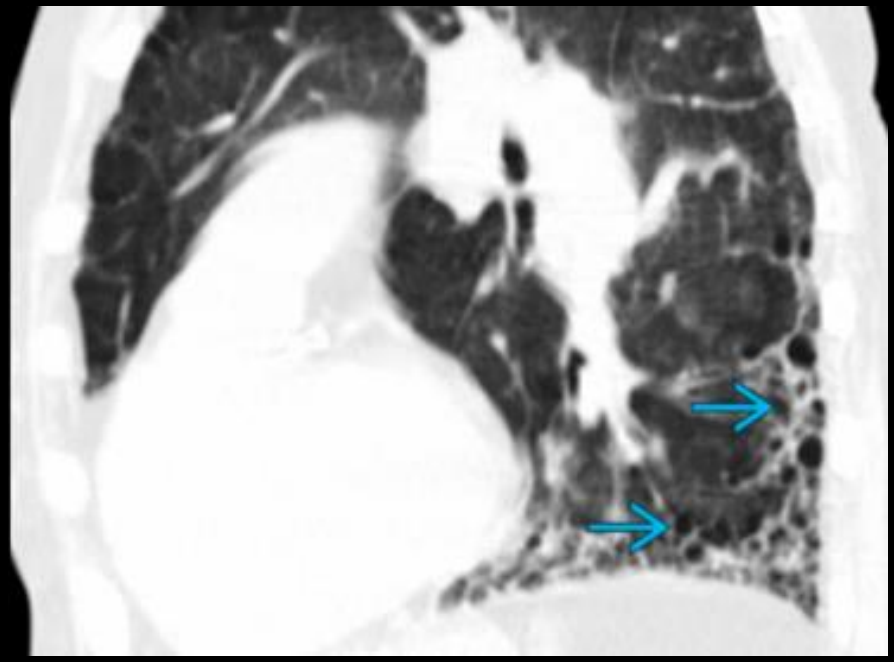
Progressive Systemic Sclerosis

Multiorgan disease characterized by overproduction and pathologic deposition of collagen.



Reduced circulating T-suppressor cells and NK cells (which may normally suppress fibroblast proliferation)





UIP, NSIP, OP, DAD.
Dilated esophagus

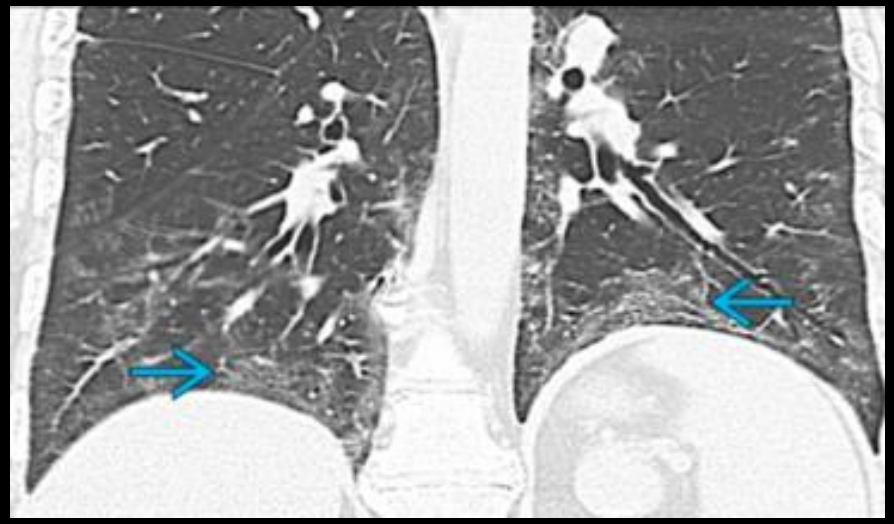
Systemic Inflammation

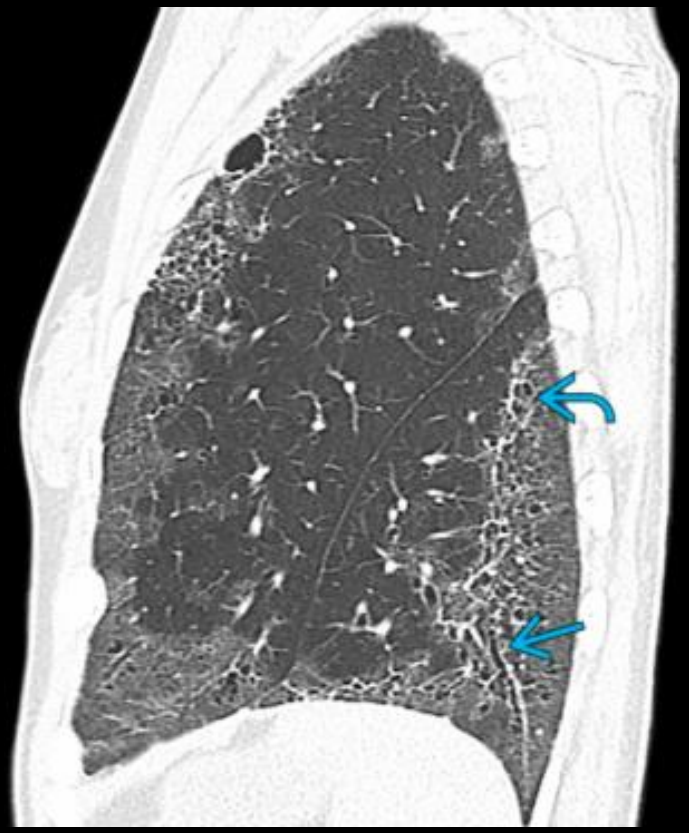
Poly-/Dermatomyositis

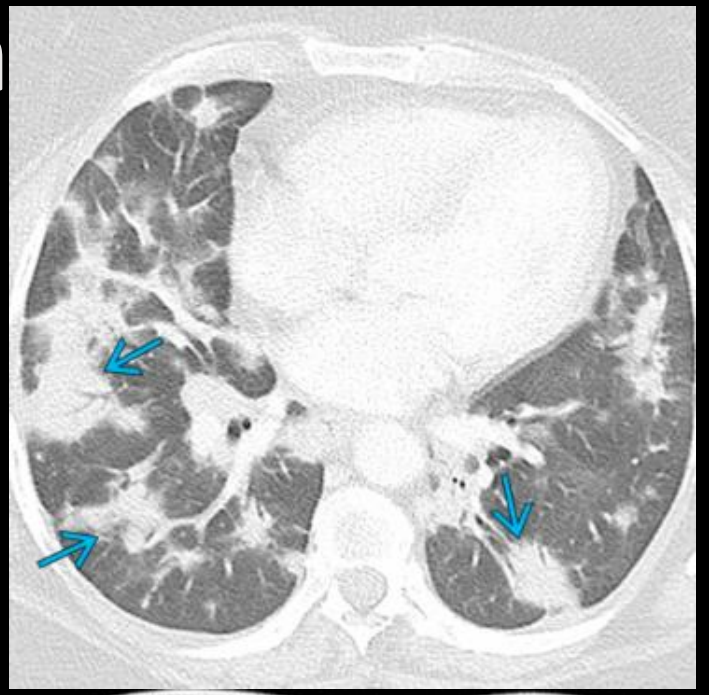
Autoimmune myositis (usually proximal) +/- skin manifestations

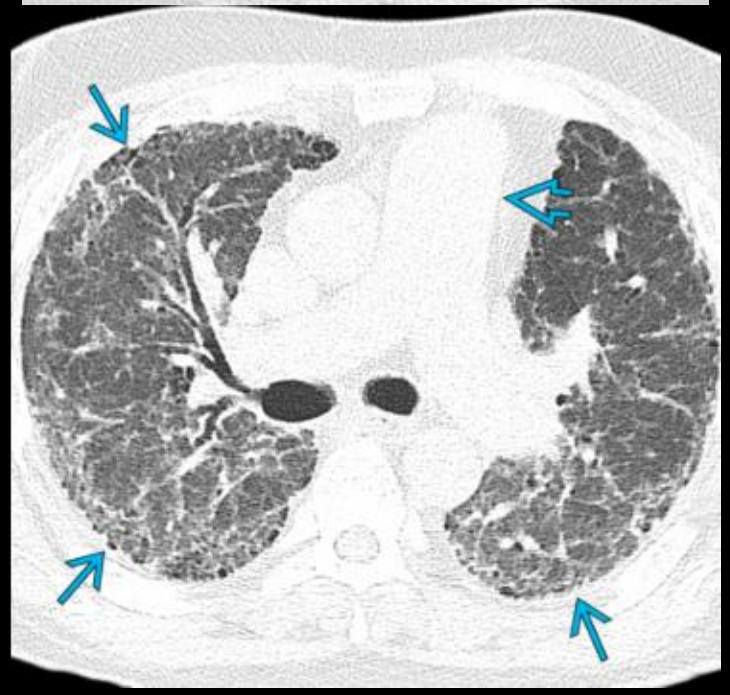
UIP, NSIP, OP, DAD

Antisynthetase variant: supradiaphragmatic pancaking







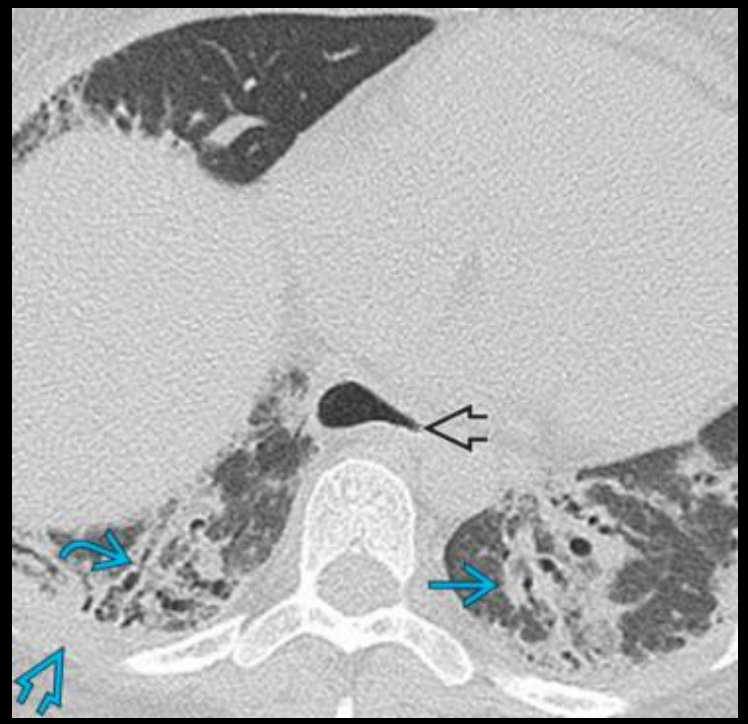


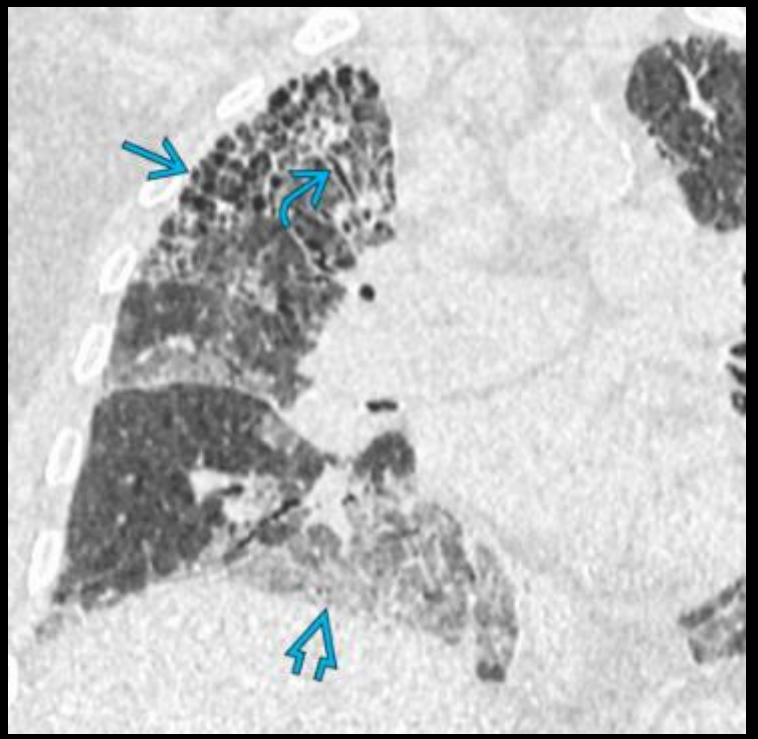
Systemic Inflammation

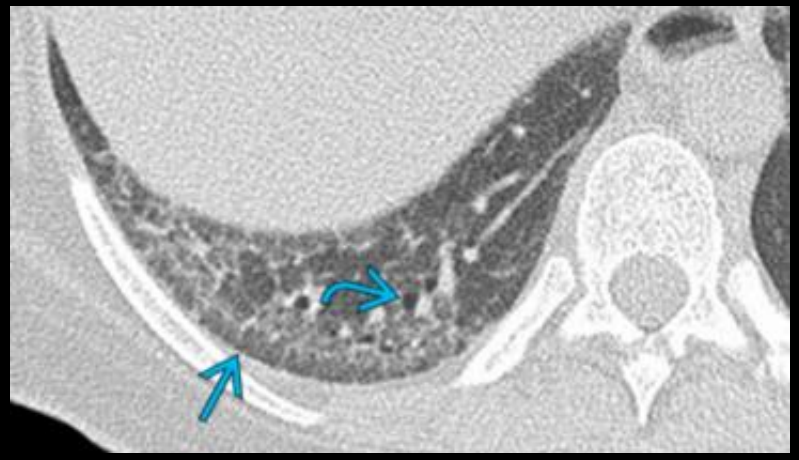
Mixed Connective Tissue Disease

Systemic syndrome with mixed clinical and laboratory findings

UIP, NSIP, OP, DAD





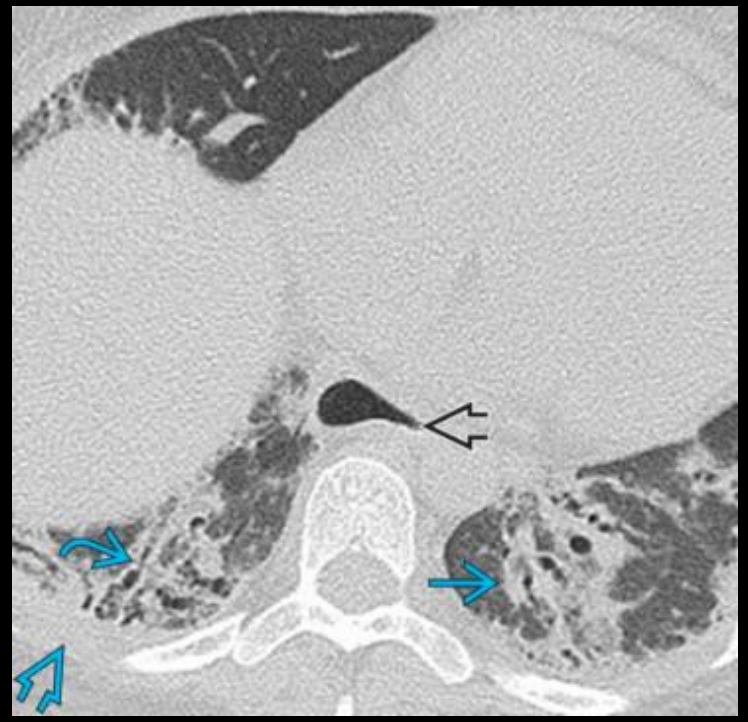


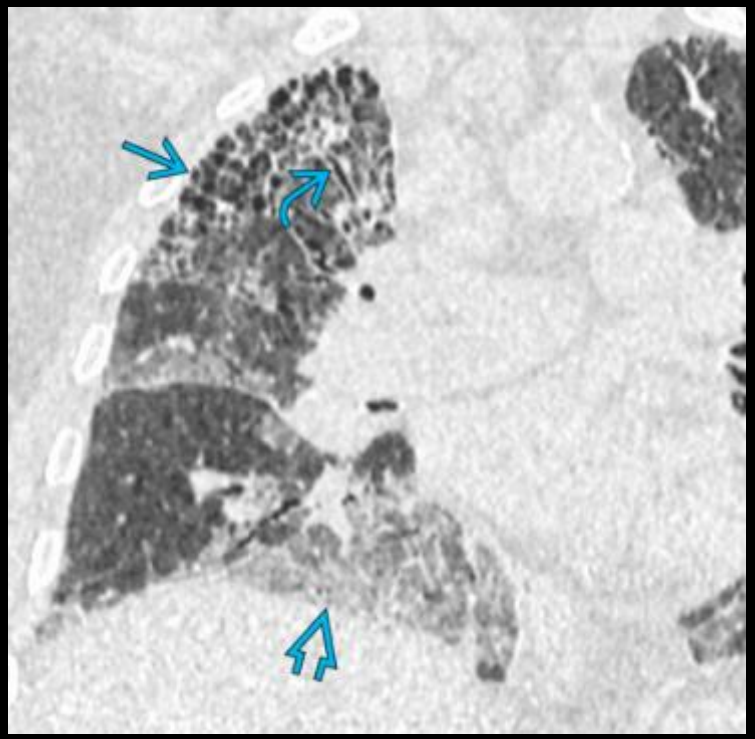
Systemic Inflammation

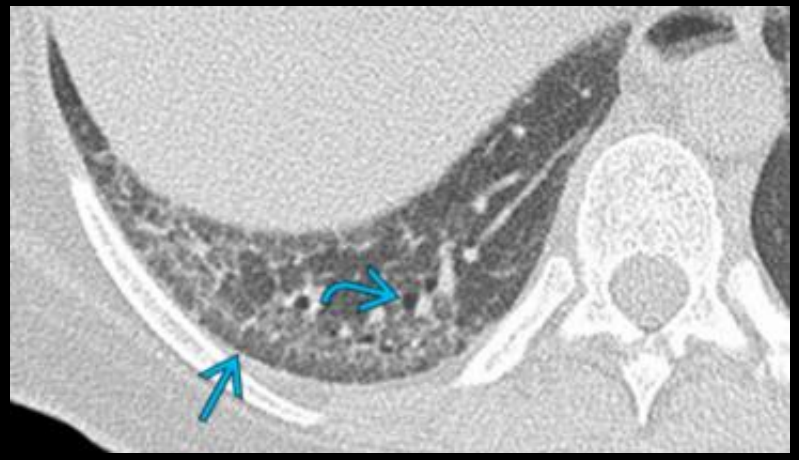
IgG4-related

Multiorgan autoimmune syndrome may involve bile ducts, pancreas, pachymeninges, etc.

UIP, NSIP, OP, DAD



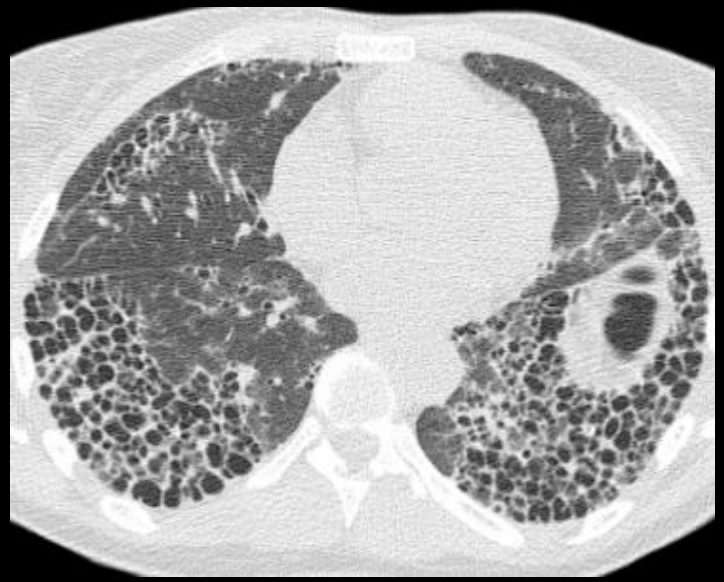




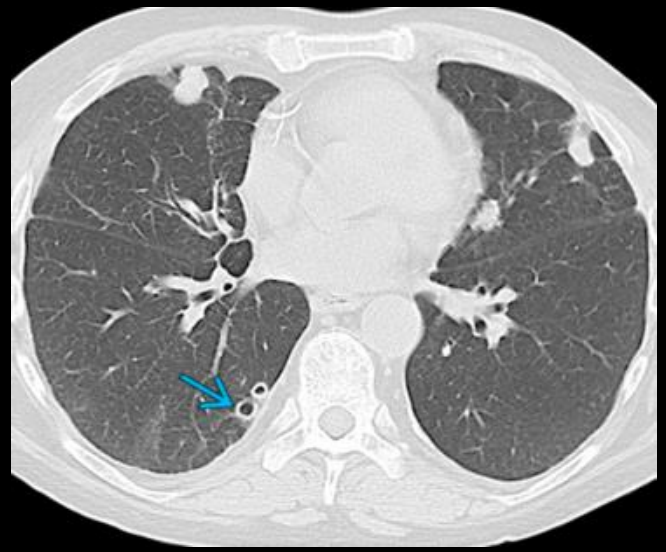
Systemic Inflammation

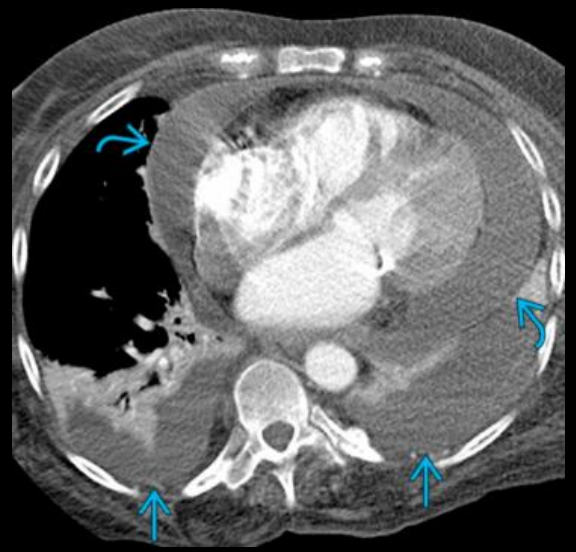
Rheumatoid Arthritis

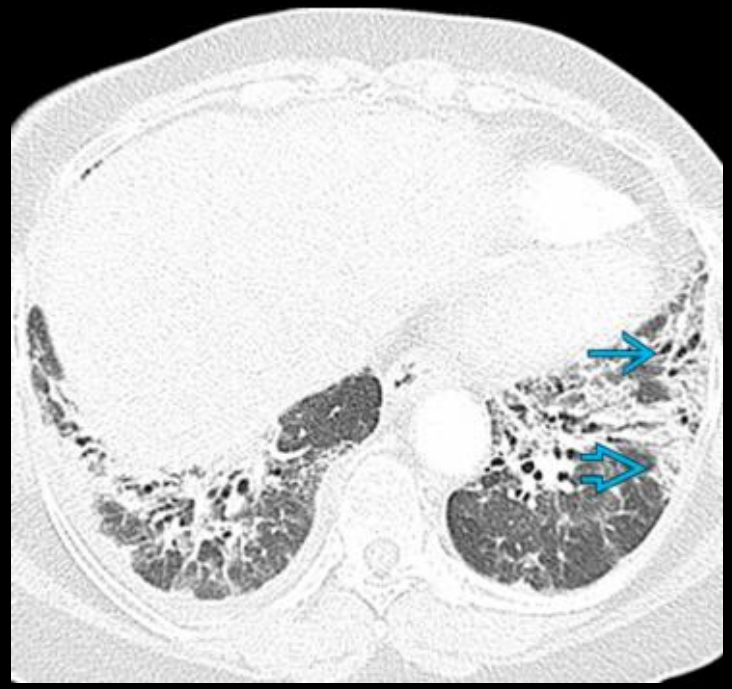
Inflammatory condition affecting articular synovium and (often) lungs, serosae, and vessels



Lymphocyte-predominant. Systemic perivasculitis. Proliferative synovitis. ILD: UIP, NSIP, OP, DAD, necrobiotic nodules







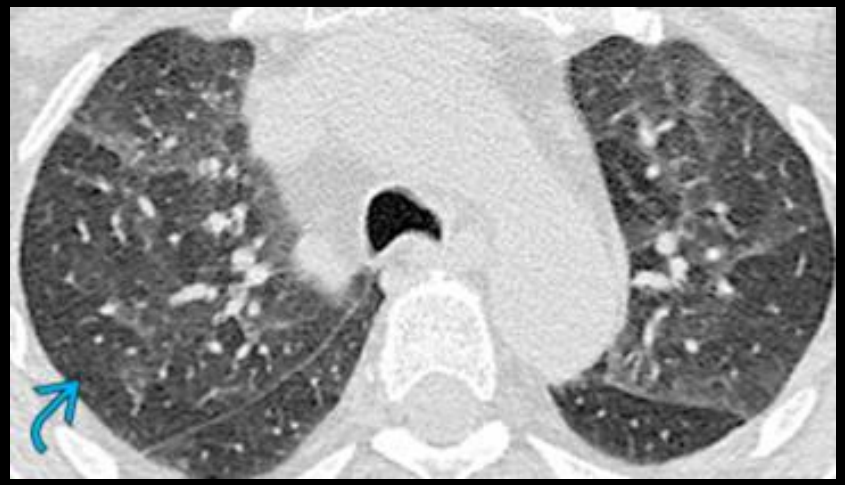
Systemic Inflammation

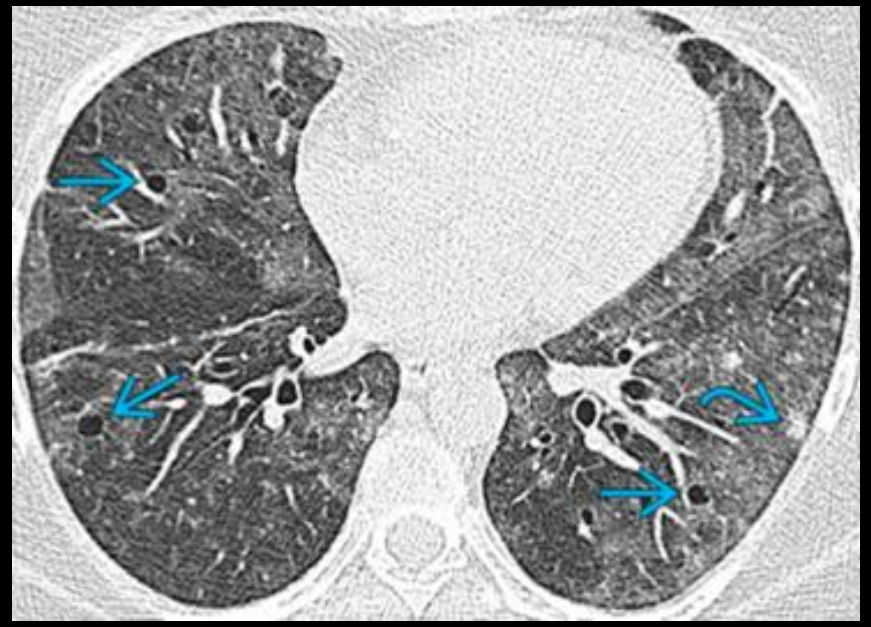
Sjogren Syndrome

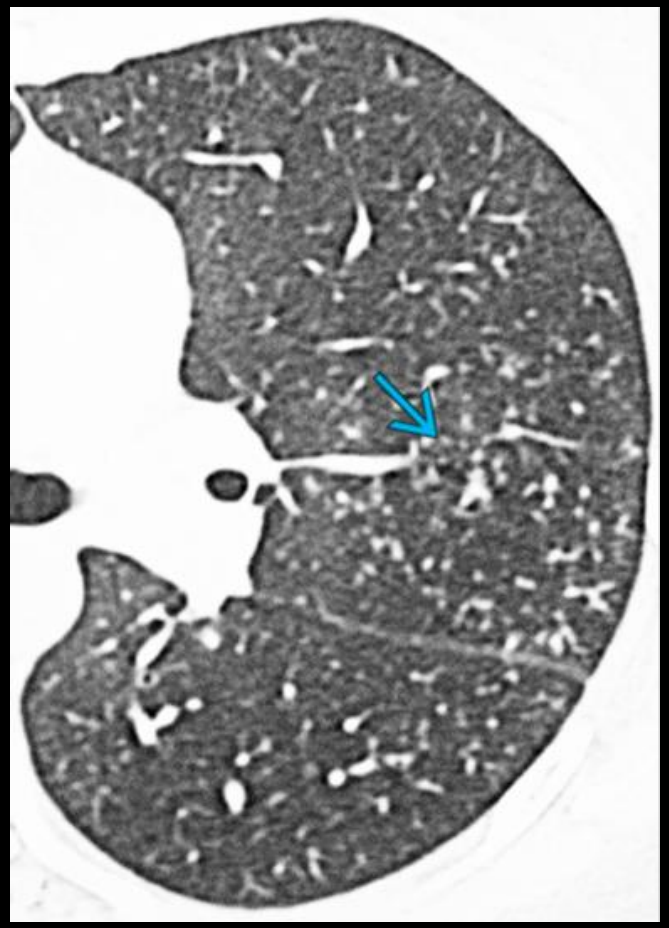
Dry cough, dry eyes, dry skin, dry mucous membranes. May be primary or associated with other rheumatologic or hematologic diseases

Lymphocytic infiltration of exocrine glands

UIP, NSIP, OP but also FB, LIP, BO



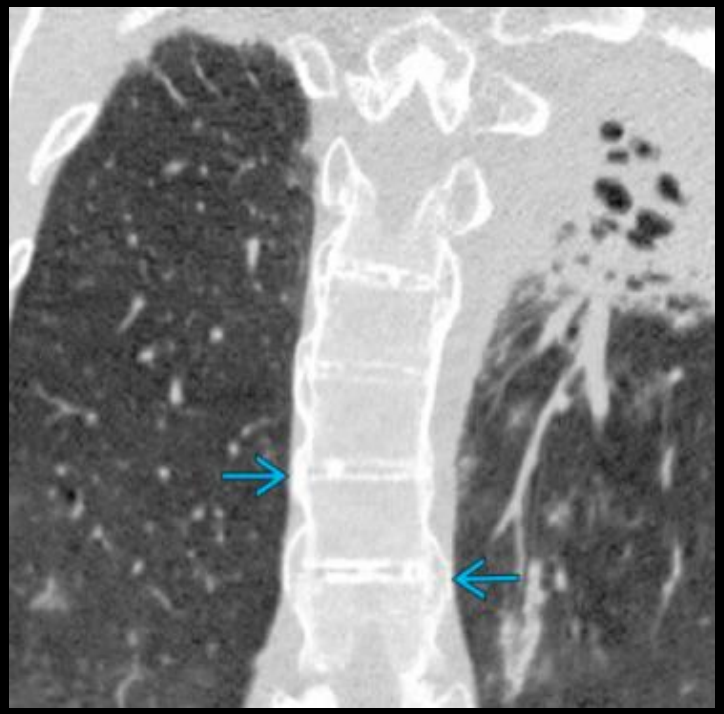


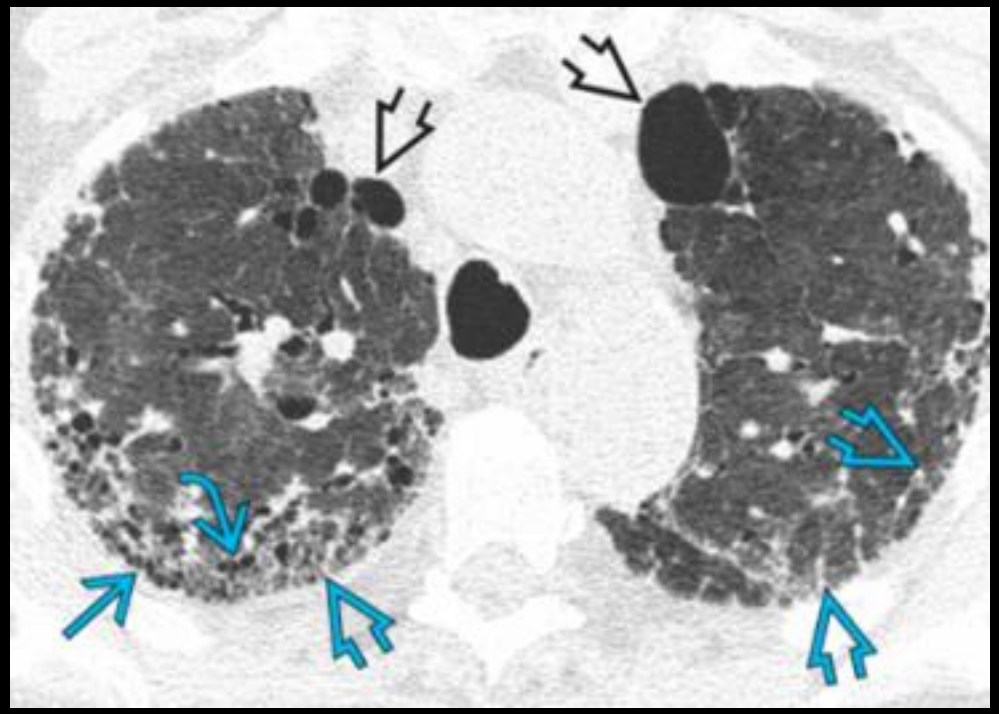


Systemic Inflammation

Ankylosing Spondylitis

Multisystem autoimmune disorder with predilection to axial skeleton. Spinal disease nearly always precedes lungs





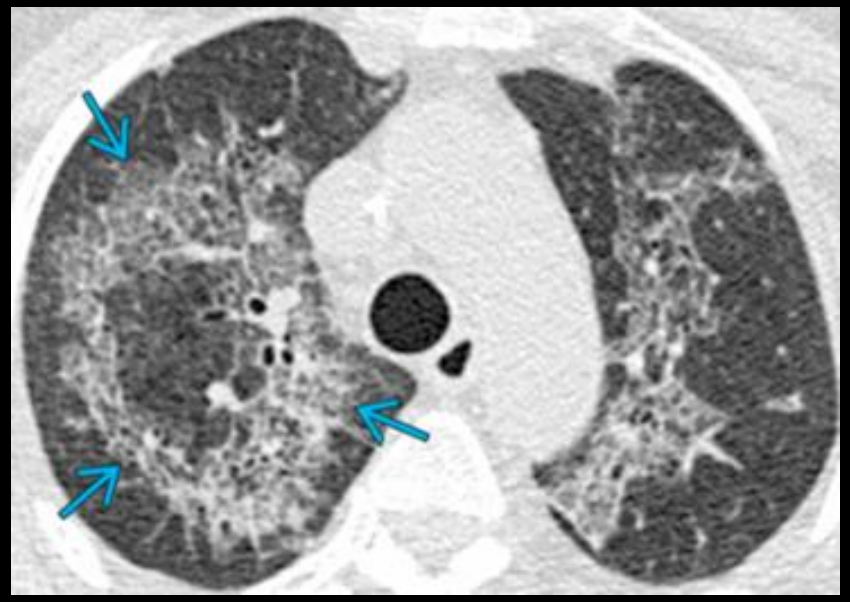
Varied airway disease (bronchial thickening, bronchieactasis, BO)

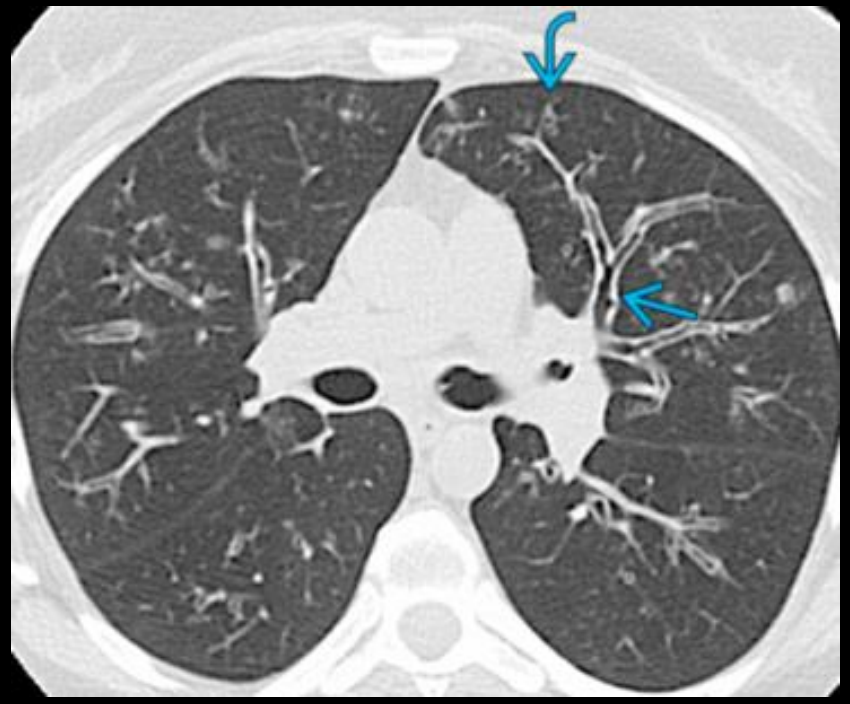
Varied ILD patterns

ULZ predominant fibrobullous disease is classic

Systemic Inflammation

Inflammatory Bowel Disease





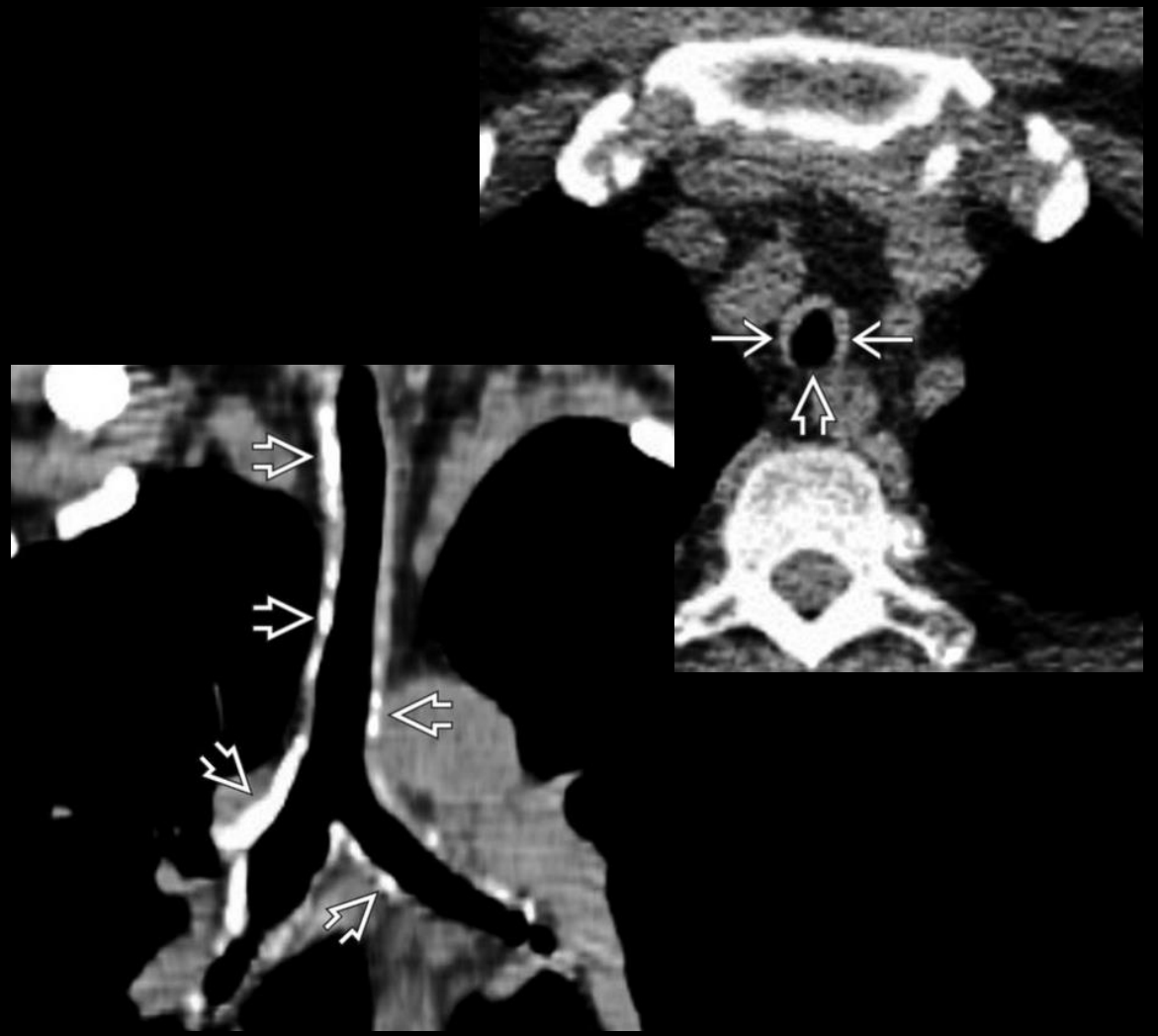
ILD, especially NSIP and OP

Bronchitis

Systemic Inflammation

Relapsing polychondritis

Rare autoimmune disorder resulting in multifocal cartilage inflammation & destruction



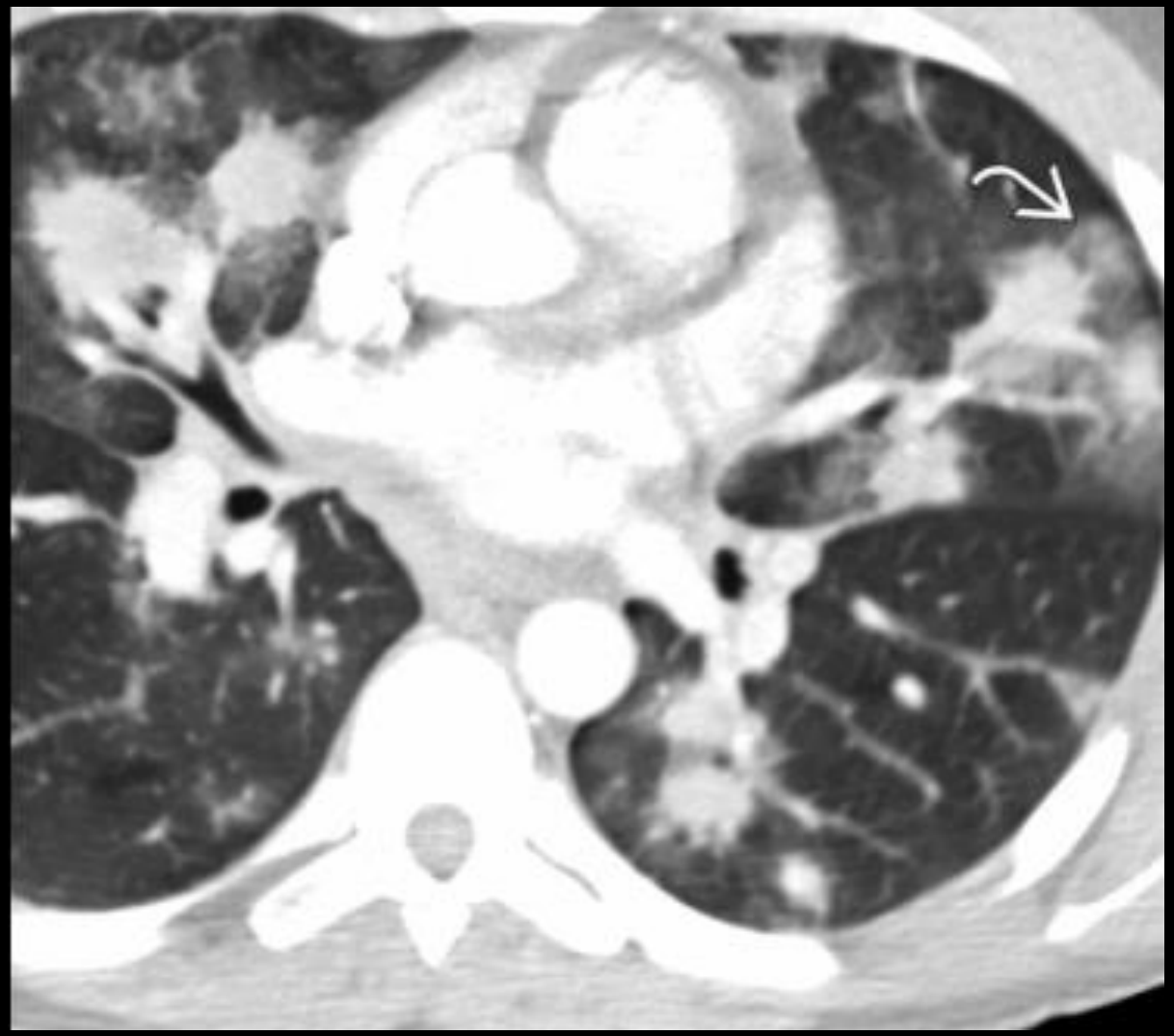
Focal or diffuse involvement airway wall thickening. Posterior tracheobronchial membrane spared.

Systemic Inflammation

Immune Reconstitution Inflammatory Syndrome

Paradoxical clinical and/or imaging deterioration due to recovery of immune function after initiation of HAART

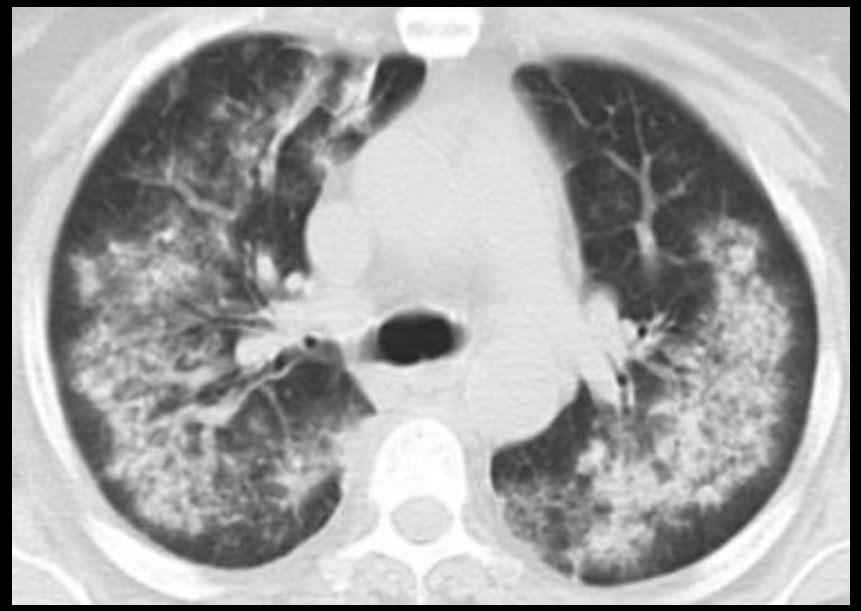
Development or worsening of pre-existing (known or occult) infection or neoplasm

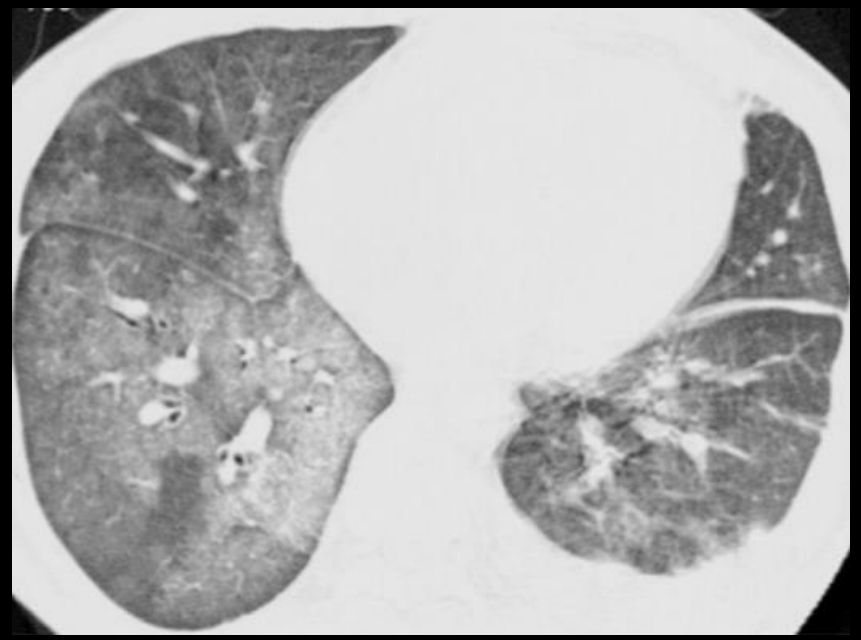


Systemic Inflammation

Small and medium vessel vasculitides

EGPA (Churg Strauss)
Essential cryoglobulinemia
Microscopic polyangiitis
GPA (Wegener)
Pulmonary-renal syndrome (Goodpasture)





- Diffuse ground glass (alveolar hemorrhage)
  - may demonstrate peribronchovascular distribution
  - often spares the extreme periphery

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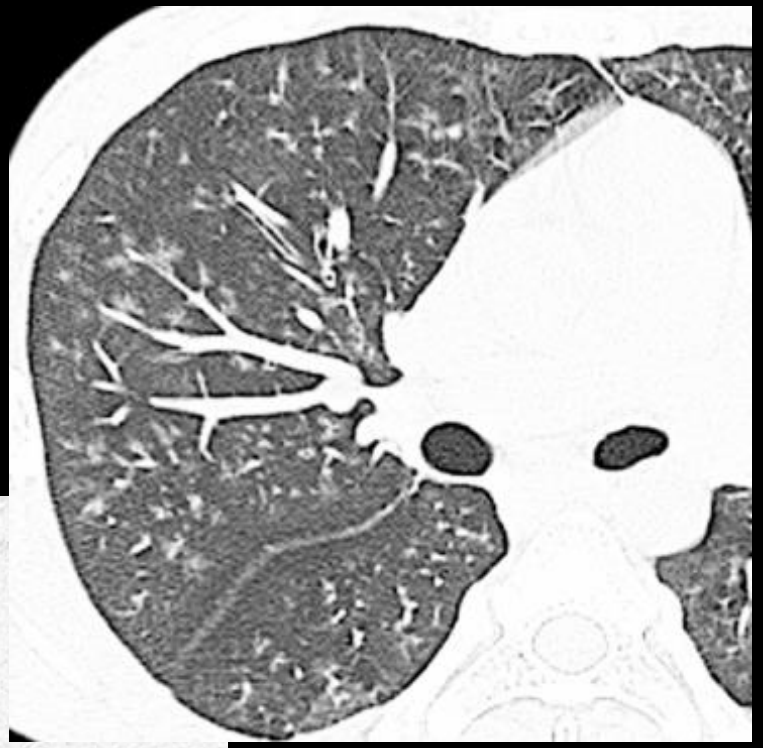

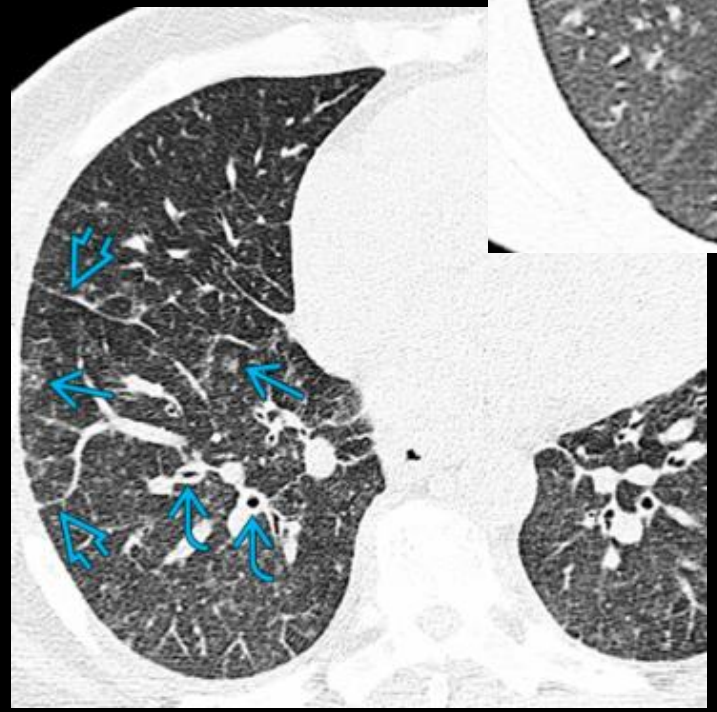

- Diffuse ground glass (alveolar hemorrhage)
- Perivascular inflammatory nodules
- Septal and peribronchial thickening (increased lymphatic return)

Systemic Inflammation

Small and medium vessel vasculitides

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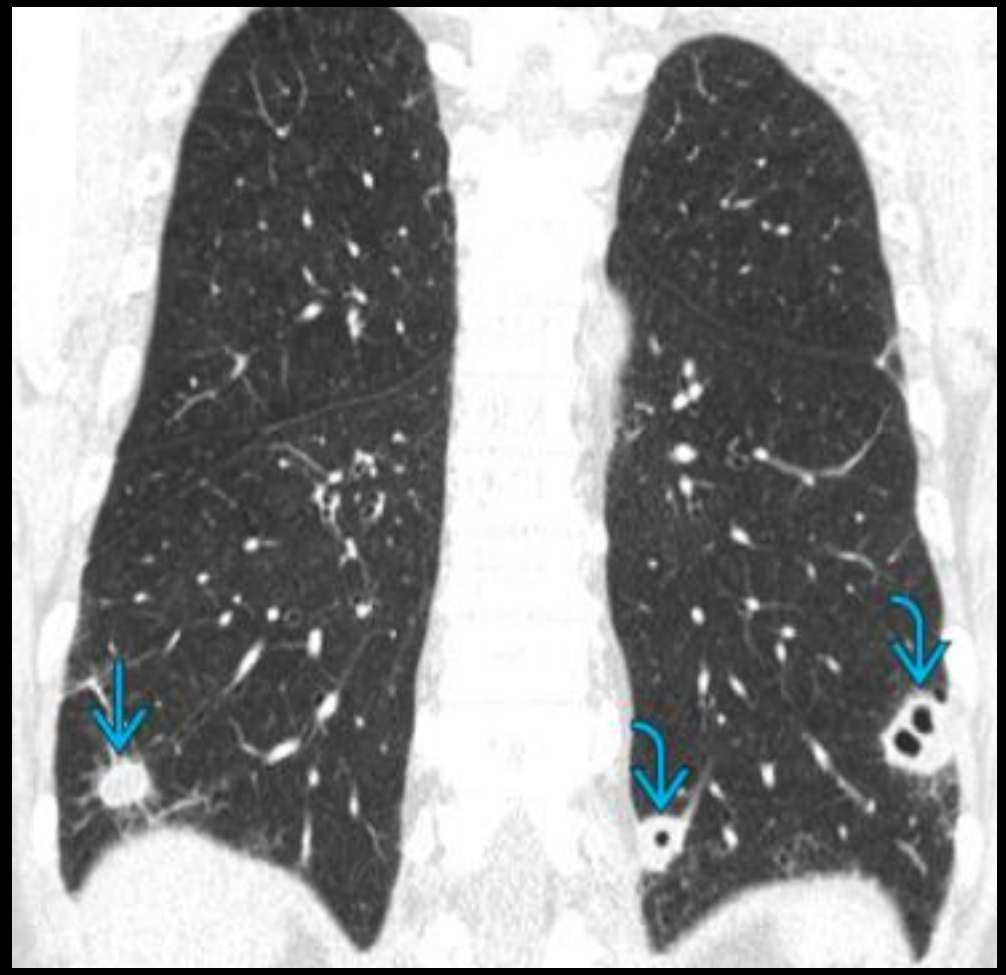
Essential cryoglobulinemia
Microscopic polyangiitis
GPA (Wegener)
Pulmonary-renal syndrome (Goodpasture)



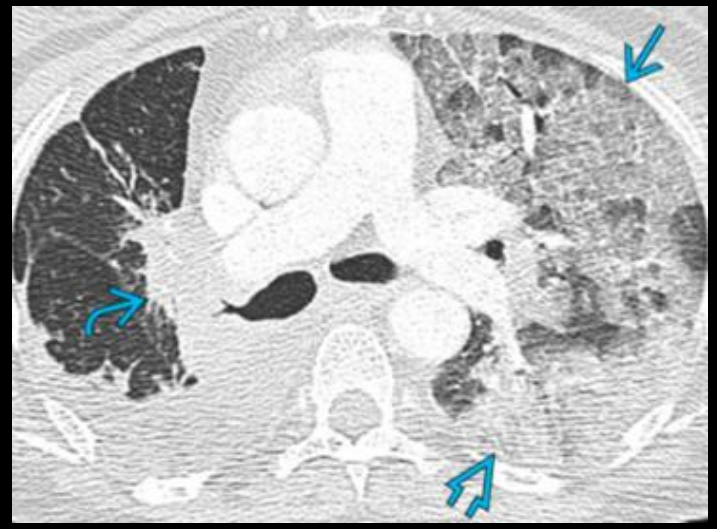
- Diffuse ground glass (alveolar hemorrhage)
- Solid and/or cavitary nodules

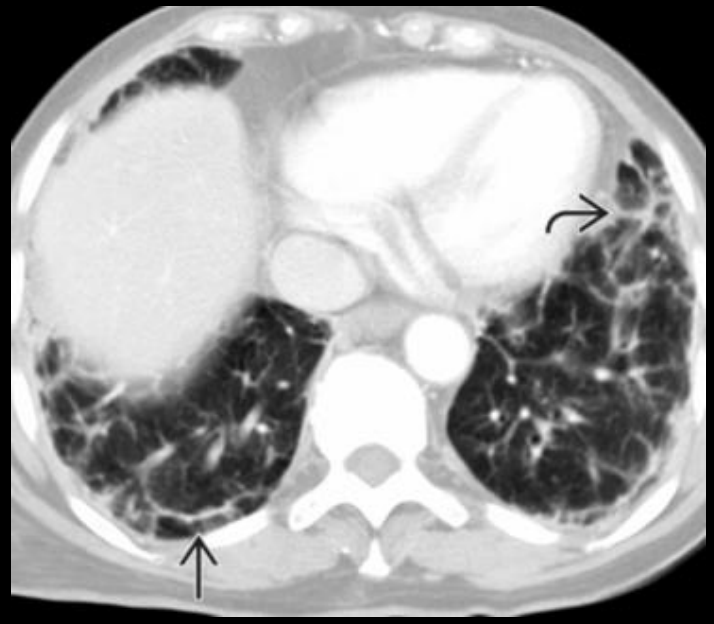
Systemic Inflammation

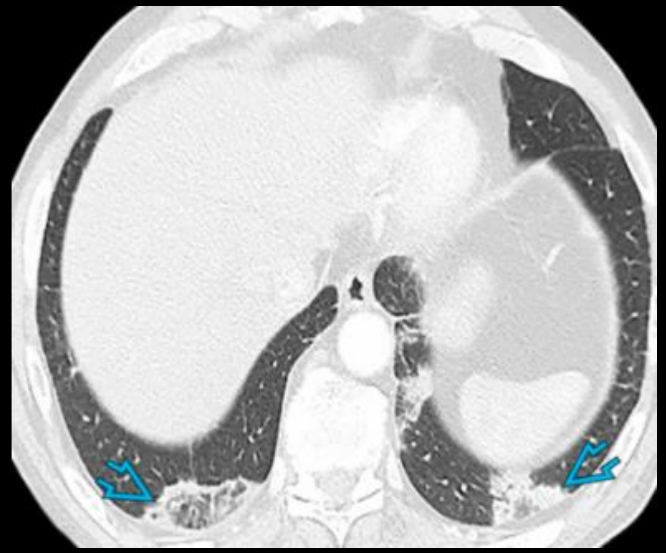
Drug Toxicity

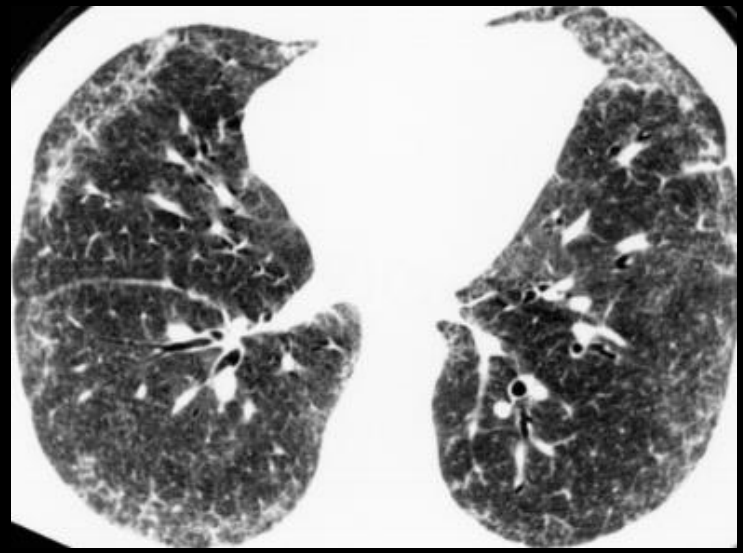
Dyspnea, cough, fever, eosinophilia
associated with multiple drug classes

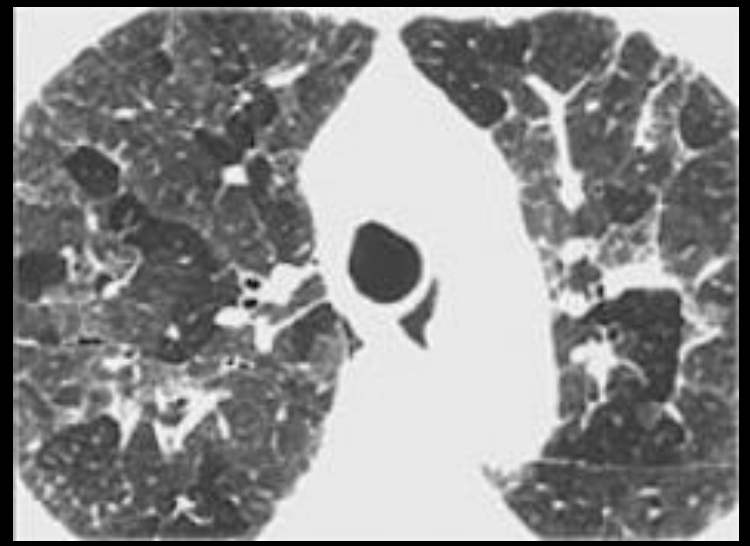
UIP, NSIP, OP, DAD, HP, EP, vasculitis, DAH





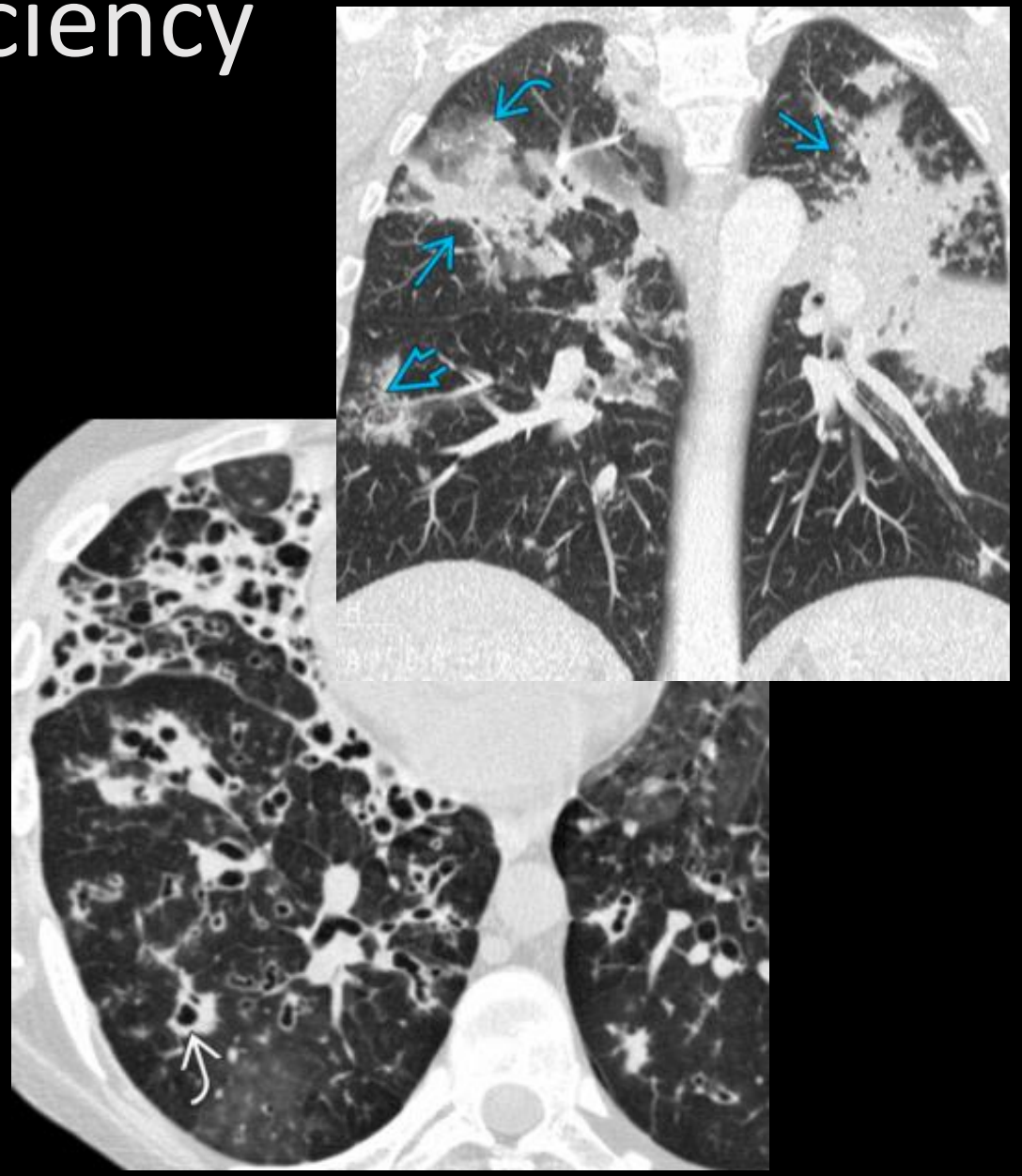






Immunodeficiency

Immunodeficiency		
B-Cell Deficiencies		**T-Cell Deficiencies**
Recurrent bacterial infections		*Severe viral, fungal, and protozoal infections*
Bruton's agammaglobulinemia—defect in B-cell development Common variable–hypogammaglobulinemia—defect in plasma cell differentiation Hyper-IgM syndrome—defect in class switching	**B and T-cell deficiencies** Severe combined immunodeficiency	Bare lymphocyte syndrome—lack of class II MHC Omenn's syndrome—defect in TCR gene rearrangement DiGeorge syndrome—thymic aplasia
Phagocytic Cell Deficiencies		**Complement Deficiencies**
Recurrent bacterial infections		*Recurrent bacterial infections* *Defects in immunocomplex clearance*
Chronic granulomatous disease—lack of respiratory burst Leukocyte adhesion deficiency—lack of PMN extravasation into tissue Chediak-Higashi syndrome—defect in neutrophil microtubule function and related phagosome/lysosome fusion		C1, C2, or C4 deficiency—defects in clearing immunocomplexes C3 or C5 deficiency—block in alternative and classical pathways C6, C7, C8, or C9—defect in MAC assembly and function



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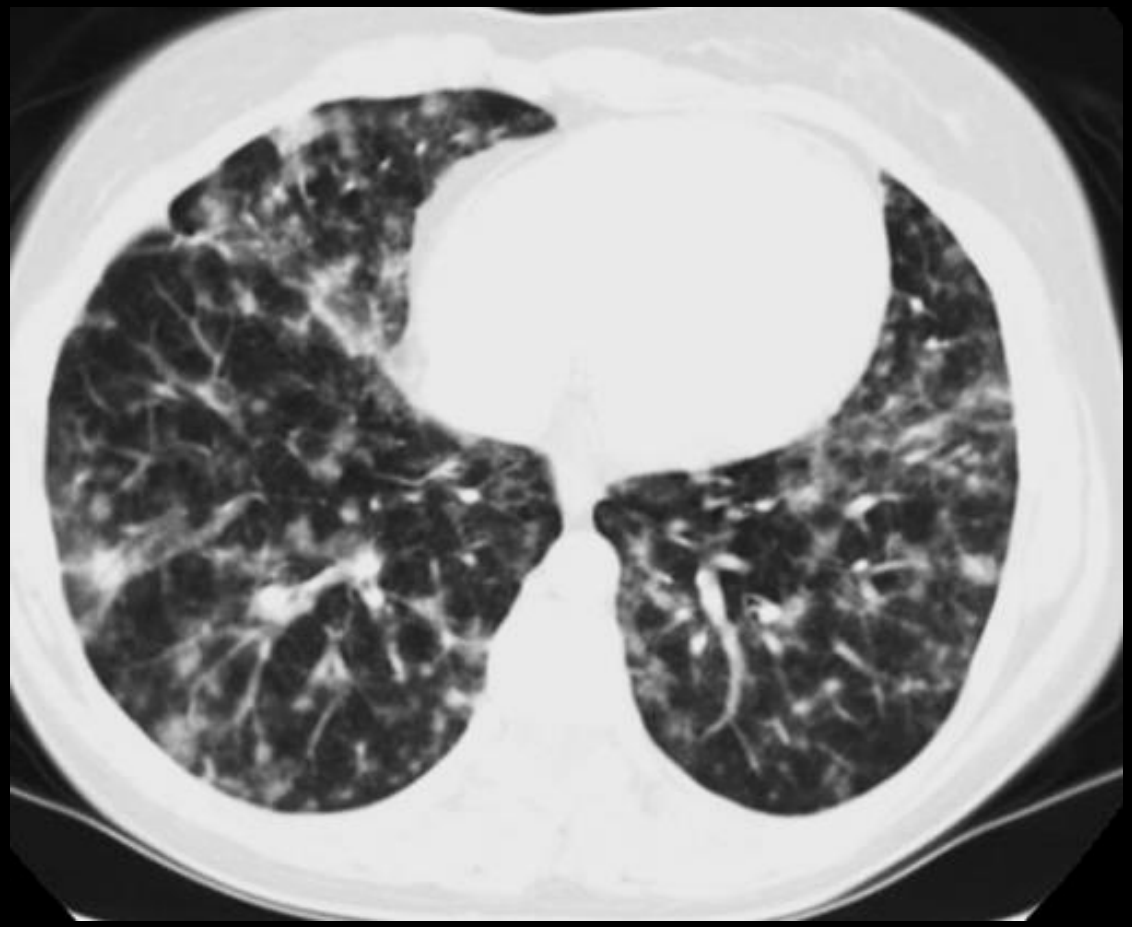
Granulomatous Lymphocytic ILD



- Sarcoid mimic but LLZ predominant
- Usually accompanied by splenomegaly

Metabolic, Deposition, & Storage

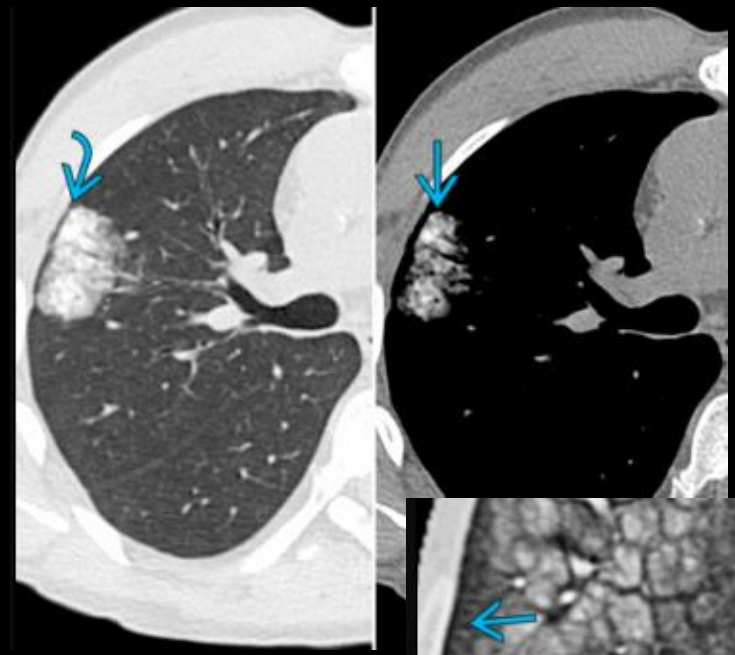
- Metastatic calcification/ossification
- Amyloidosis
- Light chain deposition disease
- Storage diseases

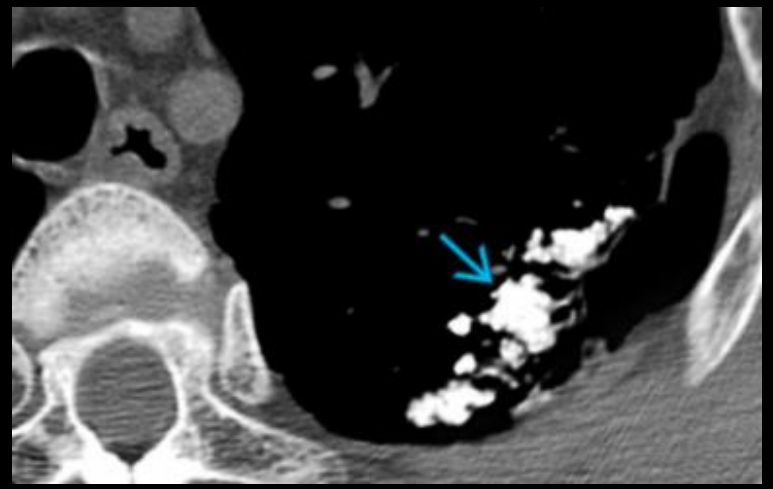
Metabolic, Deposition, & Storage

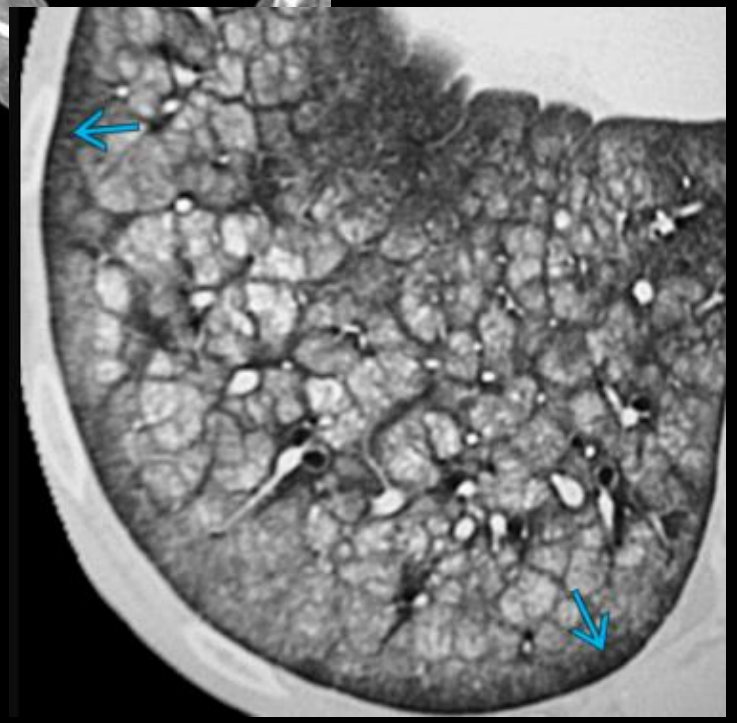
Metastatic calcification/ossification

Associated with chronic renal failure, hypercalciemia, and hyperalkalinity

- Calcified nodules/masses
- Centrilobular calcific “rosettes”







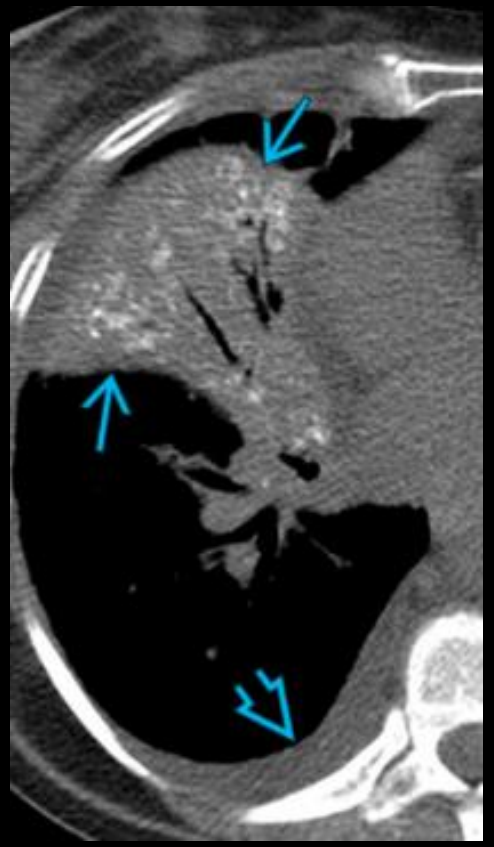
Metabolic, Deposition, & Storage

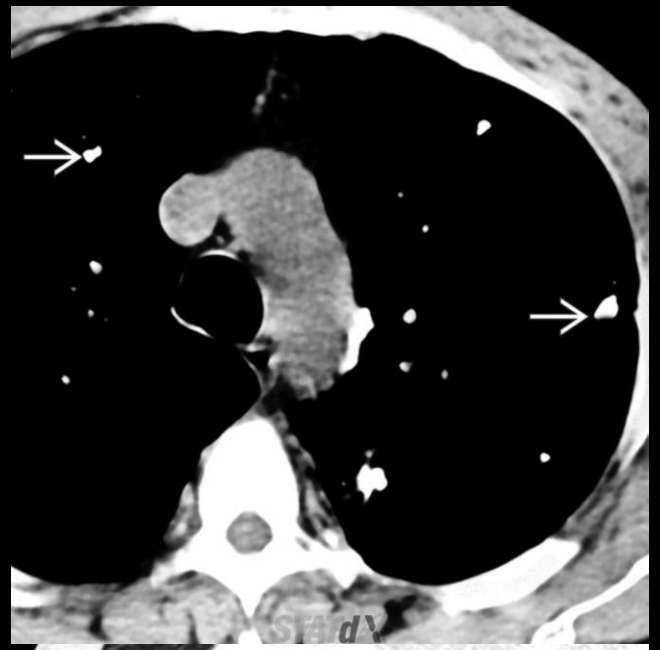
Amyloidosis

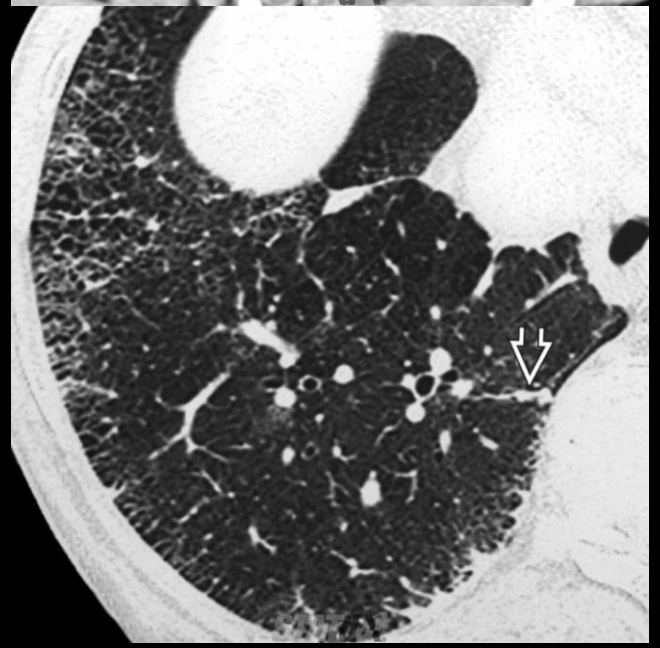
Symptoms range from asymptomatic to progressive dyspnea

Deposition of insoluble proteins

Protein deposits: Combination of serum amyloid P, glycosaminoglycans, and fibril proteins



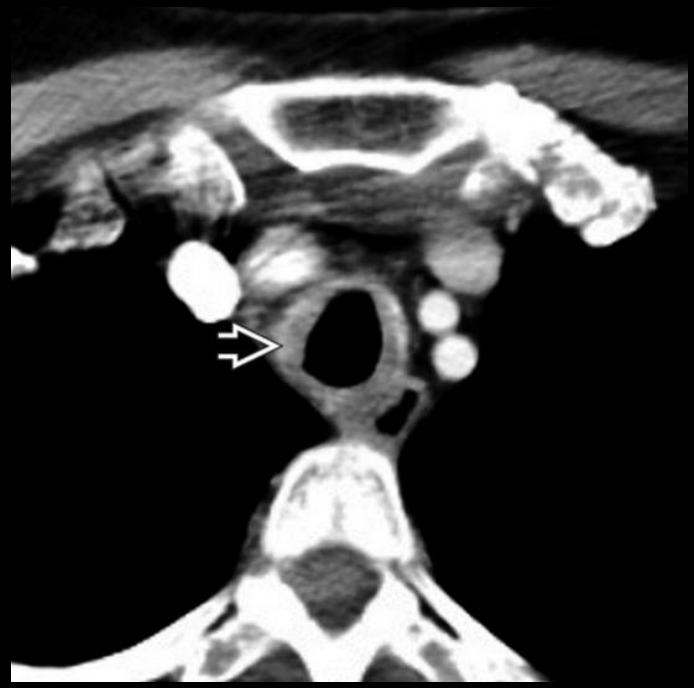


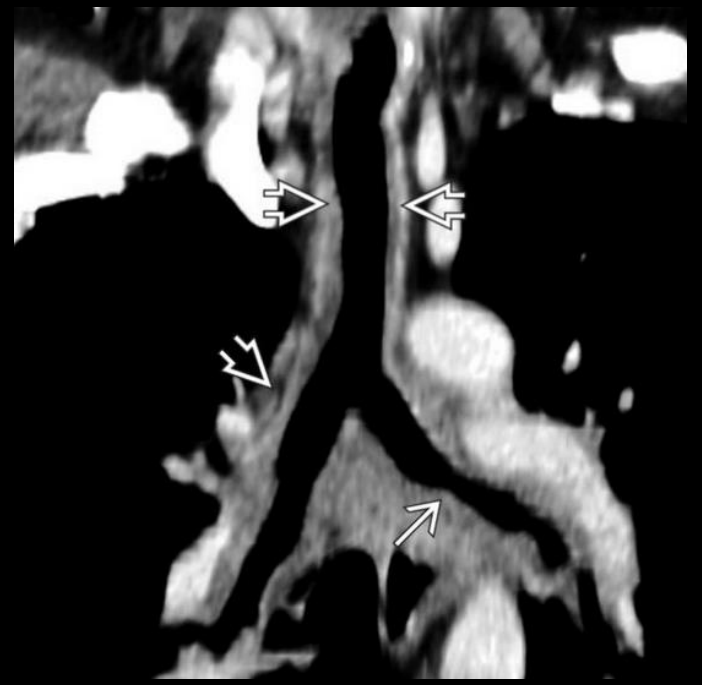


Consolidative, nodular, and interstitial presentations

Metabolic, Deposition, & Storage

Amyloidosis





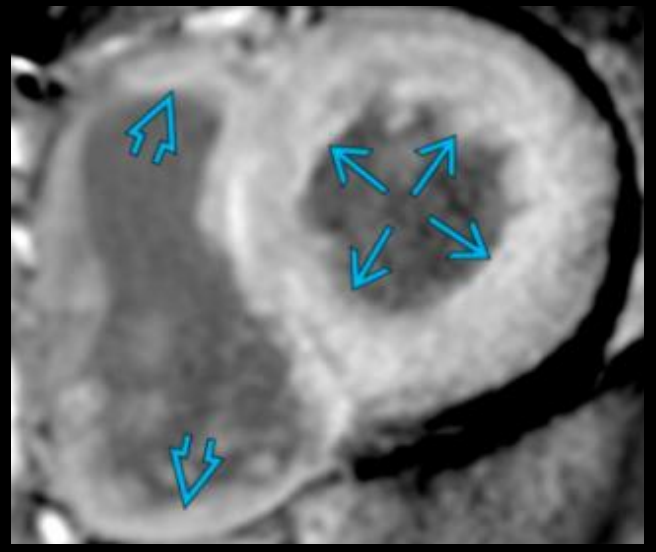
Deposition of insoluble proteins

Protein deposits: Combination of serum amyloid P, glycosaminoglycans, and fibril proteins

May also effect airways and myocardium

Wheezing/stridor

Diastolic dysfunction



Metabolic, Deposition, & Storage

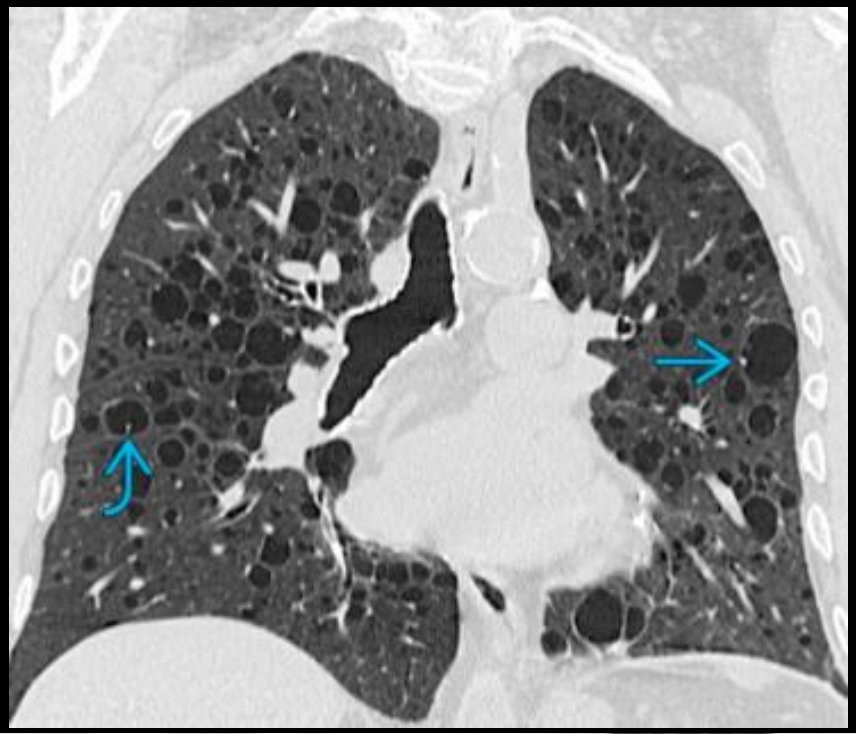
Light chain deposition disease

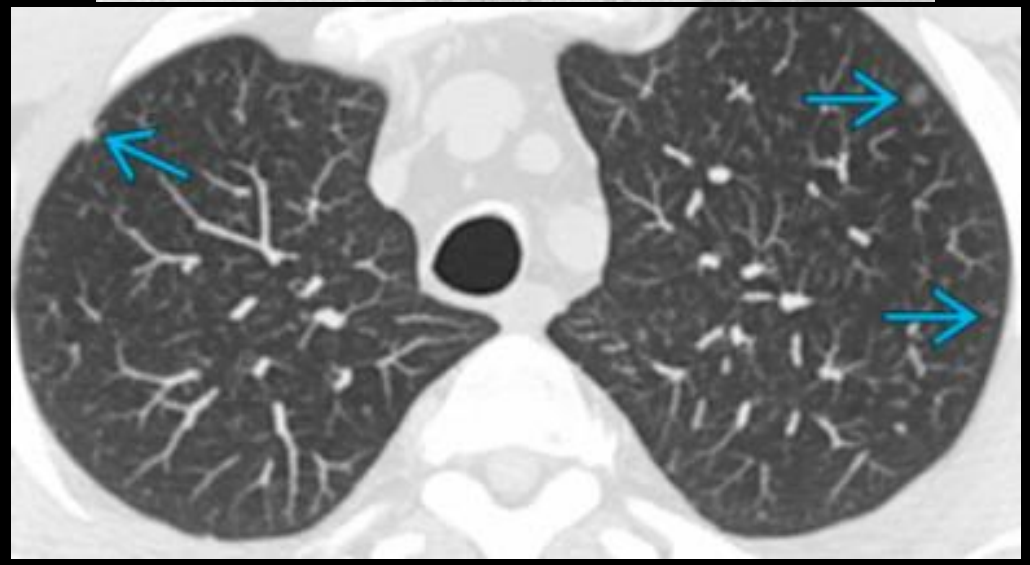
Symptoms range from asymptomatic to progressive dyspnea.

Systemic extracellular accumulation of immunoglobulin light chains due to underlying plasma cell dyscrasia

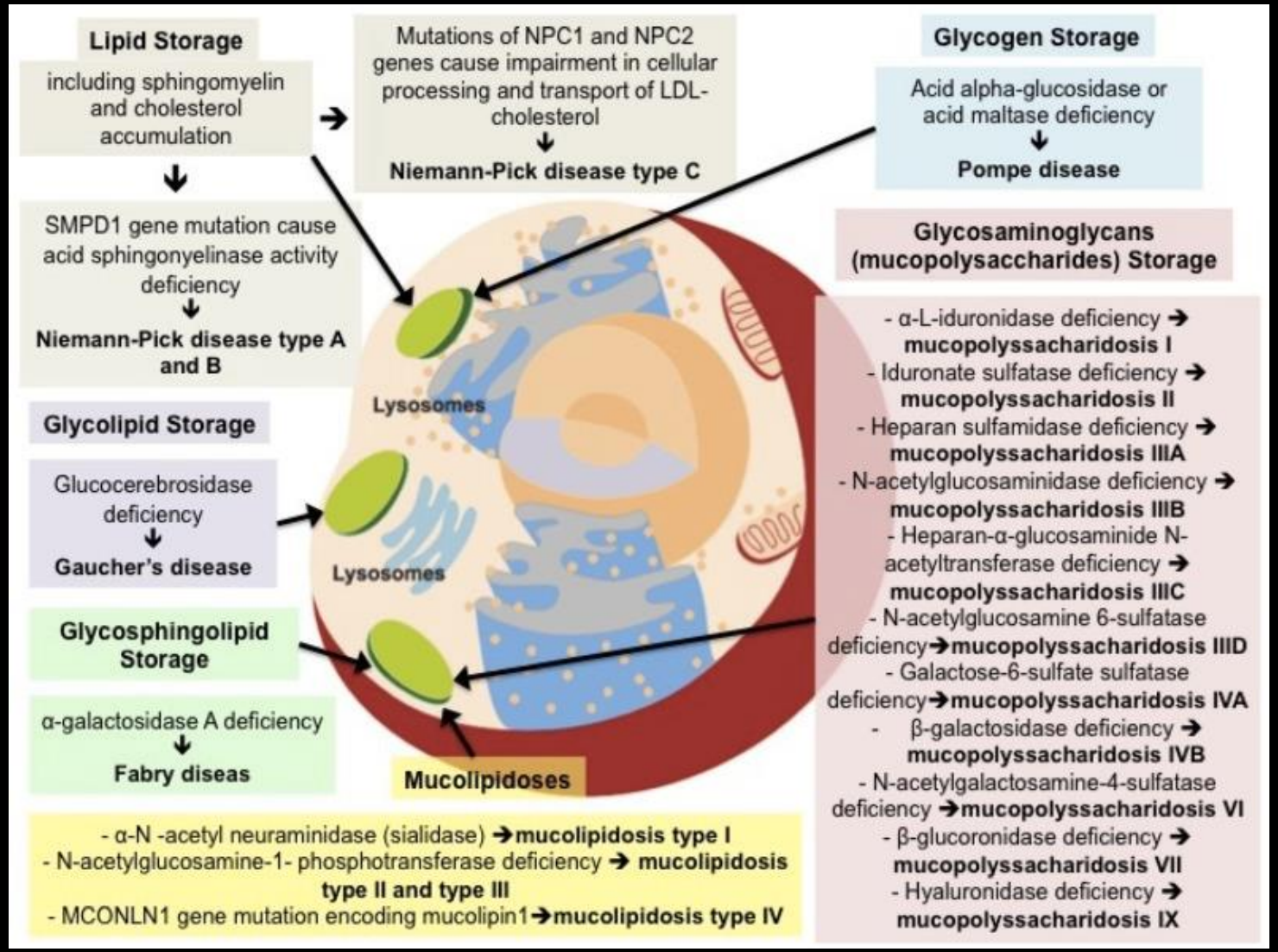
- Deposition of amorphous nonfibrillary material; does not contain amyloid fibrils
- Does not have β-pleated sheet configuration and consequently does not bind Congo red, unlike amyloidosis

Thin walled cysts and/or small nodules

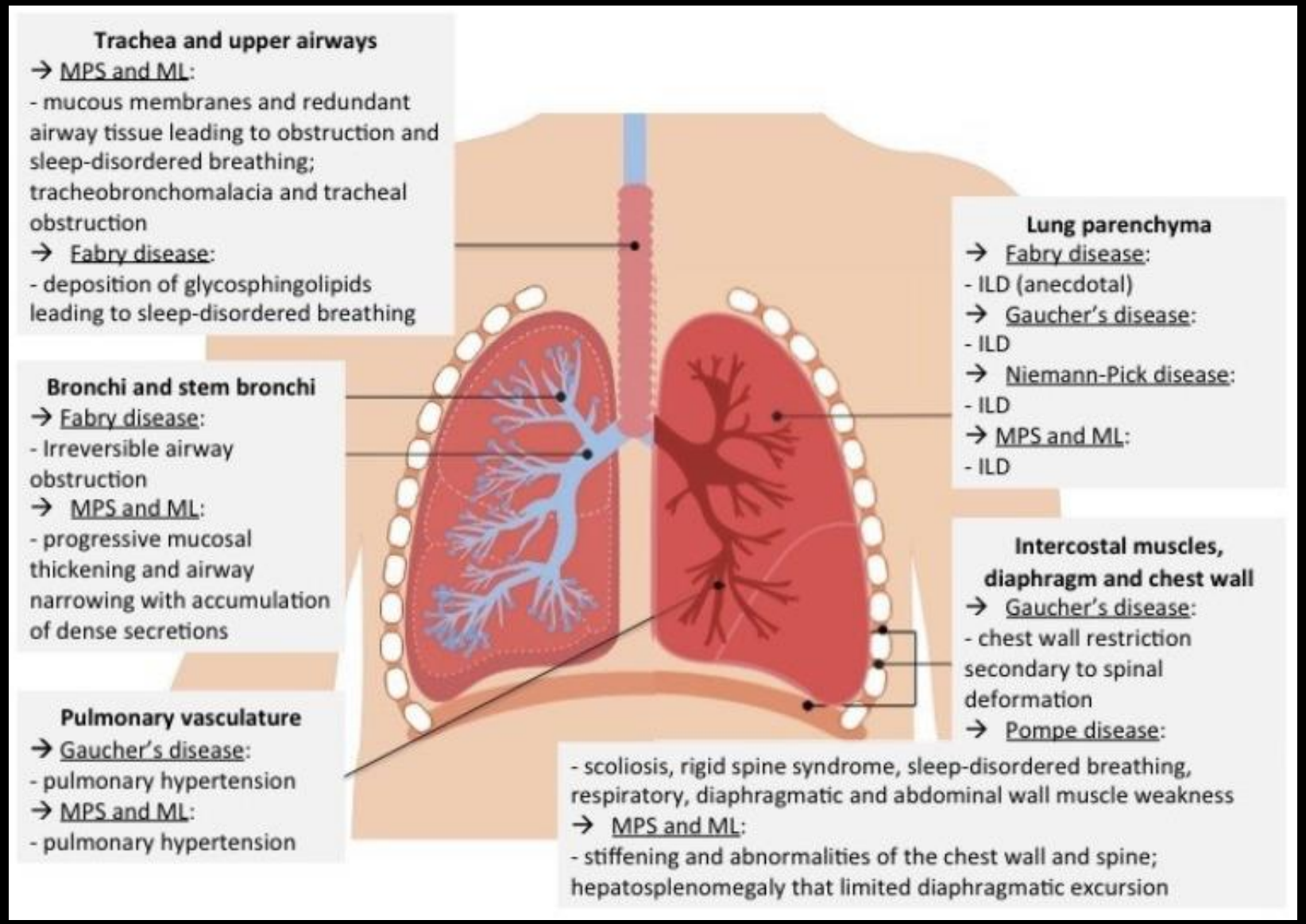




Storage Diseases



Storage Diseases



Ciliary and Mucoid

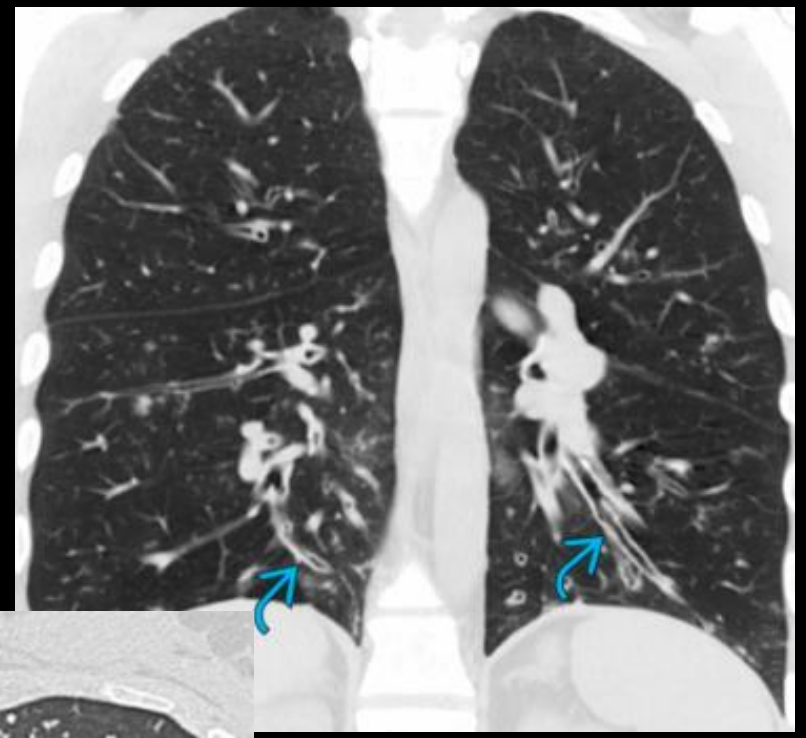
- Ciliary dyskinesia syndromes
- Cystic Fibrosis

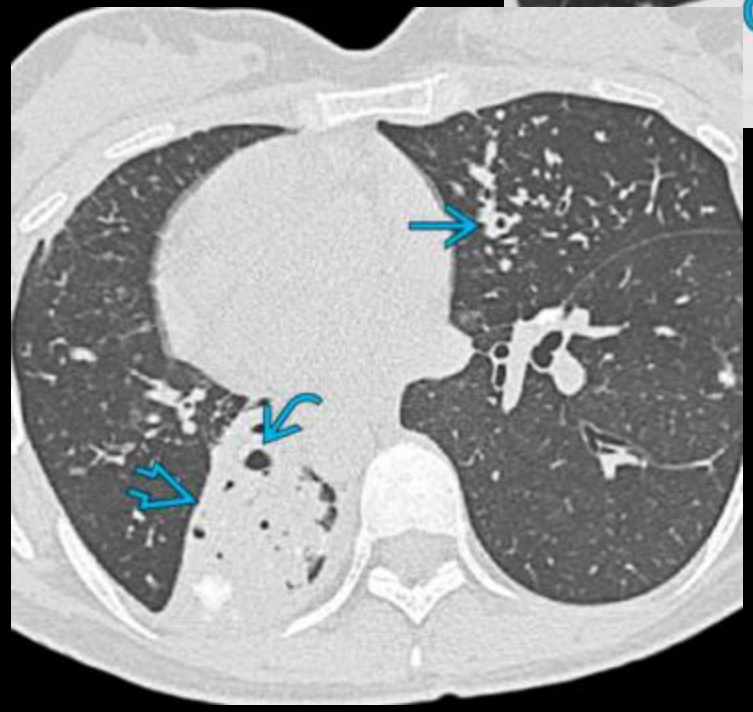
Ciliary and Mucoid

Primary ciliary dyskinesia

- Chronic/recurrent rhinitis, secretory otitis media, sinusitis
- Recurrent lower respiratory infection
- Infertility in men, lowered fertility and ectopic pregnancy in women
- Situs abnormalities with Kartagener syndrome

Bronchitis, bronchiectasis, mucous plugging, air trapping



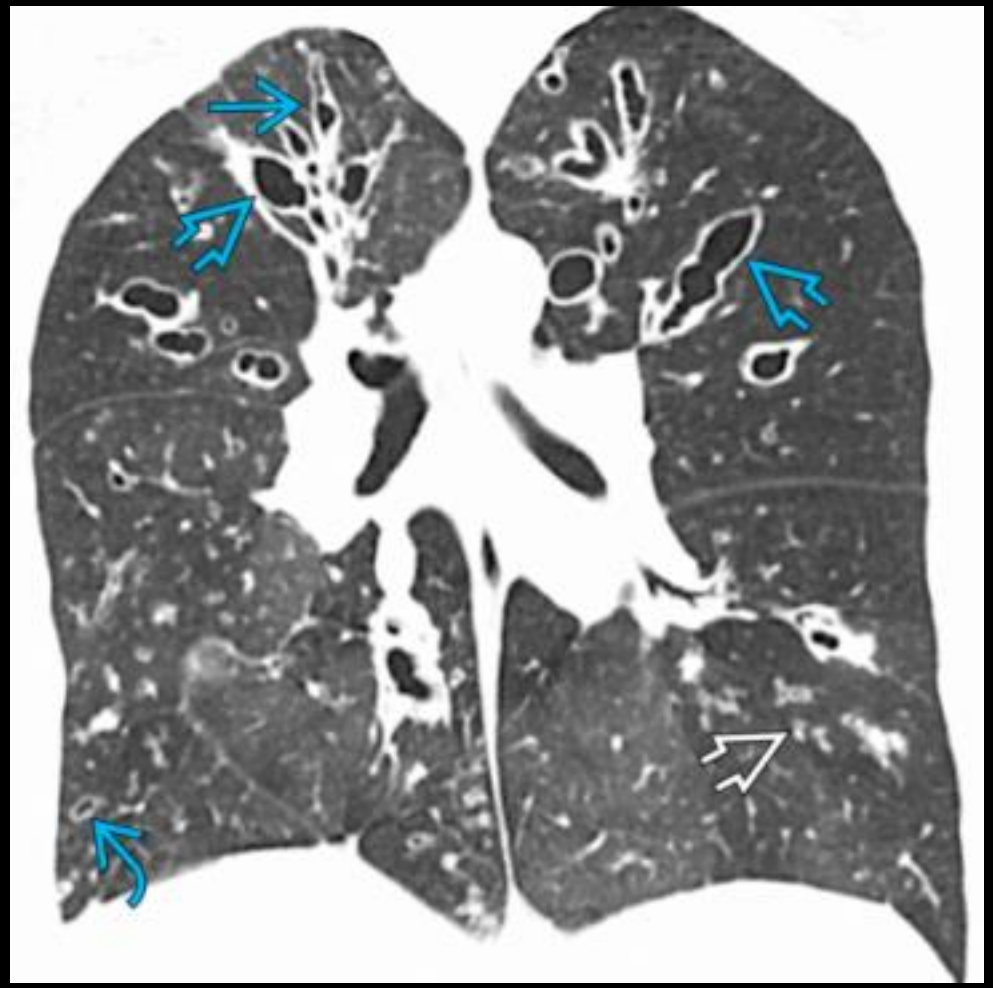


Ciliary and Mucoid

Cystic fibrosis

Autosomal recessive disorder that affects regulation of chloride transport thickened secretions

Bronchitis, bronchiectasis, mucous plugging, air trapping



Hereditary & Developmental

- LAM
- TS
- NF
- BHD
- HHT
- Lymphangiomatosis

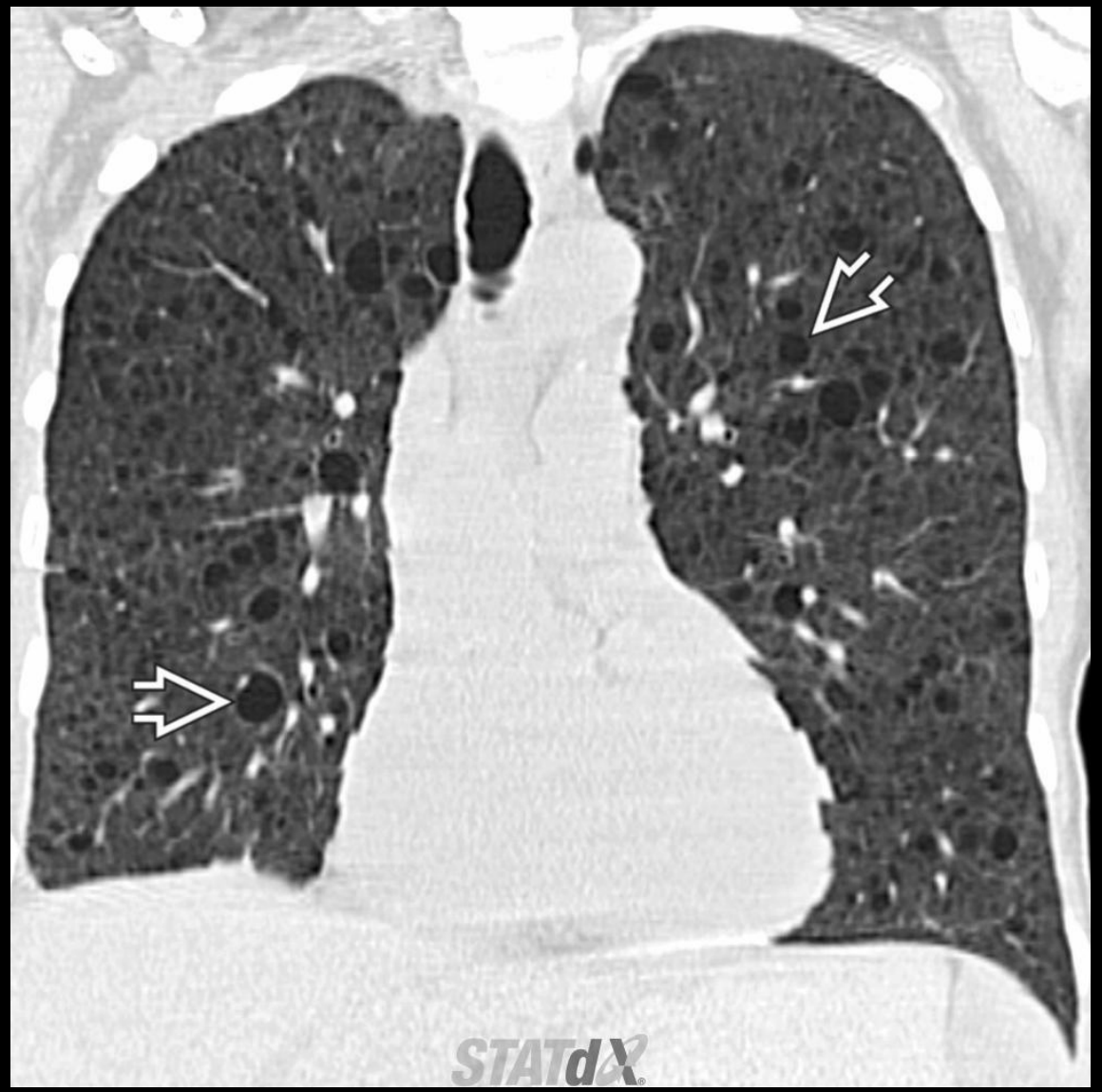
Hereditary

Lymphangioleiomyomatosis

Progressive dyspnea; spontaneous pneumothorax; females only

Pathologic smooth muscle (LAM) cell hyperplasia affecting lymphatic channels. Angiomyolipomas.

- Diffuse, thin walled cysts
- Chylous effusions



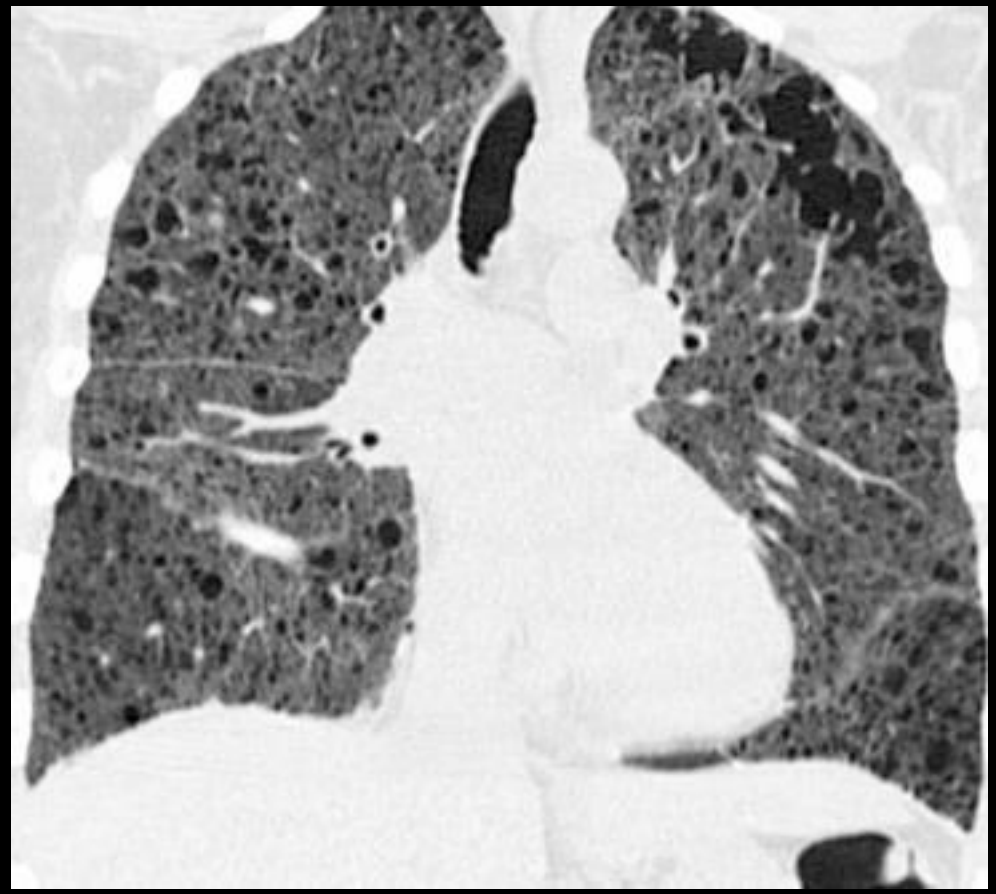
Hereditary

Tuberous Sclerosis

Multisystem autosomal dominant hereditary neurocutaneous disorder characterized by multifocal hamartomas and benign and malignant neoplasms; males and females

In the lungs, manifests as LAM cell proliferation

Imaging is identical to LAM



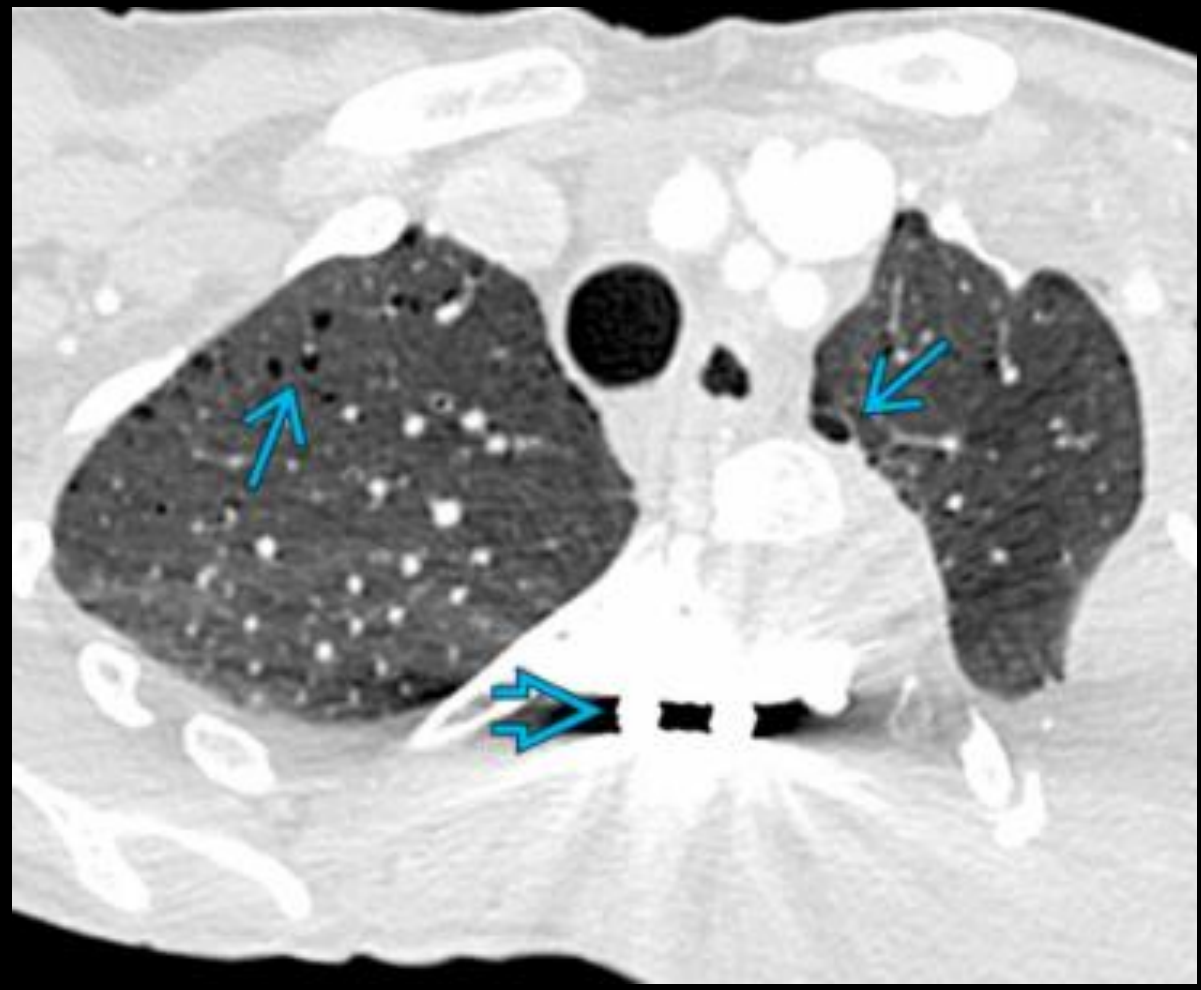
Hereditary

Neurofibromatosis

Multiple nerve sheath neoplasms (dermal, cutaneous, intraforaminal neurofibromas or diffuse plexiform neurofibromas)

Mutations in neurofibromin gene

Nodules (neurofibromas) and thin-walled cysts



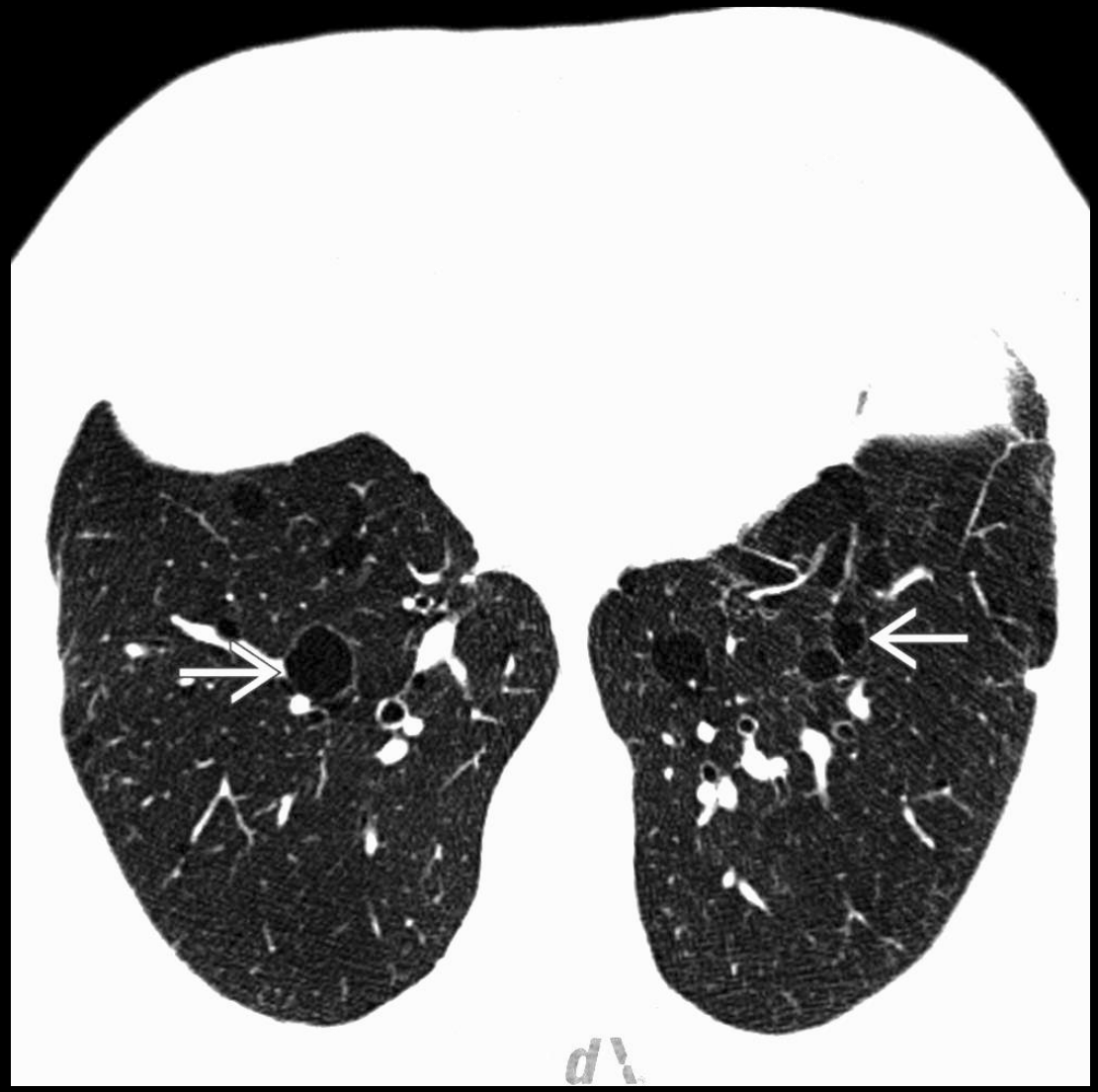
Hereditary

Birt Hogg Dube

RCC, hair follicle folliculofibromas, pulmonary cysts, spontaneous pneumothorax

AD inheritance of deletion of FLCN gene from Chromosome 17

Diffuse, thin-walled cysts



Hereditary

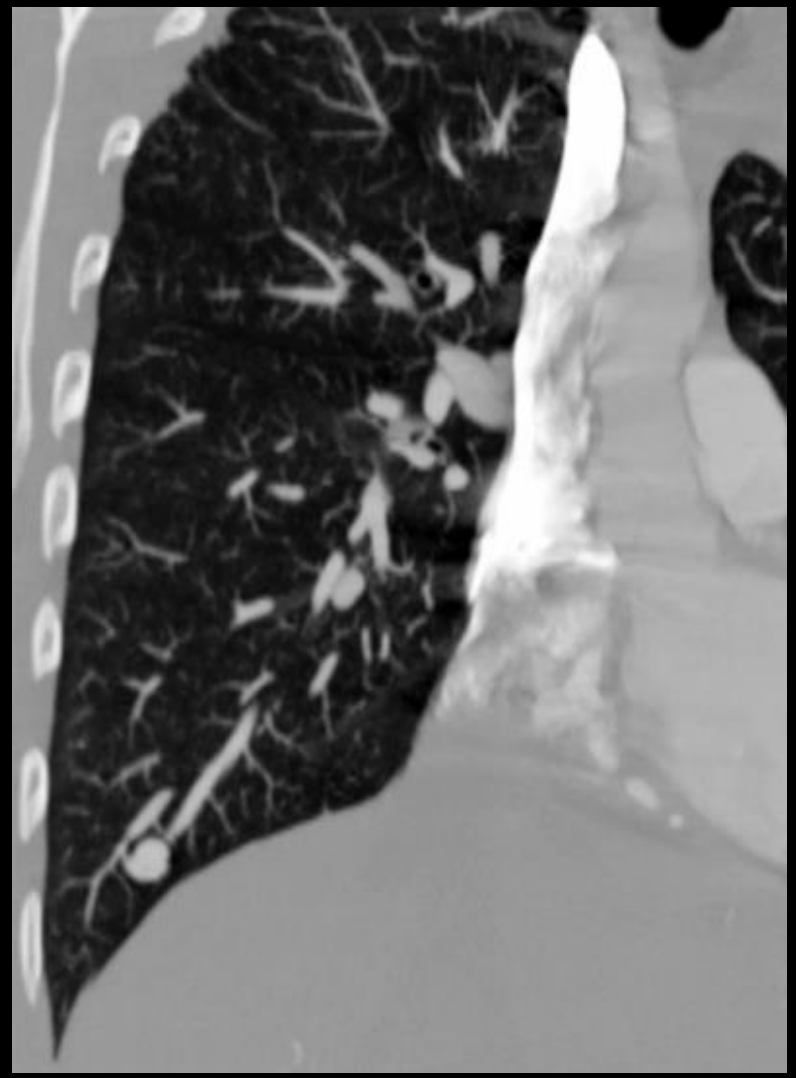
Hereditary Hemorrhagic Telangiectasia

Mucocutaneous telangiectasias with multiorgan involvement

- Nasal mucosa: Recurrent epistaxis
- CNS (cerebral or spinal AVM): Seizures, paraparesis, subarachnoid hemorrhage
- Gastrointestinal: GI bleed and angiodysplasias
- Pulmonary: Cyanosis, polycythemia, dyspnea, hemoptysis

Genetically heterogeneous disorder caused by mutations in TGF-β/BMP signaling pathway

AVMs



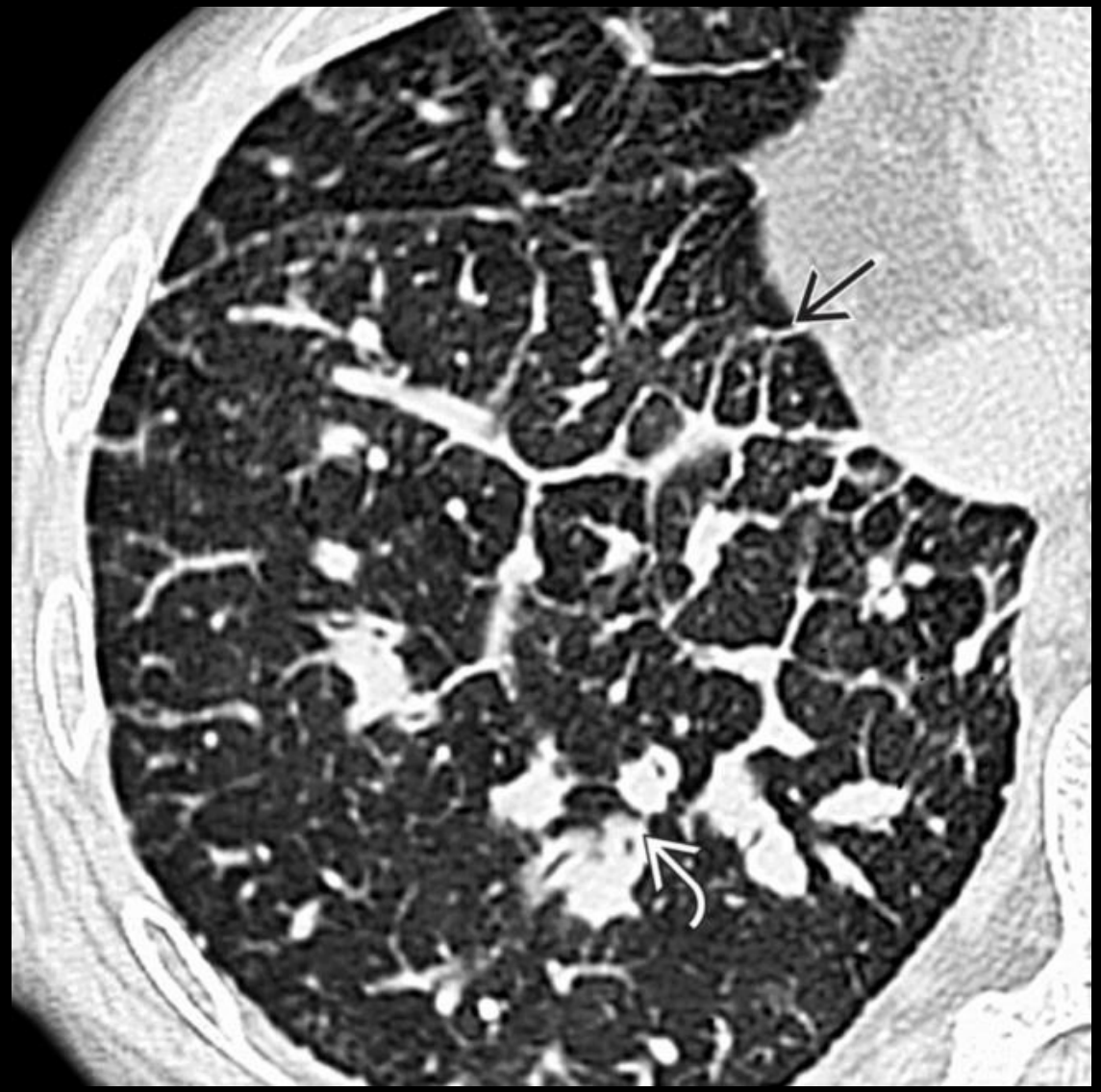
Developmental

Lymphangiomatosis

Progressive, often fatal disease affecting children and young adults presenting with dyspnea, wheezing, hemoptysis, chyloptysis, bronchial casts

Congenital lymphatic malformation with increased diameter, number, and complexity of lymphatic channels.

Lymphatic distribution thickening and chylous effusions

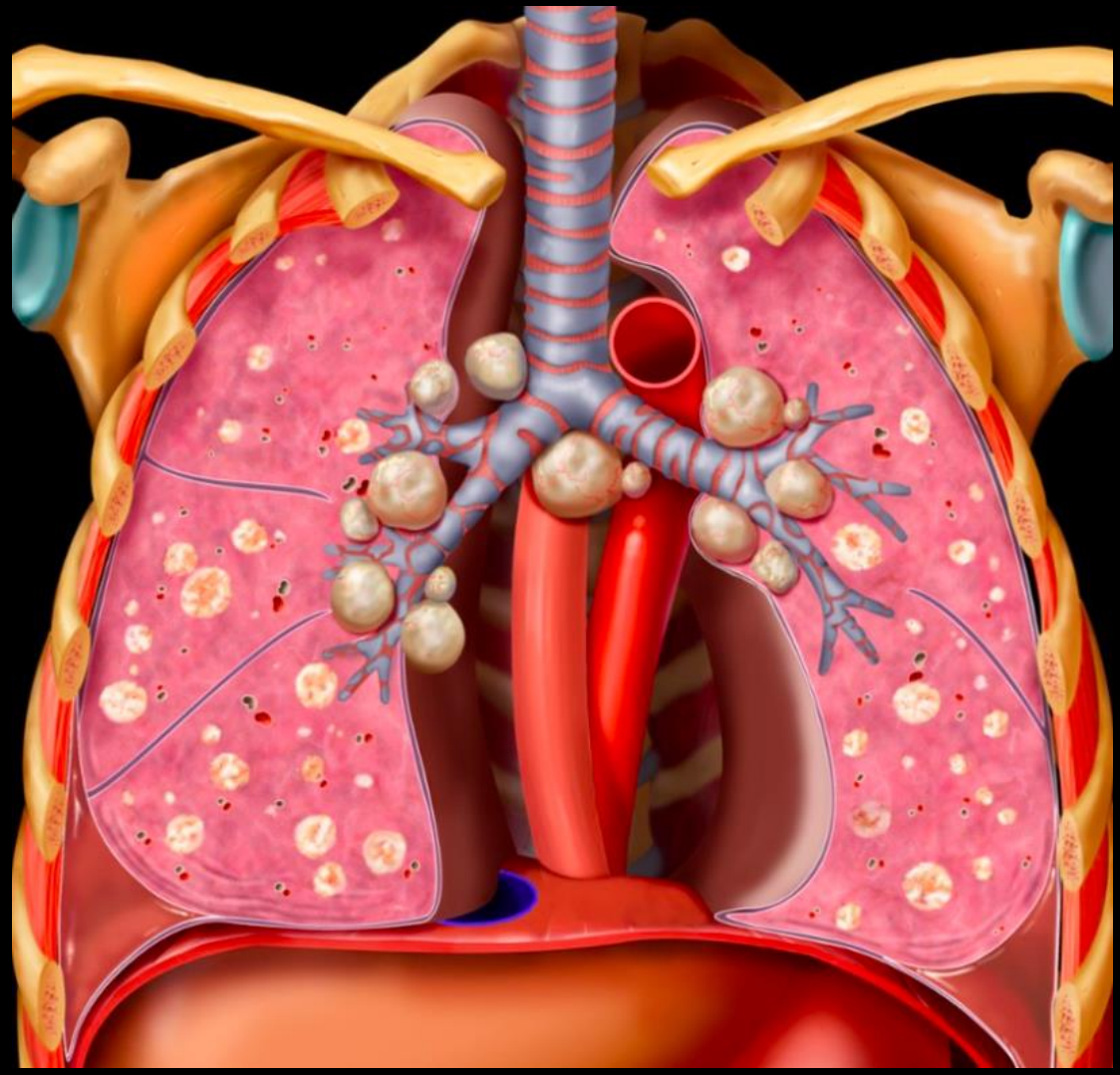


Systemic Neoplasm

- Mets
- Kaposi sarcoma
- Erdheim-Chester
- Lymphangiomatosis
- Lymphoma & lymphoproliferative
- Leukemia

Systemic Neoplasm

Metastatic Disease



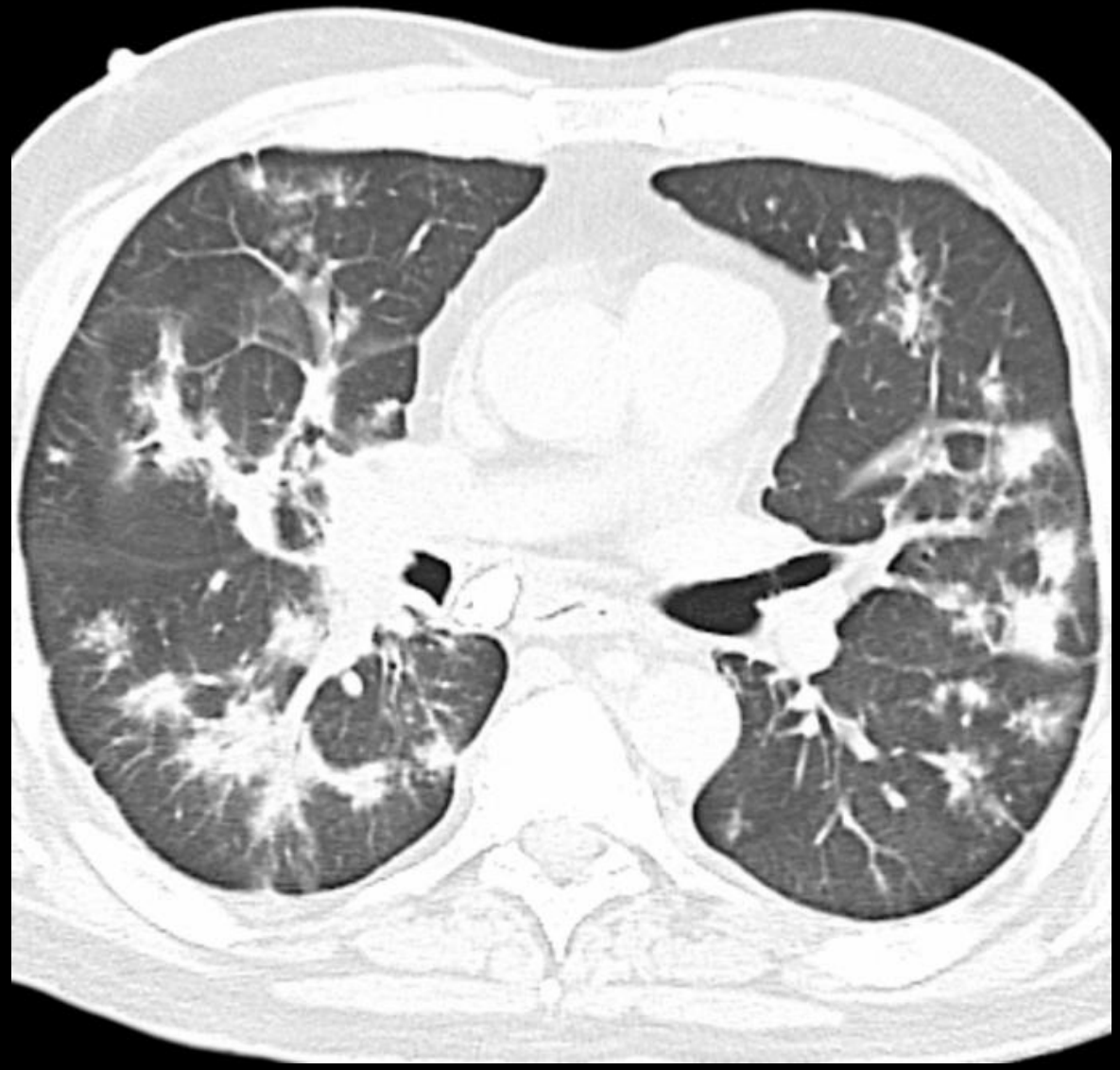
Systemic Neoplasm

Kaposi Sarcoma

HHV-8-associated malignancy in the severely immunocompromised

Spindle cell neoplasm with leaky hypervascularity

Flame-shaped and coalescent perivascular opacities +/- lymphatic distribution thickening



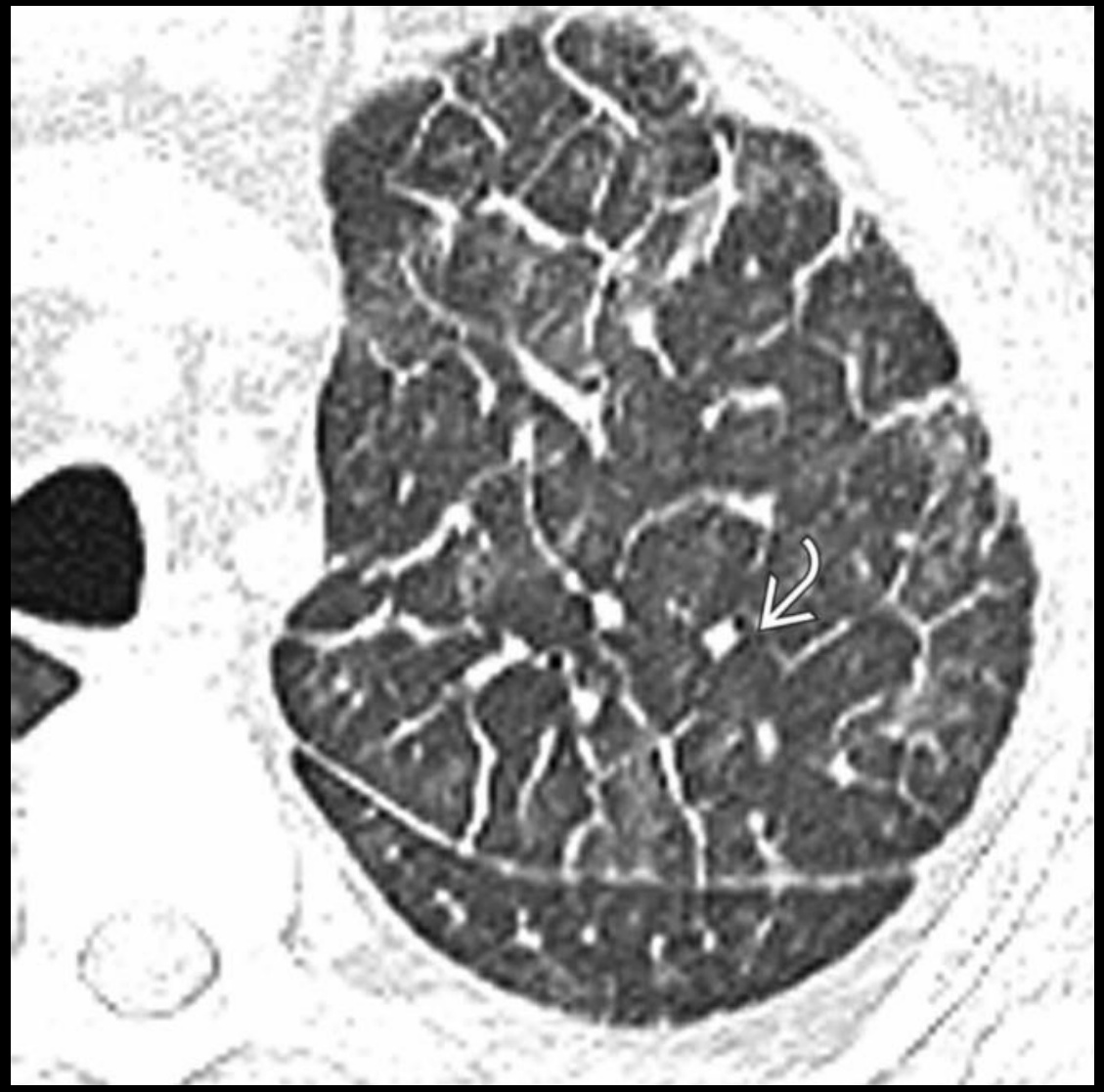
Systemic Neoplasm

Erdheim-Chester

Slowly-progressive low grade neoplasm most commonly presenting with dyspnea

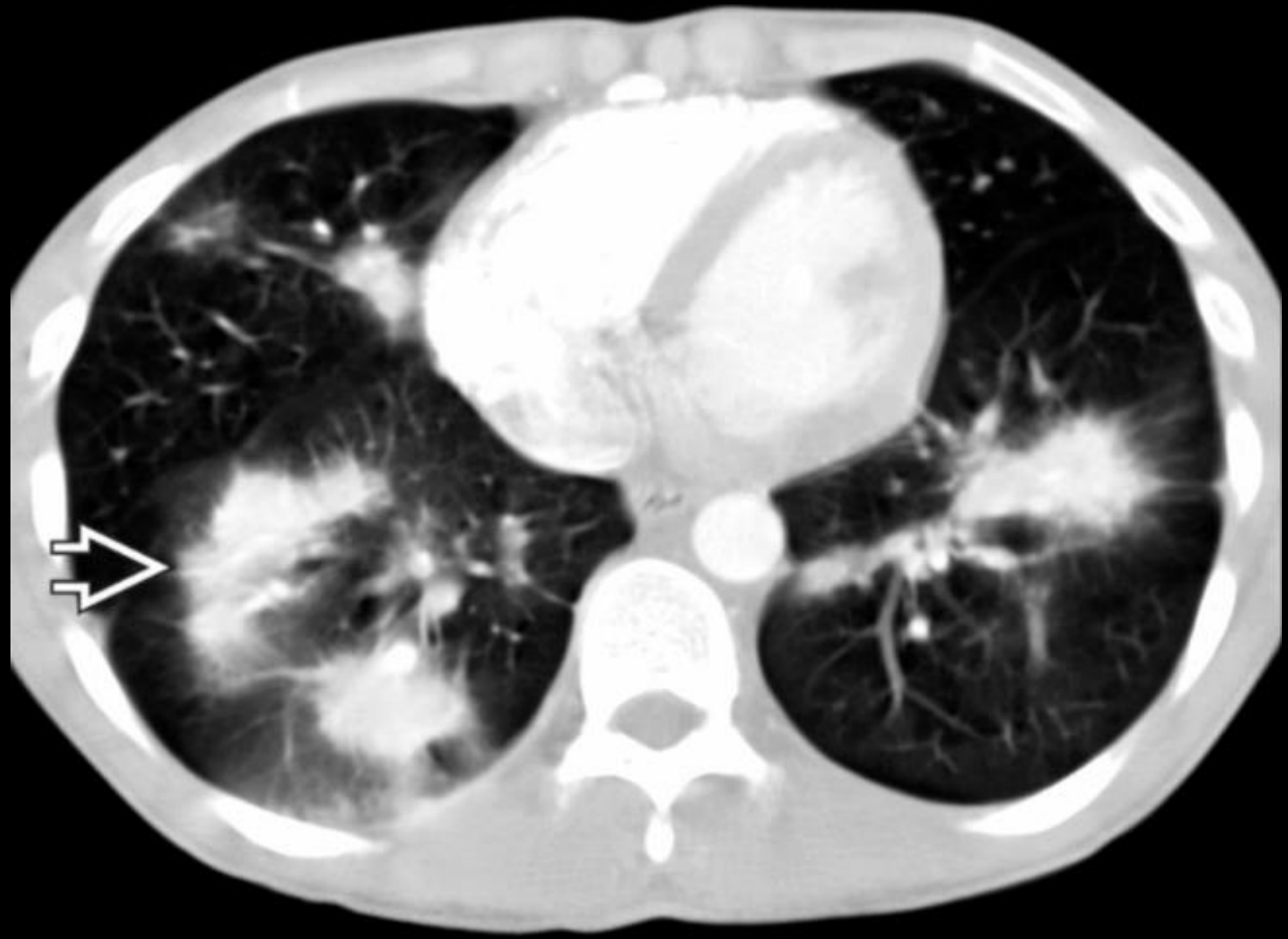
Proliferation of non-Langerhans histiocytes, lymphocytes, and plasma cells leads to lymphatic expansion, obstruction, and fibrosis

Diffuse septal thickening, pleural effusion, perirenal soft tissue encasement, and sclerotic bone lesions



Systemic Neoplasm

Lymphoma & Lymphoproliferative disorders

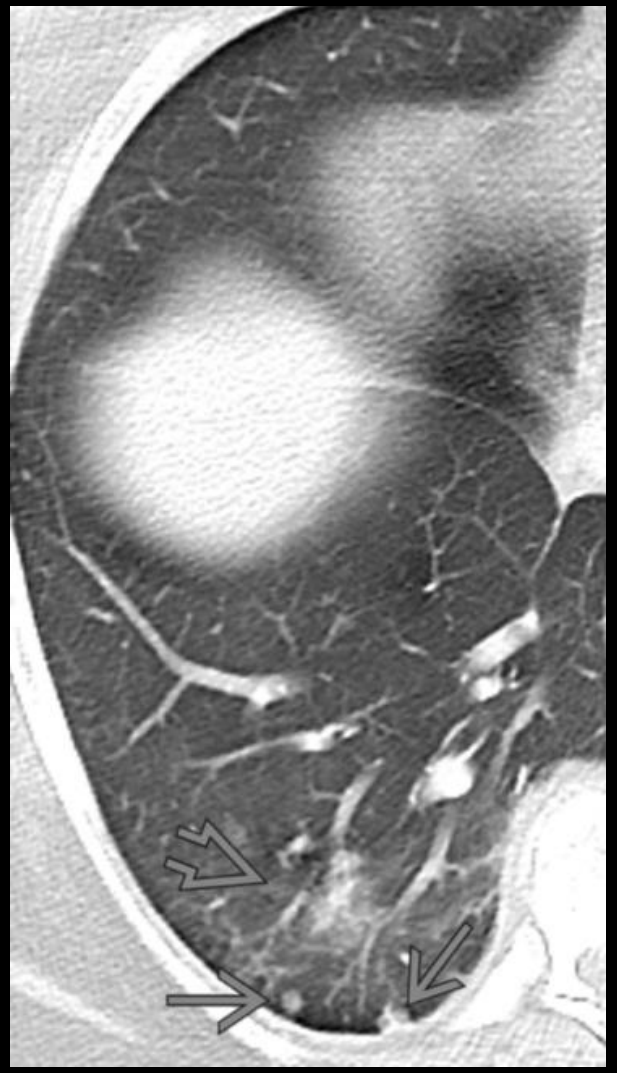


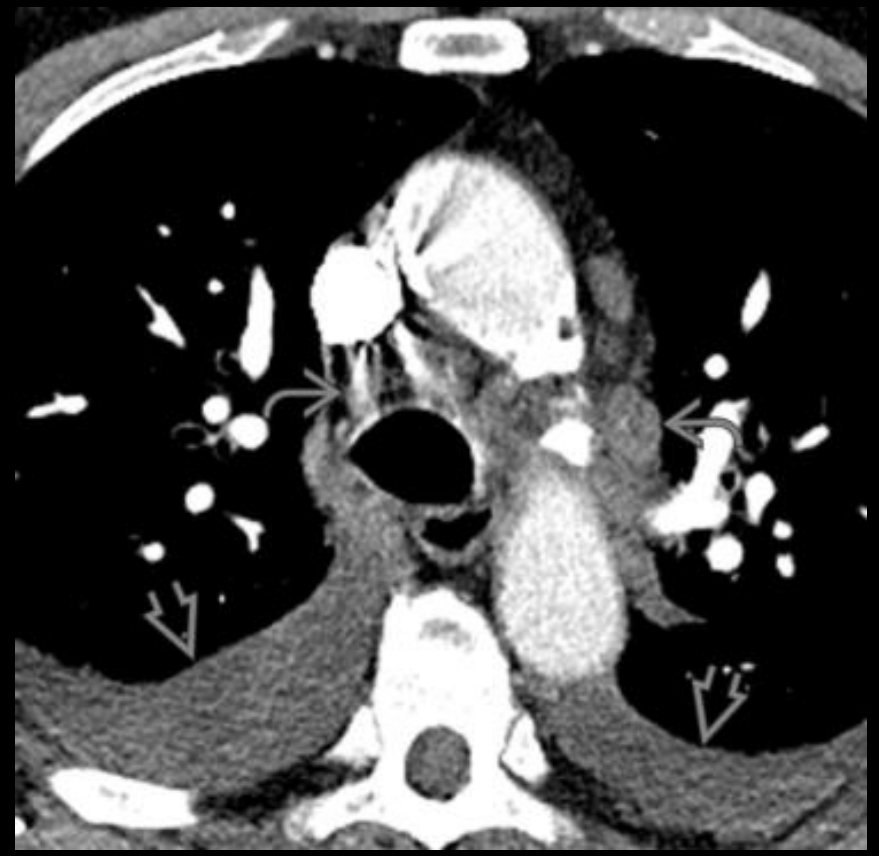
Chronic nodular and/or infiltrative airspace disease

Systemic Neoplasm

Leukemia

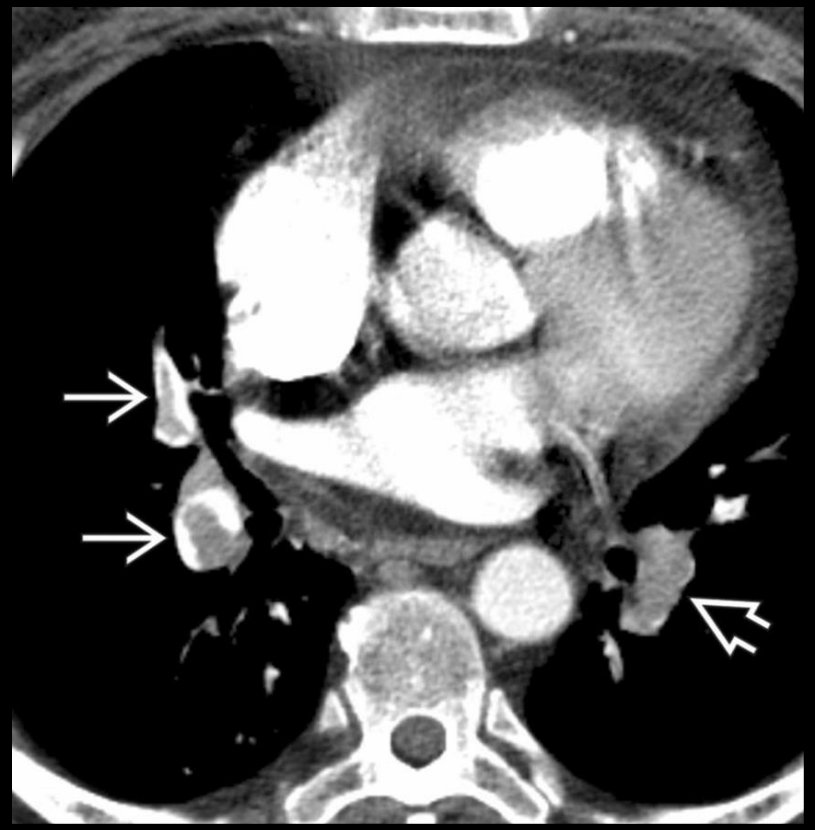
Lymphadenopathy, effusions, nodules

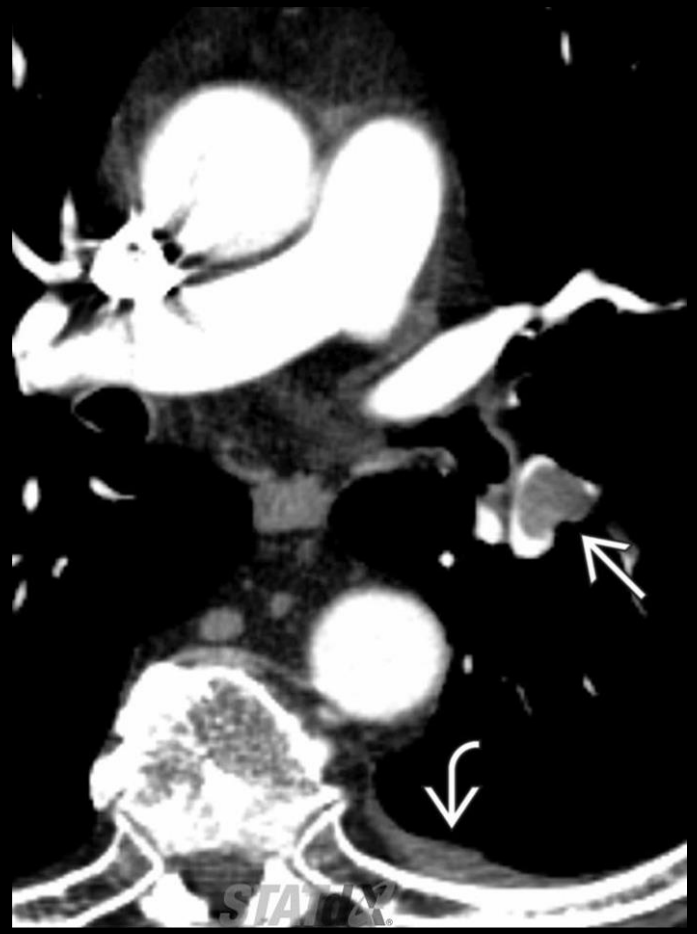


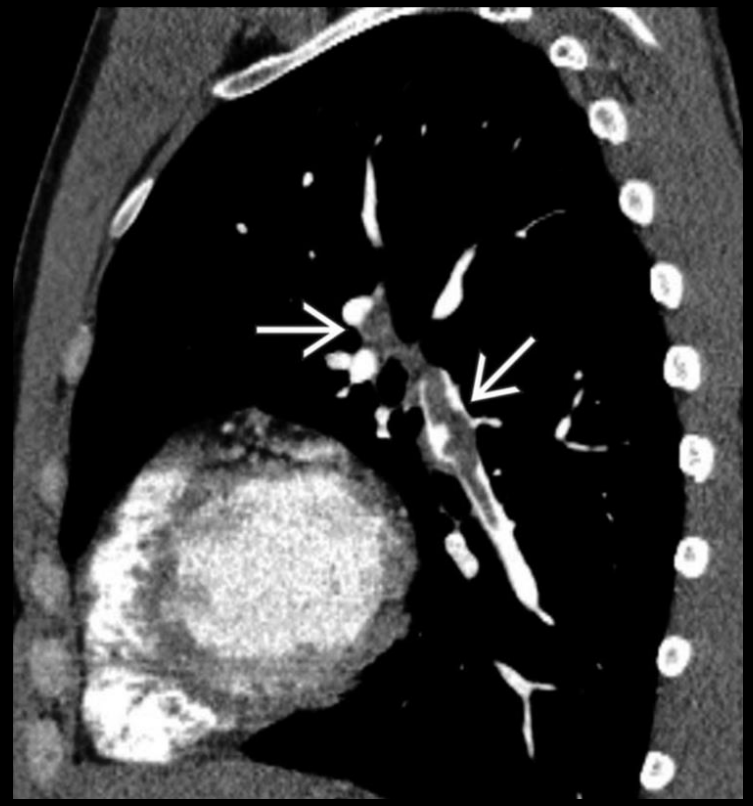


Hypercoagulable States

Pulmonary embolism

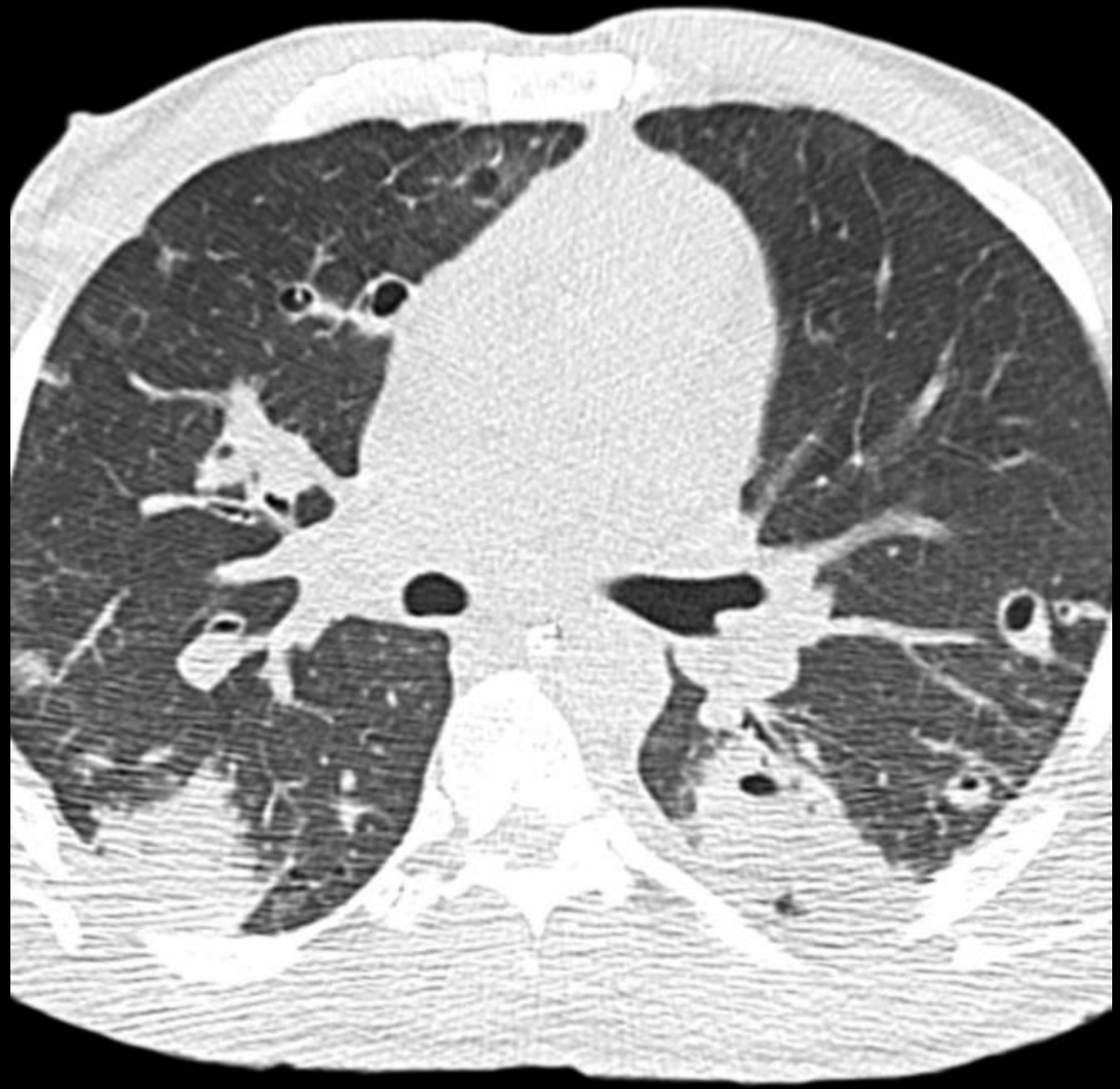






Systemic Infection

Septic embolism



Scattered (rapidly cavitating) nodules and consolidation

References

1. Most images are courtesy of StatDx (Elsevier)
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6. Postgrad Med J 2001;77:621–638
7. Renapurkar, et al. Metabolic and storage lung diseases: spectrum of imaging appearances
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