

# AMSER Rad-Path Case of the Month:

50-year-old female with a history of urothelial cancer who presents for restaging.

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# Patient Presentation

- 50-year-old female presenting for cancer surveillance imaging.
- **PMH/PSH:** Invasive high grade urothelial carcinoma of the bladder status post neoadjuvant ddMVAC chemotherapy (March 2019) and robotic-assisted laparoscopic radical cystectomy, vaginal sparing with bilateral oophorectomy and radical hysterectomy, bilateral pelvic lymph node dissections and creation of an ileal neobladder (June 2019) demonstrating ypT2a N0 disease.
- **Recent Medical History:** Hospitalized for a URI.
- **Social History:** Current cigarette smoker (1ppd x 35 years)
- **Physical Exam:** Lungs: Clear to auscultation bilaterally. Other non-contributory.

What Imaging Should We Order?

# ACR Appropriateness Criteria: Post-Treatment Surveillance of Bladder Cancer

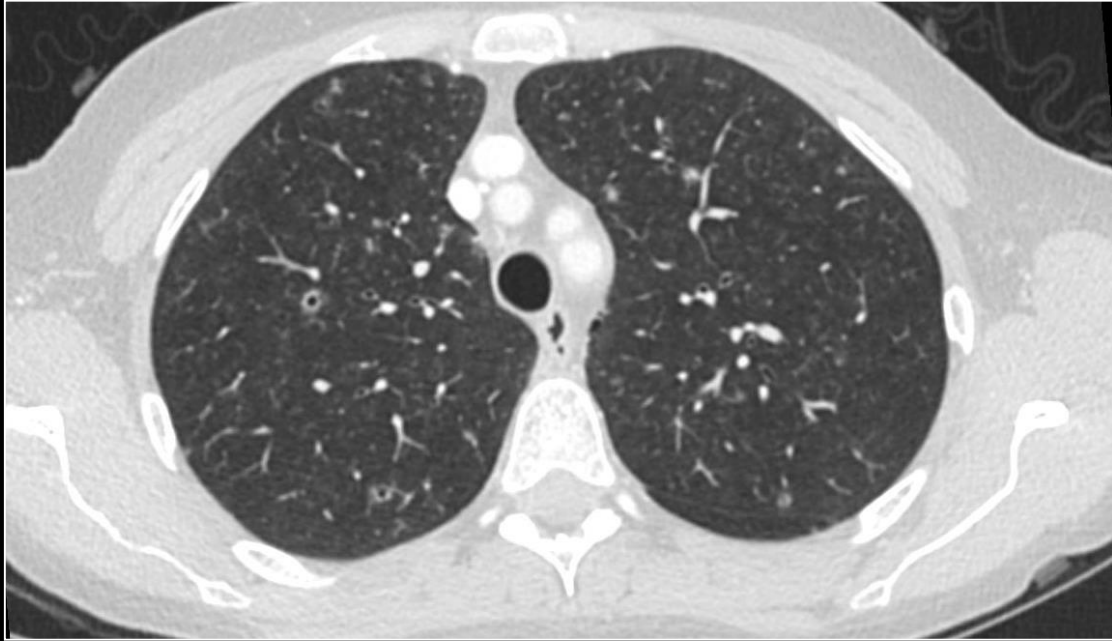
**Variant 3:** Muscle-invasive bladder cancer (MIBC) with or without cystectomy. Post-treatment surveillance.

| Procedure   | Appropriateness Category | Relative Radiation Level |
|---|--------------------------|--------------------------|
| Radiography chest                                   | Usually Appropriate      | ⊕                        |
| CT abdomen and pelvis without and with IV contrast  | Usually Appropriate      | ⊕⊕⊕⊕                     |
| Fluoroscopy abdomen loopogram                       | Usually Appropriate      | ⊕⊕⊕                      |
| CT abdomen and pelvis with IV contrast              | Usually Appropriate      | ⊕⊕⊕                      |
| MRI abdomen and pelvis without and with IV contrast | Usually Appropriate      | ○                        |
| CT chest with IV contrast                           | May Be Appropriate       | ⊕⊕⊕                      |
| CT chest without IV contrast                        | May Be Appropriate       | ⊕⊕⊕                      |
| FDG-PET/CT skull base to mid-thigh                  | May Be Appropriate       | ⊕⊕⊕⊕                     |
| MRI abdomen and pelvis without IV contrast          | May Be Appropriate       | ○                        |
| CT abdomen and pelvis without IV contrast           | Usually Not Appropriate  | ⊕⊕⊕                      |
| Radiography intravenous urography                   | Usually Not Appropriate  | ⊕⊕⊕                      |
| US pelvis (bladder)                                 | Usually Not Appropriate  | ○                        |
| CT chest without and with IV contrast               | Usually Not Appropriate  | ⊕⊕⊕                      |

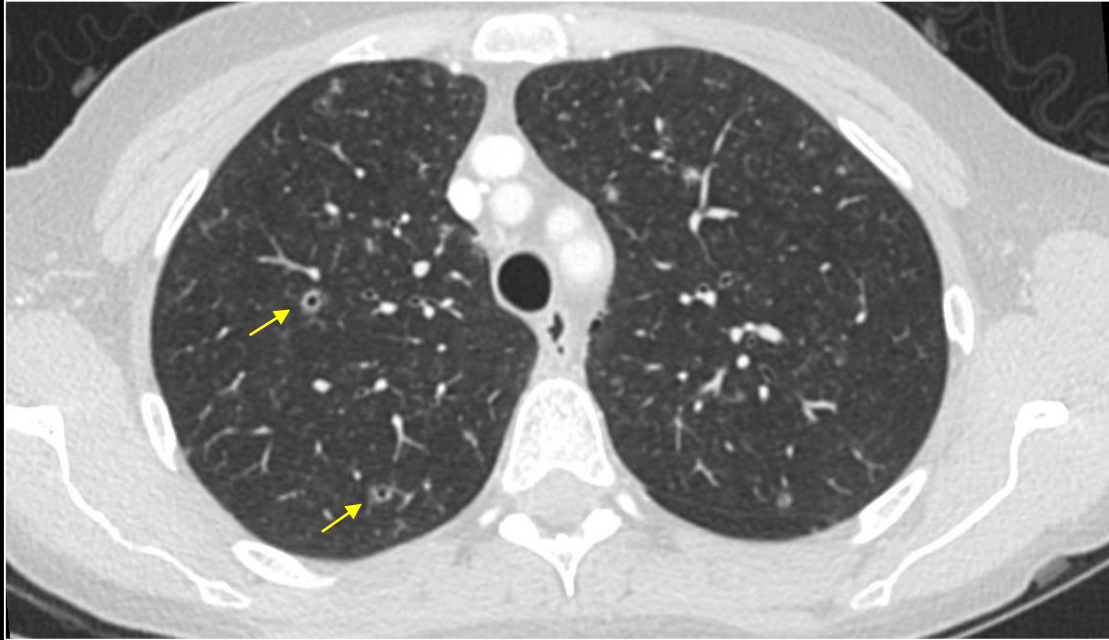
← These scans were ordered by the oncologist.



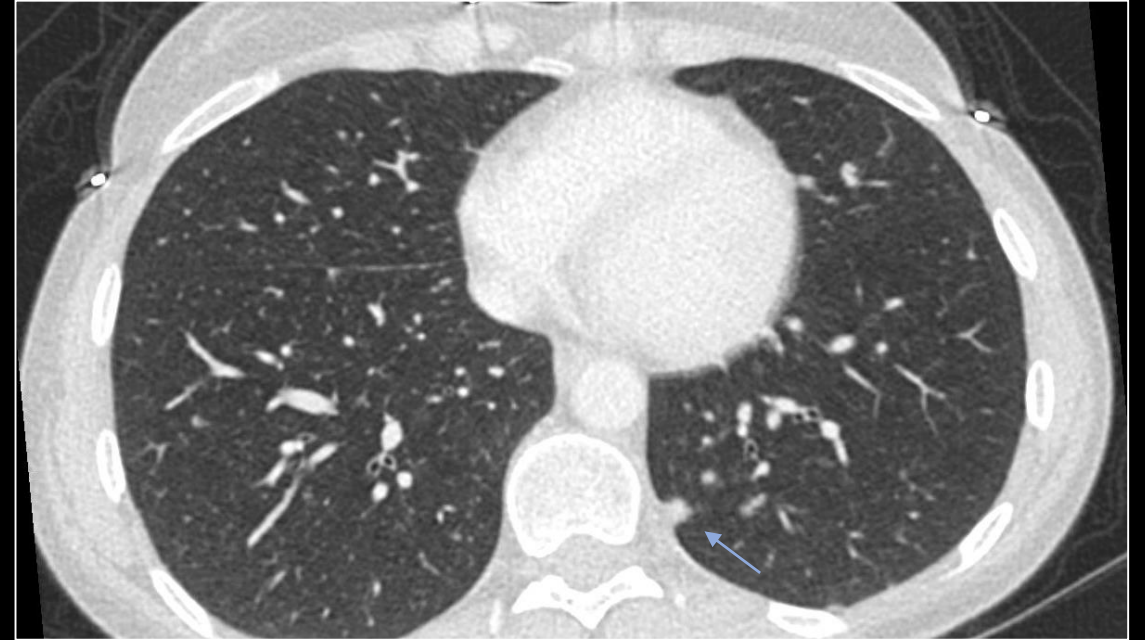
# Chest CT (unlabeled)



# Chest CT (labeled)



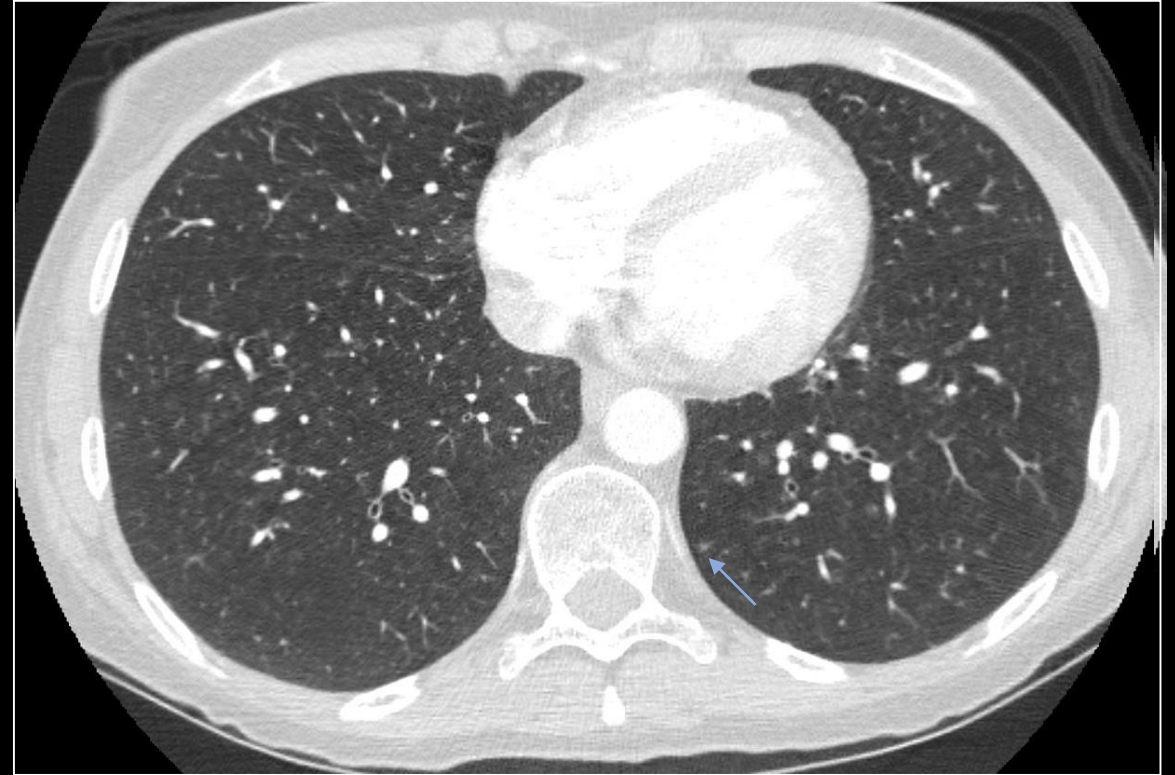
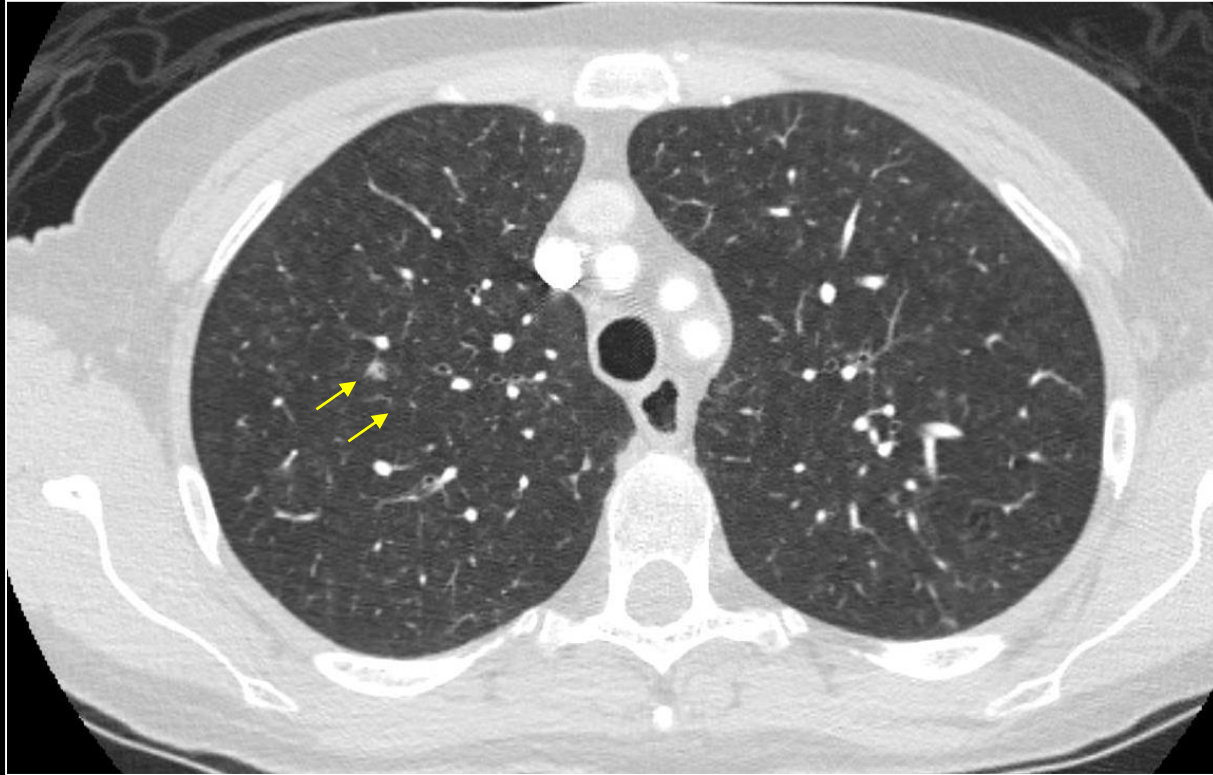
Axial CT demonstrated multiple solid and cavitory nodules. On the left image, there are 2 subcentimeter cavitory nodules in the right upper lobe (yellow arrows).



Axial CT demonstrated multiple solid and cavitory nodules. On the right image, there is a 6mm solid nodule in the left lower lobe (blue arrow).



# More information - Chest CT



The nodules had increased in size compared to a CT scan done two months previously with selected images above.

# DDX of Cavitory Pulmonary Nodules (based on imaging):

- Metastases
- Wegener's disease / Vasculitis Nodules
- Septic Emboli
- Pulmonary Langerhans Cell Histiocytosis (PLCH)
- Cavitated *P. Jiroveci* lesions
- Primary Lung Malignancy (Squamous cell carcinoma)

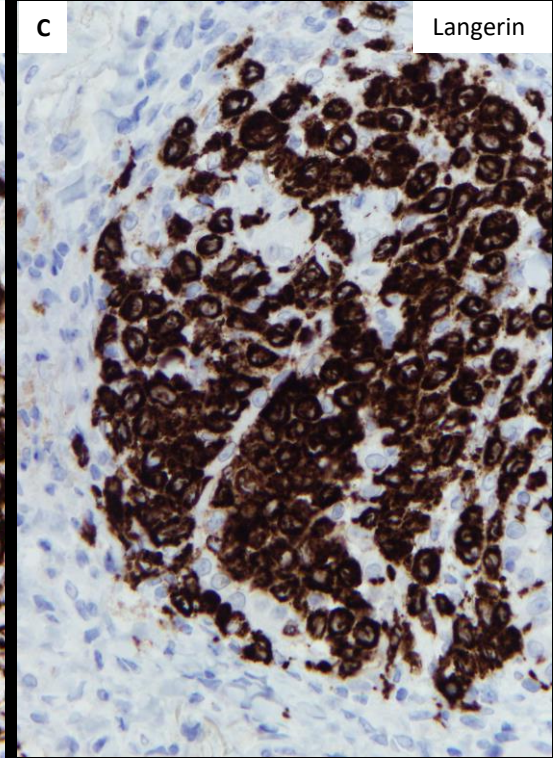
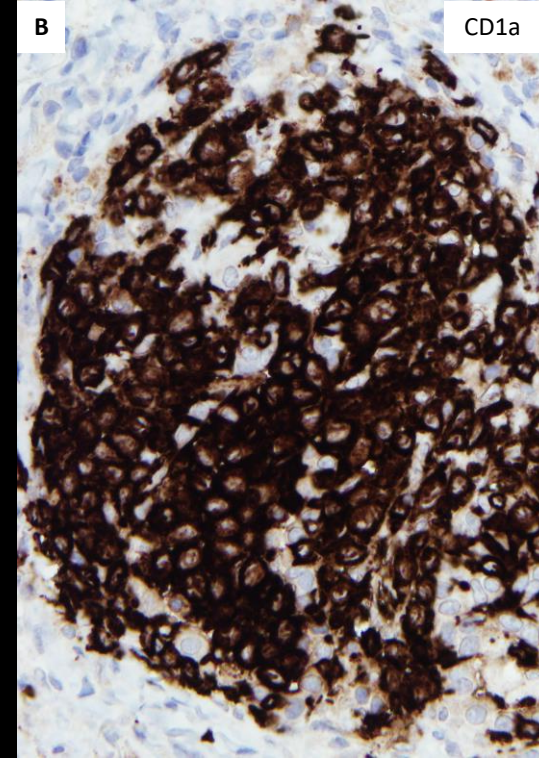
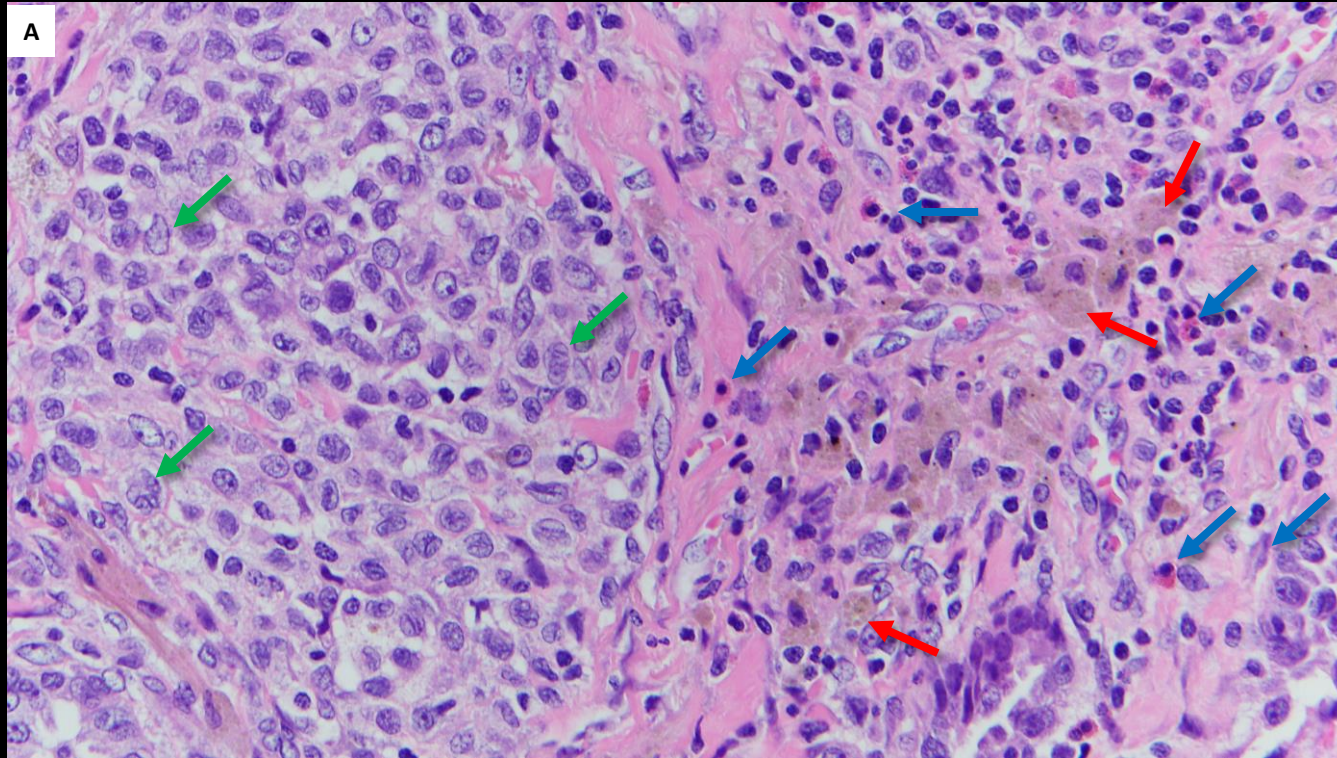
A right VATS wedge resection was performed for definitive diagnosis.



# DDX of Solid Pulmonary Nodules / Micronodules (based on imaging):

- Metastases
- Infection/Inflammatory (Tuberculosis)
- Sarcoidosis / Silicosis
- Wegener's disease / Vasculitis Nodules
- Pulmonary Langerhans Cell Histiocytosis (PLCH)
- RB-ILD
- Primary Lung Malignancy

A right VATS wedge resection was performed for definitive diagnosis.



(A) Photomicrograph (hematoxylin-eosin stain) from a right wedge resection show a nodular proliferation of **Langerhans cells** admixed with frequent **eosinophils** and **pigmented (smoker's) macrophages**. (B, C) CD1a and Langerin immunohistochemistry stains highlight abundant Langerhans cells.

Final Dx:

Pulmonary Langerhans Cell Histiocytosis (PLCH)

# Pulmonary Langerhans Cell Histiocytosis (PLCH)

- **Overview**

- PLCH is distinct from systemic Langerhans Cell Histiocytosis (LCH)
- A rare interstitial granulomatous disease occurring primarily in young smokers characterized by an accumulation of Langerhans and other inflammatory cells in small airways resulting in the formation of nodular lesions

- **Pathophysiology**

- Accumulation of activated Langerhan cells (LC) organized into loose granulomas that develop in, and destroy, the distal bronchiole walls is the pathologic hallmark of PLCH. Lymphocytes and inflammatory cells, including eosinophils and macrophages are also found.
- It is unclear if PLCH is a reactive or neoplastic process

- **Epidemiology**

- Rare disorder of unknown etiology
- Young adults, 20-40 years
- No gender predilection
- Caucasians > African Americans or Asians
- Smoking history in 95% of patients

# Pulmonary Langerhans Cell Histiocytosis (PLCH)

- **Presentation**

- 2/3 of patients are symptomatic at presentation
- Most common symptoms: non-productive cough, dyspnea, chest pain, fatigue, weight loss, fever
- Spontaneous pneumothorax in 15% of patients

- **Treatment**

- Smoking cessation
- Corticosteroids (prednisone 0.5-1.0mg/kg daily with a slow taper over months)
- Cladribine, cyclophosphamide, or methotrexate for progressive disease
- Lung transplant for advanced disease refractory to immunosuppressants

- **Prognosis**

- Spontaneous resolution or stabilization in 50% of cases
- Progression to end-stage pulmonary fibrosis in 20% of cases

- **Associations**

- Acute lymphoblastic leukemia
- Acute myeloid leukemia



# Imaging Findings of PLCH

- **Early**

- bilateral reticulonodular pattern of few to innumerable nodules
- nodules 1-10mm in size with irregular margins
- predilection for upper to mid lungs
- centrilobular distribution
- preserved lung volumes or hyperinflation

- **Late**

- cysts are more pronounced in the later stages of disease
- cysts usually less than 10mm in diameter but may measure up to 2-3 cm
- coalescent cysts, honeycombing and fibrosis



# Pulmonary Langerhans Cell Histiocytosis (PLCH)

- **Gross description**

- Early disease with fine nodular infiltrate
- Advanced disease with cysts and honeycombing

- **Microscopic description**

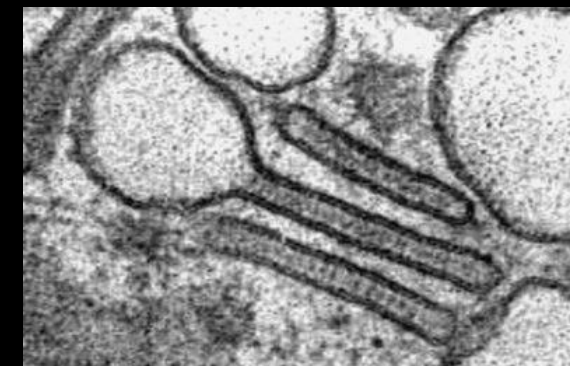
- Interstitial proliferation of Langerhans cells distributed along small airways, often resulting in bronchiolocentric stellate-shaped nodules
- Numerous admixed eosinophils, lymphocytes, plasma cells, pigmented macrophages, and giant cells
- Abundance of Langerhans cells – relatively large cells with a granular, mildly eosinophilic cytoplasm and “crumpled tissue paper” nuclear outlines
- Late or more advanced lesions may be almost totally devoid of Langerhans cells and replaced by stromal fibrosis
- Nodules may cavitate and form cysts
- ~60% of cases show BRAF V600E mutation

- **Positive stains**

- CD1a, Langerin, and S100

- **Electron microscopy**

- Birbeck granules (intracytoplasmic structures that are shaped like tennis rackets)



# References

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