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
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?
2 View Chest Radiograph

- Left lingula and lower lung consolidation
- Burring of costophrenic angle
- Cardiac silhouetting
- Tracheal deviation towards lesion
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.
Sagittal CT Chest
- lung window

Coronal CT Chest

Occlusion of left bronchus
Axial CT Non-Contrast - lung window

Axial CT With Contrast
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
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
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
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

<table>
<thead>
<tr>
<th>Variant 3:</th>
<th>Chronic cough lasting more than 8 weeks. Persistent symptoms despite initial clinical evaluation and empiric treatment. Initial imaging.</th>
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</thead>
<tbody>
<tr>
<td><strong>Procedure</strong></td>
<td><strong>Appropriateness Category</strong></td>
</tr>
<tr>
<td>Radiography chest</td>
<td>Usually Appropriate</td>
</tr>
<tr>
<td>CT chest with IV contrast</td>
<td>Usually Appropriate</td>
</tr>
<tr>
<td>CT chest without IV contrast</td>
<td>Usually Appropriate</td>
</tr>
<tr>
<td>CT maxillofacial without IV contrast</td>
<td>May Be Appropriate</td>
</tr>
<tr>
<td>Fluoroscopy biphasic esophagram</td>
<td>Usually Not Appropriate</td>
</tr>
<tr>
<td>MRI heart function and morphology without and with IV contrast</td>
<td>Usually Not Appropriate</td>
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<tr>
<td>CT maxillofacial with IV contrast</td>
<td>Usually Not Appropriate</td>
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<tr>
<td>CT chest without and with IV contrast</td>
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<tr>
<td>CT maxillofacial without and with IV contrast</td>
<td>Usually Not Appropriate</td>
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<tr>
<td>V/Q scan lung</td>
<td>Usually Not Appropriate</td>
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<tr>
<td>FDG-PET/CT skull base to mid-thigh</td>
<td>Usually Not Appropriate</td>
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<tr>
<td>SPECT or SPECT/CT MPI rest and stress</td>
<td>Usually Not Appropriate</td>
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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.

