Rady 403 Case Presentation

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Focused Patient History

4 y.o female with history of asthma presenting for 1 month of persistent cough

- 1 month ago presented to PCP with fever, cough, congestion, and sinusitis leading to a diagnosis of viral pneumonia
- 2-3 week later she presented again for persistent symptoms. She received a chest Xray, which led to the diagnosis of left lower lobe pneumonia and was prescribed antibiotics
- She presented to the ED 10 days later for persistent cough despite antibiotic treatment

Physical exam was significant for diminished lung sounds in left lower lobe. Other exams were within normal limits

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List of imaging studies

Chest X Ray

CT chest

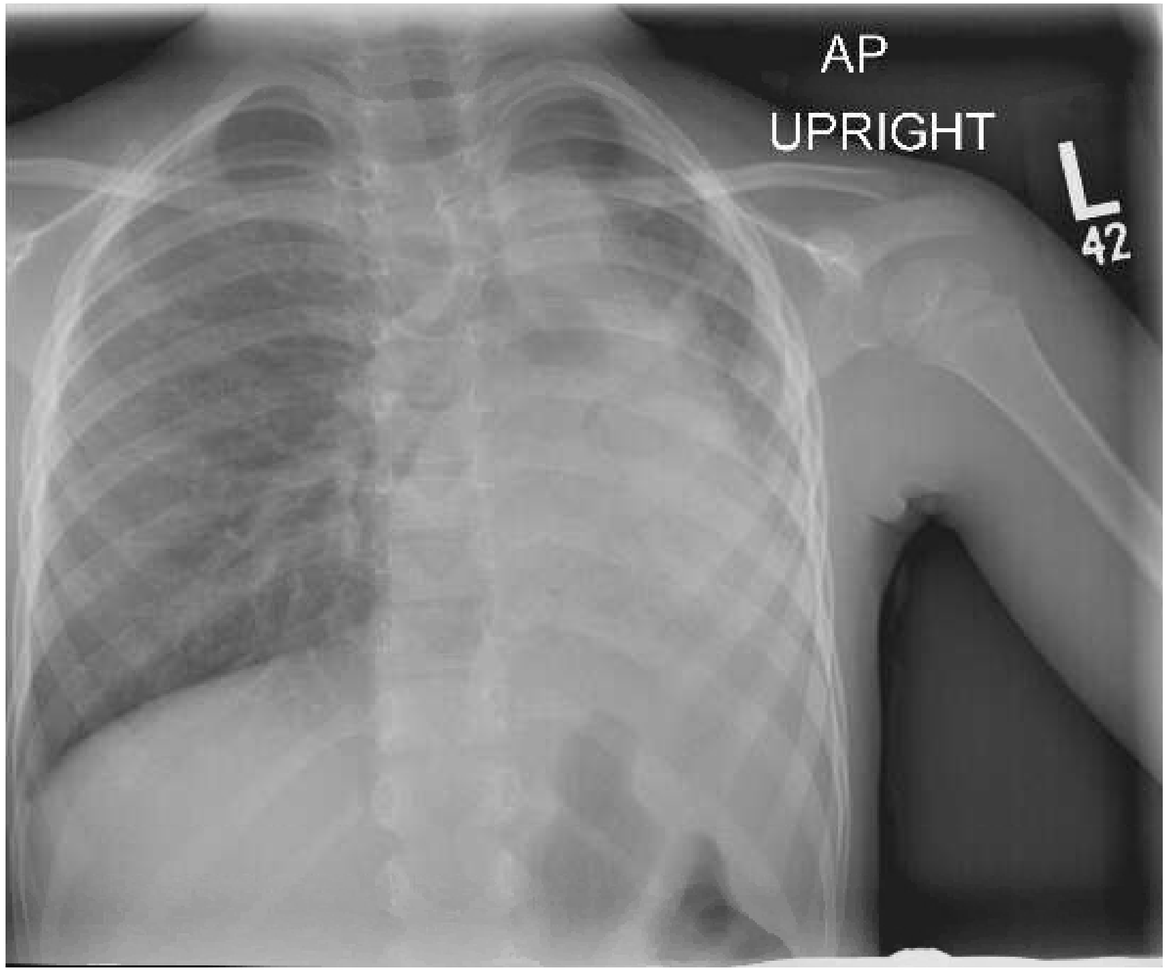
Bronchoscopy

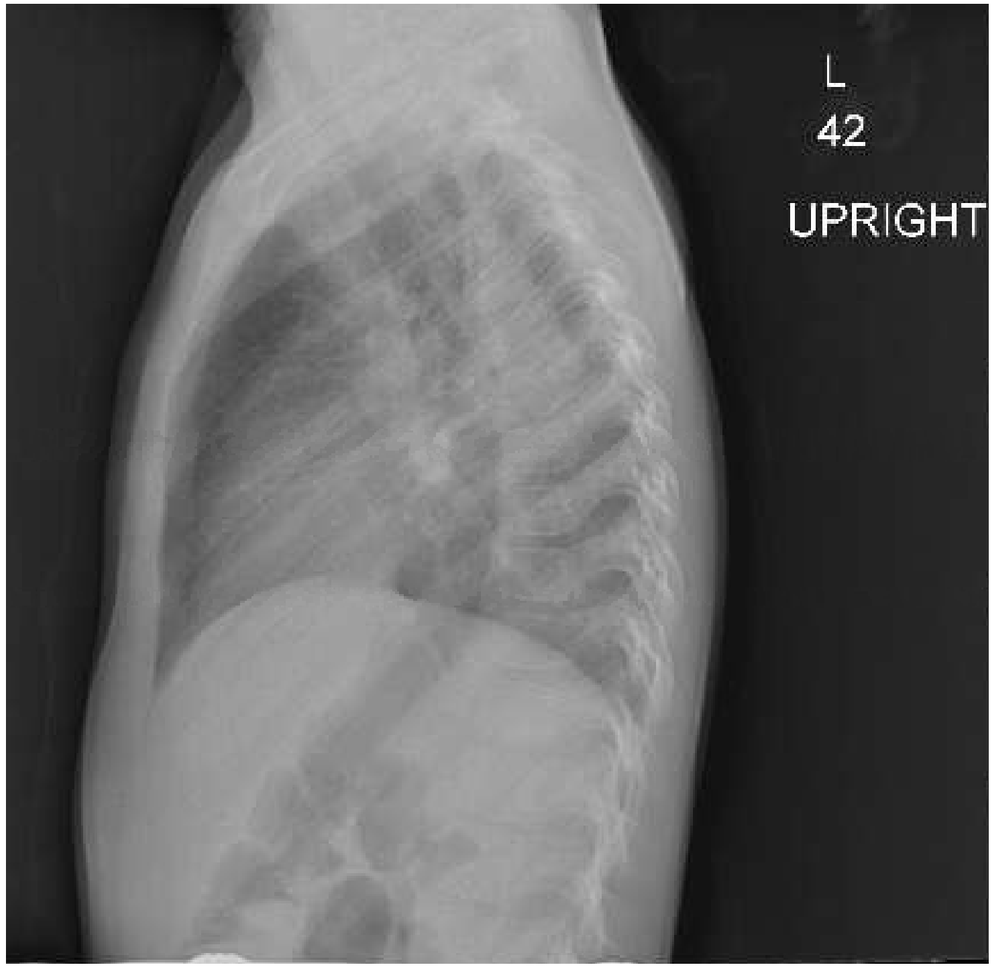
MRI Chest

MRI Abdomen and Pelvis

CT abdomen and pelvis

2 View Chest Radiograph





What do you see?

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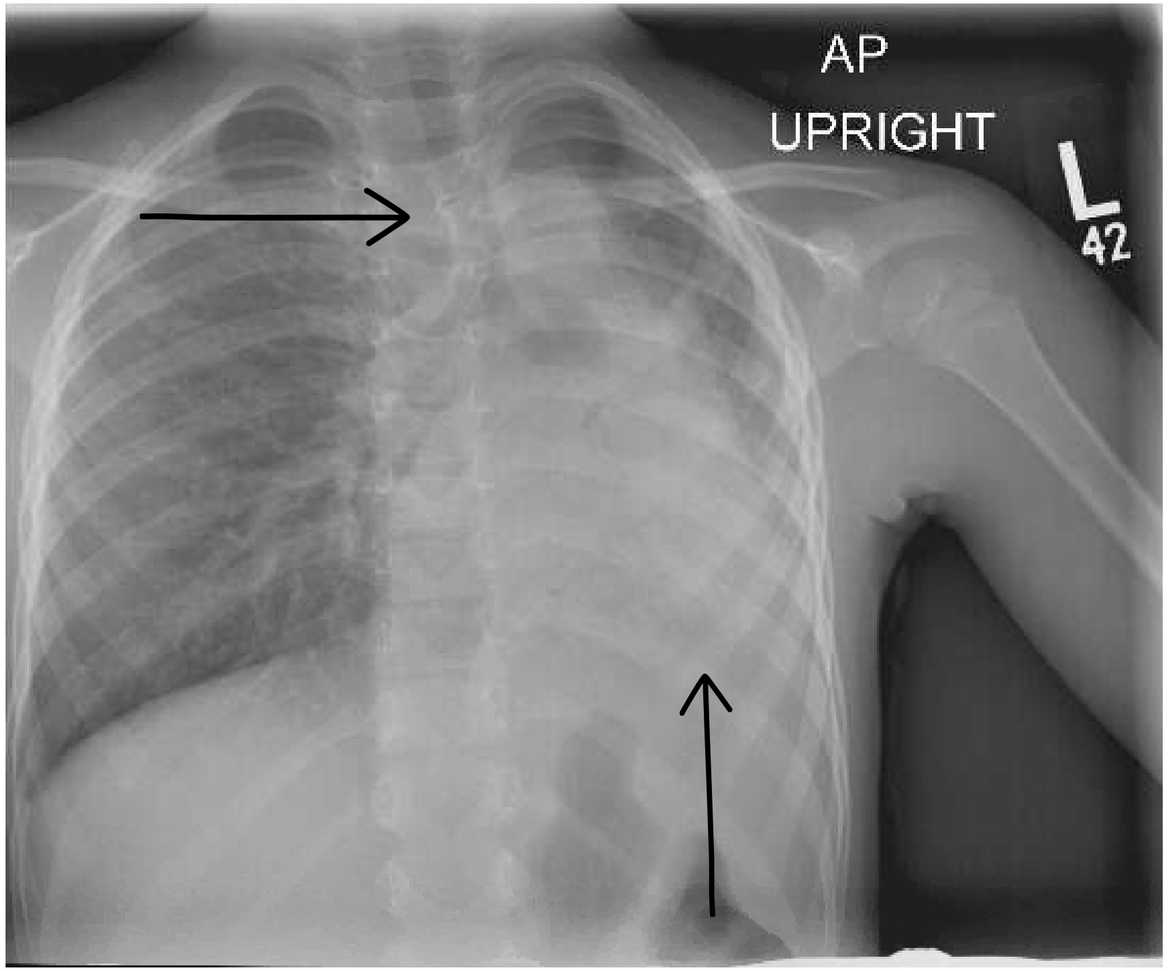
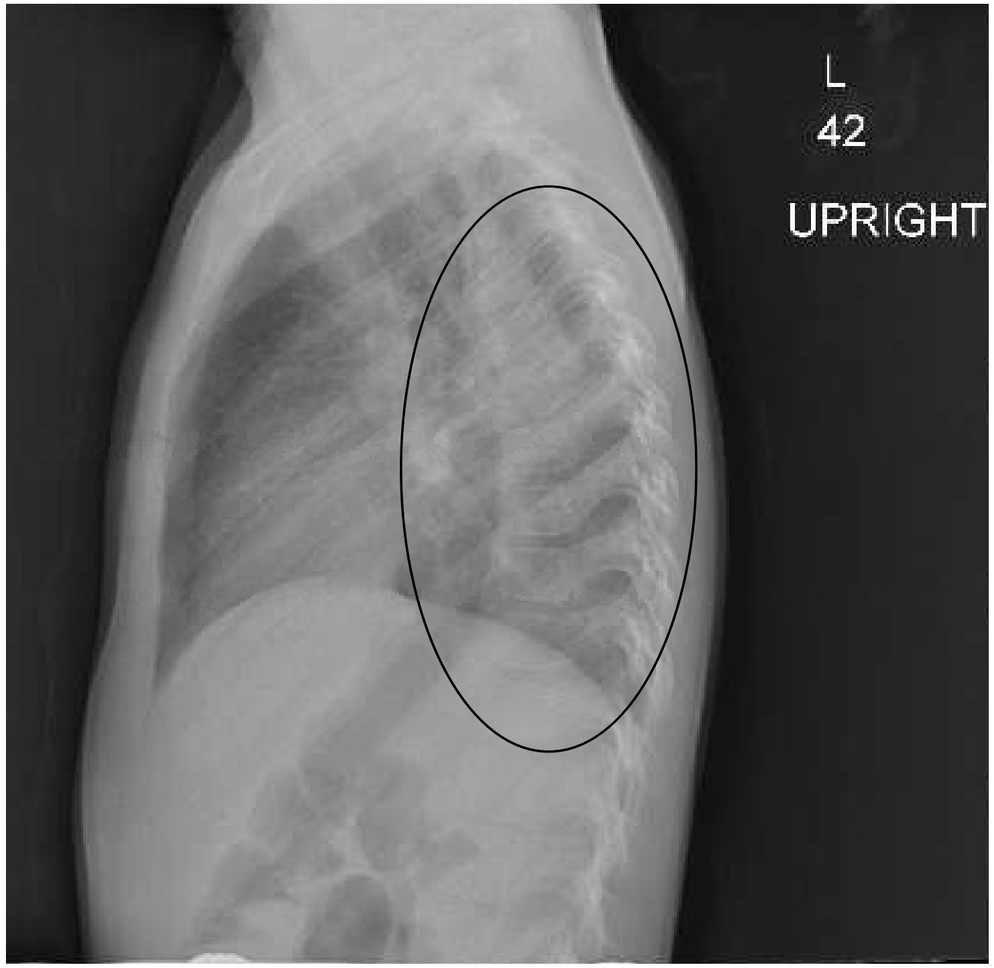
2 View Chest Radiograph





Left lingula and lower lung consolidation

Burring of costophrenic angle

Cardiac silhouetting

Tracheal deviation towards lesion

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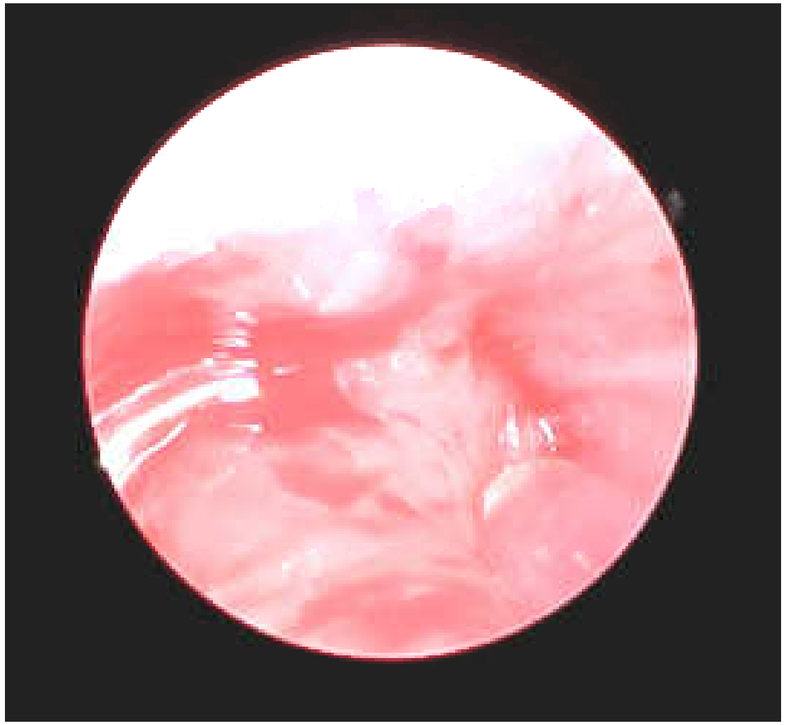
What's Next?

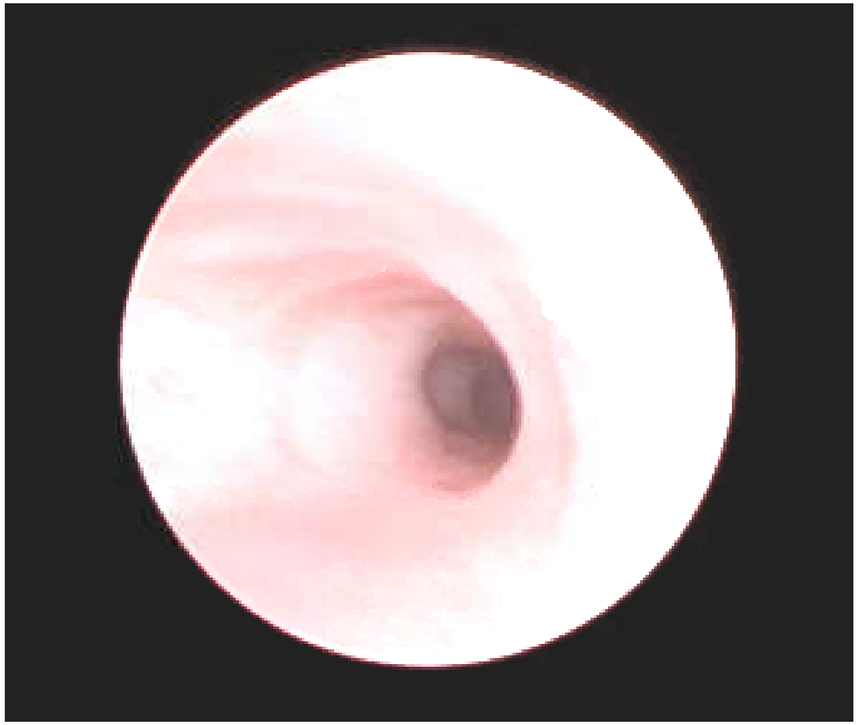
Pt was discharged with antibiotics to help clear what was presumed to be sequelae of lobar pneumonia and followed up with a bronchoscopy to assess for mucus plug obstruction.

Bronchoscopy Images

Occluded Left Mainstem Bronchus



Patent Right Mainstem Bronchus



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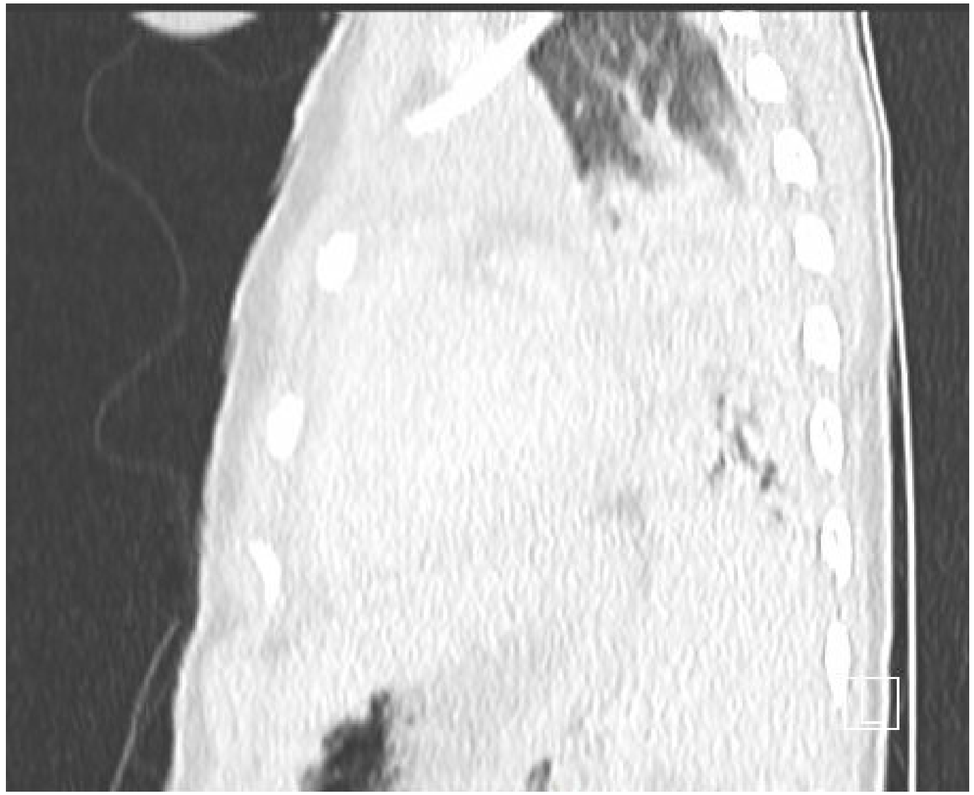
Sagittal CT Chest
- lung window

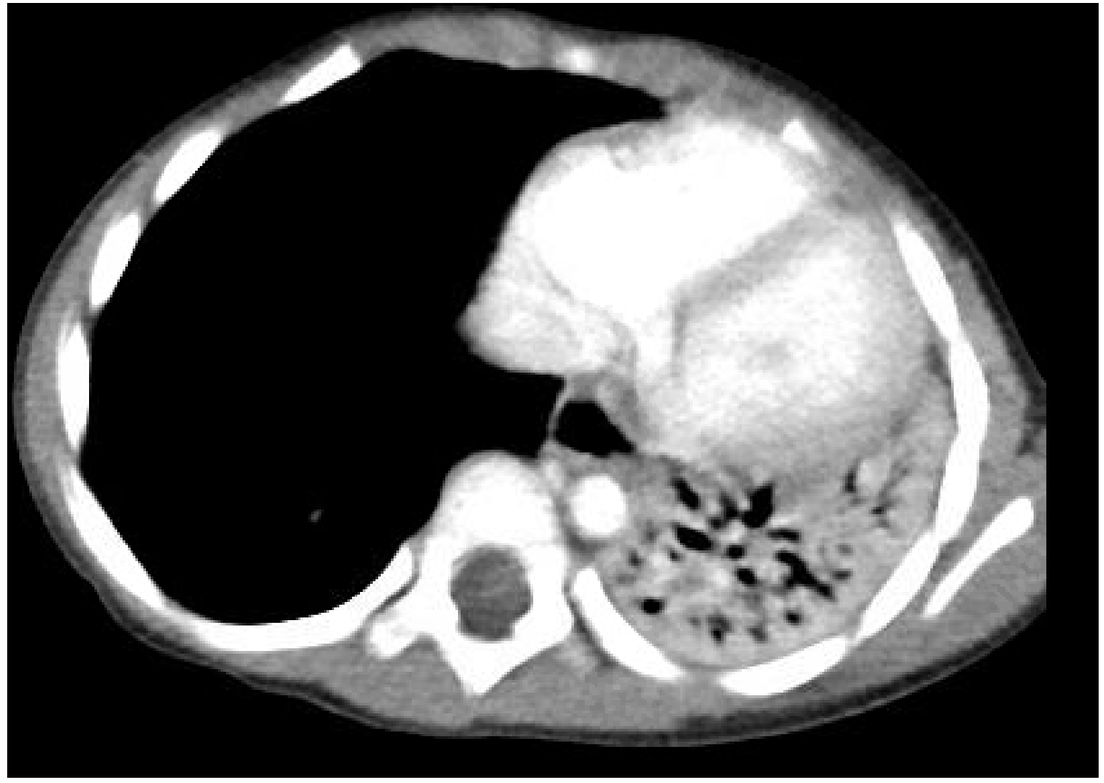


Coronal CT Chest

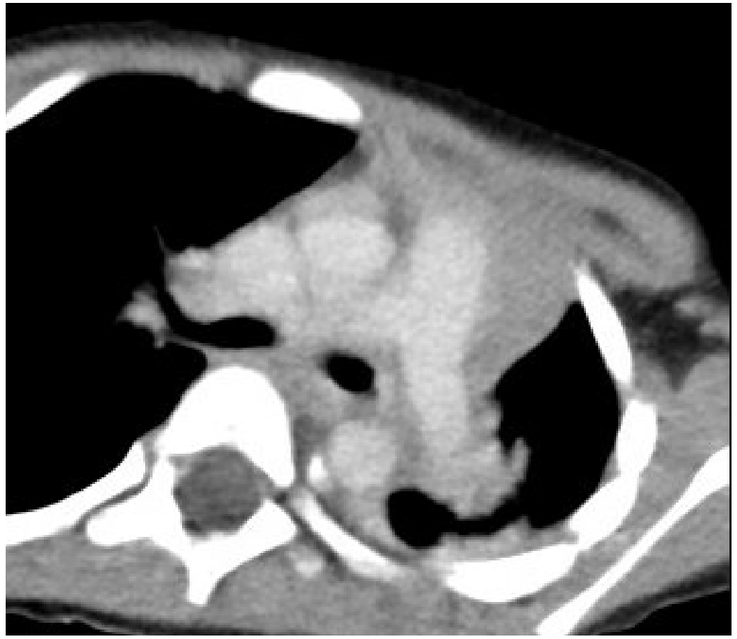
Occlusion of left bronchus

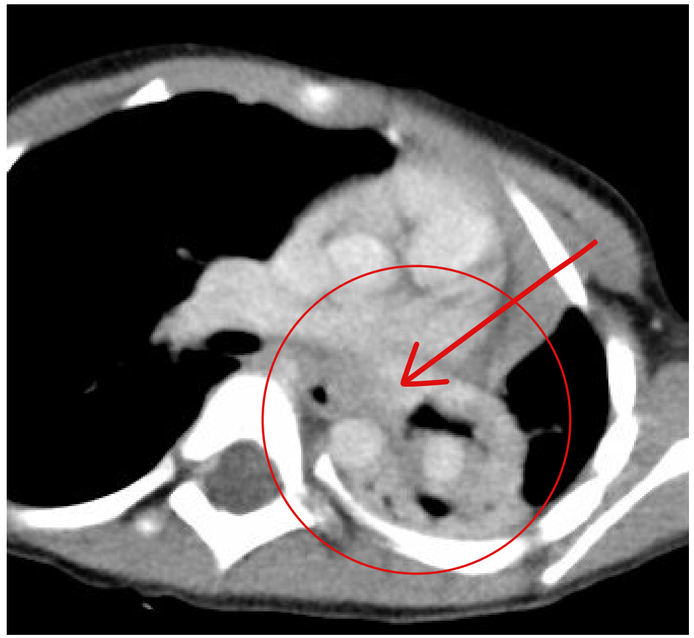
Axial CT Non-Contrast - lung window

Axial CT With Contrast



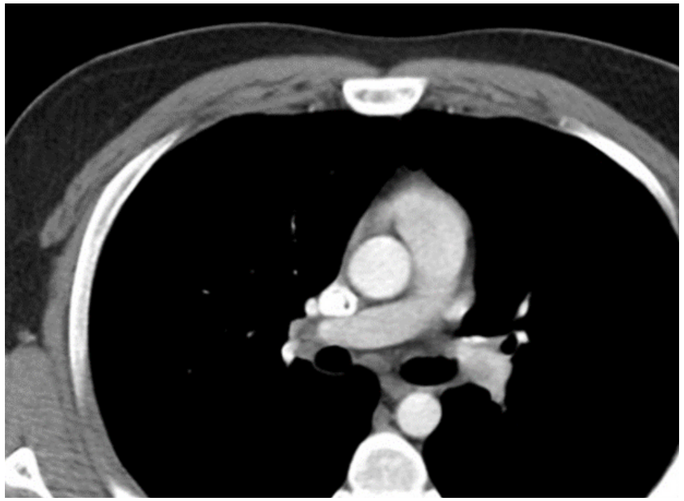
Patient Axial CT

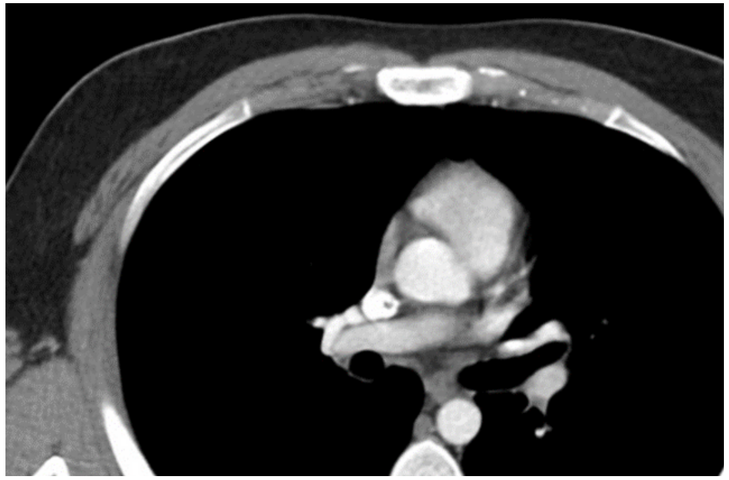




Airway occluded

Normal Reference

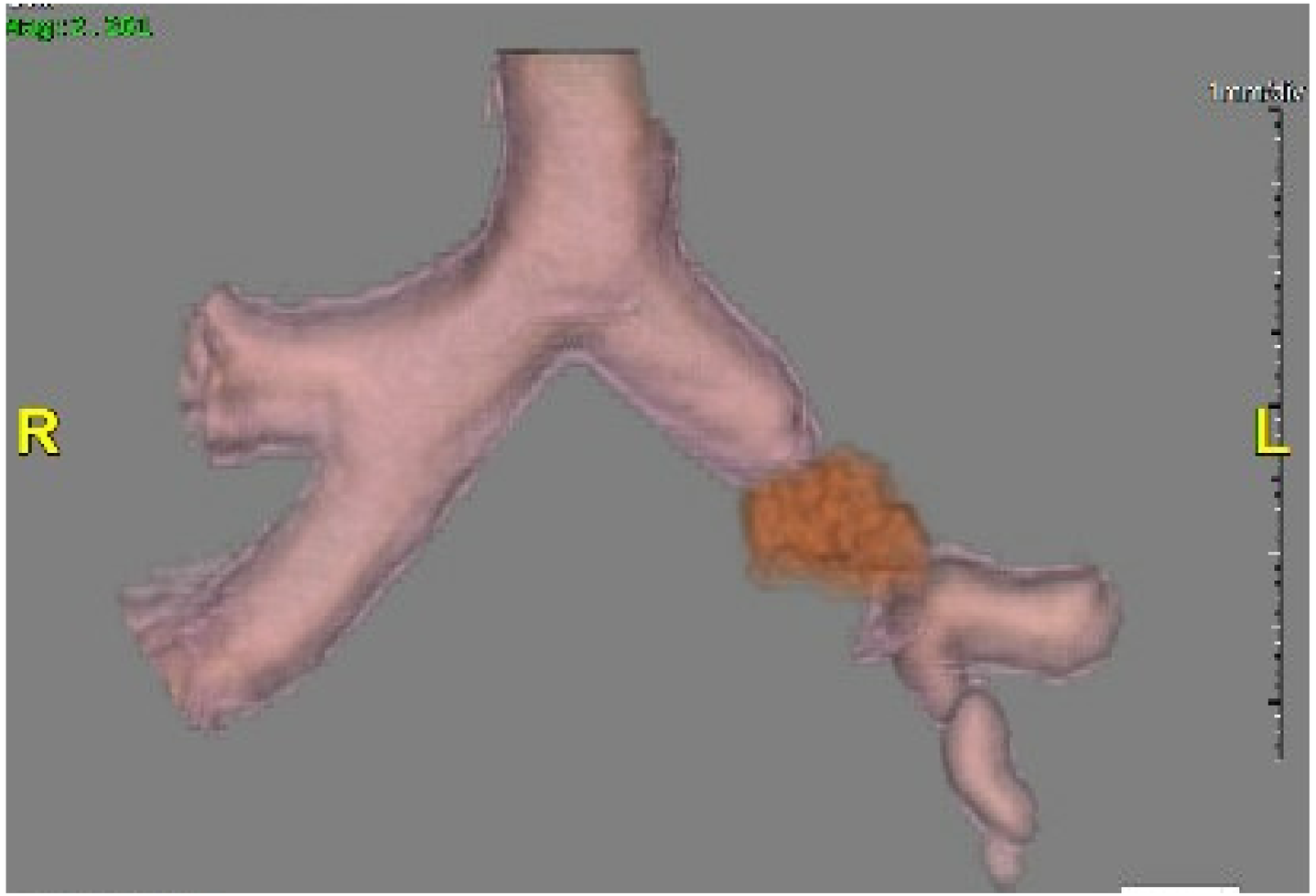




Impression from CT images

- There is occlusion of the left mainstem bronchus secondary to a mass/lesion
- The left lower lobe appears completely atelectatic with air bronchograms, suggestive of postobstructive syndrome.
- Preserved aeration of the left upper lobe

3D Airway Reconstruction



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Inflammatory Myofibroblastic Tumor (IMT)

Although rare (150-200 cases per year), IMT is one of the most common primary lung tumors in pediatric patients. Etiology of IMT is unclear but theories suggest inflammatory reaction to an infection versus underlying low grade malignancy. Most tumors are benign but some can be malignant and invade surrounding structures [1-2].

- Can occur anywhere in the body but are commonly in lung, abdomen/pelvis, and retroperitoneum.
- Can be in any age, but more common in children
- ALK negative IMTs may be more aggressive with a higher frequency of metastasis compared to ALK positive IMT

Inflammatory Myofibroblastic Tumor

Symptoms:

- Patients can be asymptomatic (70%) but some can have cough, dyspnea, chest pain, fever, night sweats or hemoptysis [1,3].

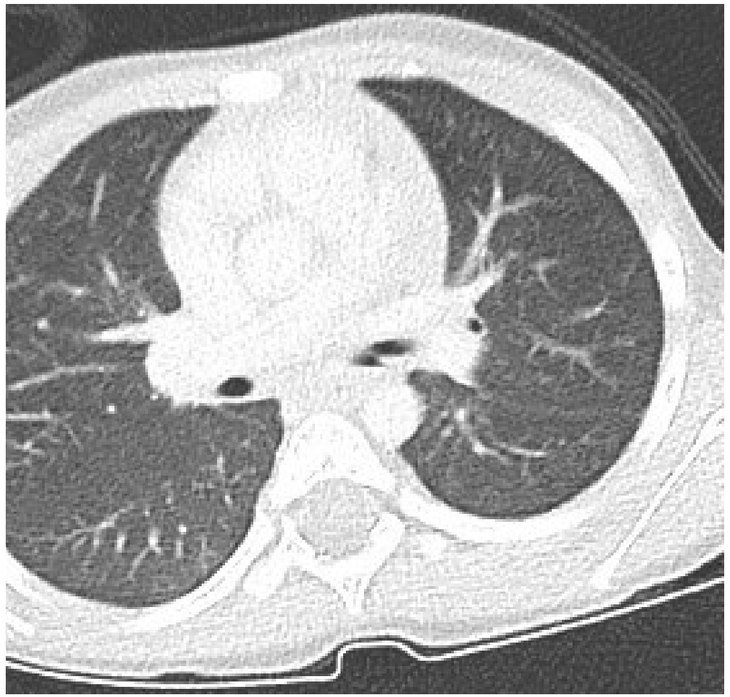
Diagnostics:

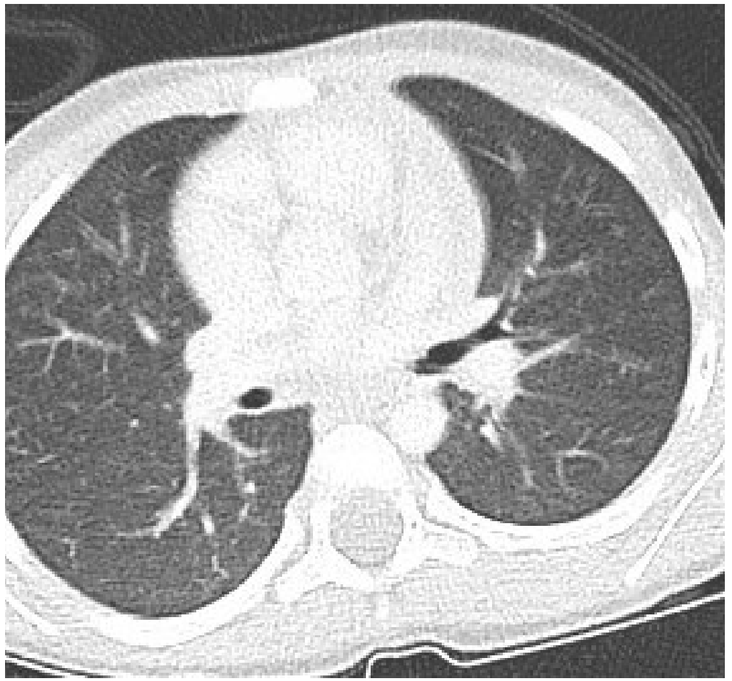
- CT, ultrasound, MRI and biopsy

Management:

- Surgical resection
- Chemo therapy or glucocorticoids for those who cannot tolerate surgery
- Tyrosine Kinase inhibitors such as crizotinib

CT Chest with Contrast - lung window





after 2-3 months on Crizotinib

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IMT and ALK (anaplastic lymphoma kinase) expression

- About 50% of IMT have AlK expression by immunohistochemistry [2]
- Most common mechanism of ALK expression involves structural rearrangements in the ALK gene, leading to the formation of a chimeric fusion proteins [2]
- Many fusion proteins have been studied and can be targeted by medication
- ALK+ patients respond to tyrosine kinase inhibitors such as Crizotinib

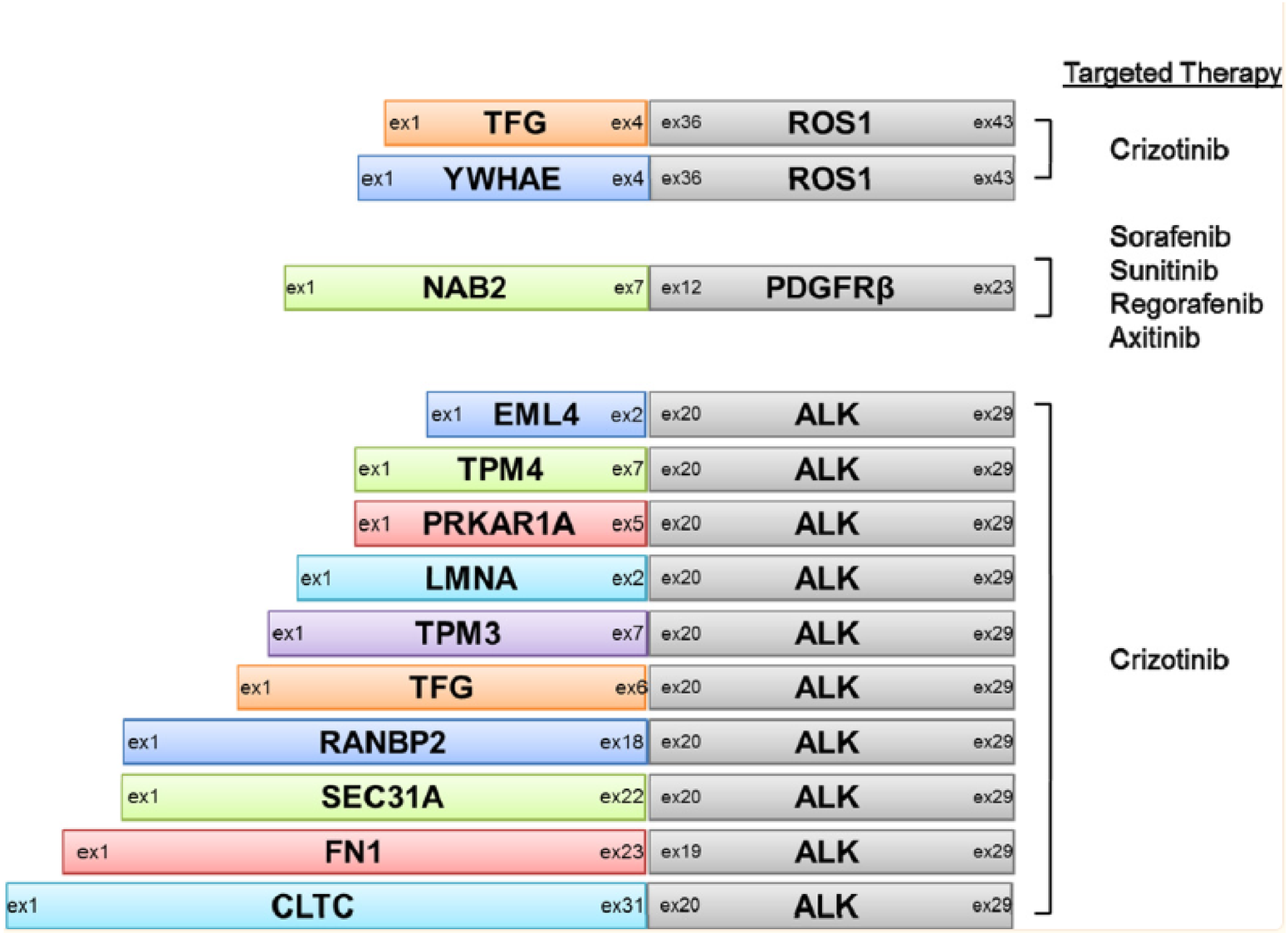

image from Lovly CM, Gupta A, Lipson D, et al. Inflammatory myofibroblastic tumors harbor multiple potentially actionable kinase fusions. Cancer Discov. 2014;4(8):889-895. doi:10.1158/2159-8290.CD-14-0377

Patient Treatment or Outcome

Pt had a biopsy via bronchoscopy that showed ALK+ inflammatory myofibroblastic tumor

- She received and is currently on Crizotinib
- She is now s/p left mainstem bronchial sleeve resection
- Positive margins found on post op biopsy despite resection of some normal appearing lung
- Possible lobectomy or pneumonectomy in the future for definitive treatment

Although cough was documented for less than 8 weeks in this pt, this criteria fit the best

Variant 3: **Chronic cough lasting more than 8 weeks. Persistent symptoms despite initial clinical evaluation and empiric treatment. Initial imaging.**

Procedure	Appropriateness Category	Relative Radiation Level
Radiography chest	Usually Appropriate	☢
CT chest with IV contrast	Usually Appropriate	☢☢☢
CT chest without IV contrast	Usually Appropriate	☢☢☢
CT maxillofacial without IV contrast	May Be Appropriate	☢☢
Fluoroscopy biphasic esophagram	Usually Not Appropriate	☢☢☢
MRI heart function and morphology without and with IV contrast	Usually Not Appropriate	O
CT maxillofacial with IV contrast	Usually Not Appropriate	☢☢
CT chest without and with IV contrast	Usually Not Appropriate	☢☢☢
CT maxillofacial without and with IV contrast	Usually Not Appropriate	☢☢☢
V/Q scan lung	Usually Not Appropriate	☢☢☢
FDG-PET/CT skull base to mid-thigh	Usually Not Appropriate	☢☢☢☢
SPECT or SPECT/CT MPI rest and stress	Usually Not Appropriate	☢☢☢☢

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UNC Top 3

- Although CT is appropriate for IMT, it has non specific findings such as a heterogenous mass. It can also be misleading on chest radiograph
- Have suspicion for tumors or abnormal physiology in previously healthy patients presenting with chronic cough despite treatment
- ALK+ IMT is less aggressive and has targeted therapy

References

1. Weinberger, SE. Inflammatory myofibroblastic tumor (plasma cell granuloma) of the lung. In: UpToDate, Post, TW (Ed), UpToDate, Waltham, MA, 2020.
2. Lovly CM, Gupta A, Lipson D, et al. Inflammatory myofibroblastic tumors harbor multiple potentially actionable kinase fusions. Cancer Discov. 2014;4(8):889-895. doi:10.1158/2159-8290.CD-14-0377
3. Inflammatory Myofibroblastic Tumor. National Cancer Institute. Published February 27, 2019. Accessed July 20, 2023. https://www.cancer.gov/pediatric-adult-rare-tumor/rare-tumors/rare-soft-tissue-tumors/inflammatory-myofibroblastic-tumor
4. Inflammatory myofibroblastic tumor of the lung. Radiopaedia. Accessed July 20, 2023. https://radiopaedia.org/articles/inflammatory-myofibroblastic-tumour-of-the-lung?lang=us