AMSER Case of the Month:
July 2019
Febrile with Recurrent Pneumonia

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Patient Presentation

• HPI: 46 year old female with Kartagener’s Syndrome and recent Pseudomonas infection s/p cefepime treatment, presented to ED with worsening shortness of breath. In the ED, the patient was febrile to 102 F and tachypnic. She was started on vancomycin, vefepime, doxycycline, and albuterol nebulizer and admitted to the ICU.
• PMH: Kartagener’s Syndrome, Bronchiectasis, Sinusitis, Hyperlipidemia
• Surg Hx: Sinus surgery, appendectomy
• Med: Flonase, Advair, Singulair, Spiriva, Low-Ogestrel
• Allergies: Bactrim, Sulfa
• Family Hx: Father – CAD, Mother – Gallbladder disease
• Social Hx: No tobacco, alcohol, or drug use
What Imaging Should We Order?
### ACR Appropriateness Criteria

**Variant 2:** Acute respiratory illnesses in immunocompetent patients with positive physical examination, abnormal vital signs, organic brain disease, or other risk factors. Initial imaging.

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Appropriateness Category</th>
<th>Relative Radiation Level</th>
</tr>
</thead>
<tbody>
<tr>
<td>Radiography chest</td>
<td>Usually Appropriate</td>
<td>☀</td>
</tr>
<tr>
<td>US chest</td>
<td>May Be Appropriate</td>
<td>☀</td>
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<tr>
<td>CT chest with IV contrast</td>
<td>Usually Not Appropriate</td>
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<tr>
<td>CT chest without and with IV contrast</td>
<td>Usually Not Appropriate</td>
<td>☀ ☀ ☀</td>
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<tr>
<td>CT chest without IV contrast</td>
<td>Usually Not Appropriate</td>
<td>☀ ☀ ☀ ☀</td>
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<tr>
<td>MRI chest without and with IV contrast</td>
<td>Usually Not Appropriate</td>
<td>☀</td>
</tr>
<tr>
<td>MRI chest without IV contrast</td>
<td>Usually Not Appropriate</td>
<td>☀</td>
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</tbody>
</table>
Findings: unlabeled
Findings: labeled

- Cardiac apex pointing to the RIGHT
- Cardiac knob
- Gastric bubble in RUQ
- Elevation of LEFT hemidiaphragm
ACR Appropriateness Criteria

**Variant 3:** Acute respiratory illness in immunocompetent patients with positive physical examination, abnormal vital signs, organic brain disease, or other risk factors and negative or equivocal initial chest radiograph. Next imaging study.

<table>
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</thead>
<tbody>
<tr>
<td>CT chest without IV contrast</td>
<td>Usually Appropriate</td>
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<tr>
<td>CT chest with IV contrast</td>
<td>May Be Appropriate (Disagreement)</td>
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</tr>
<tr>
<td>US chest</td>
<td>May Be Appropriate</td>
<td>🌟</td>
</tr>
<tr>
<td>CT chest without and with IV contrast</td>
<td>Usually Not Appropriate</td>
<td>🌟🌟🌟🌟</td>
</tr>
<tr>
<td>MRI chest without and with IV contrast</td>
<td>Usually Not Appropriate</td>
<td>🌟</td>
</tr>
<tr>
<td>MRI chest without IV contrast</td>
<td>Usually Not Appropriate</td>
<td>🌟</td>
</tr>
</tbody>
</table>
Findings: unlabeled
CT of the abdomen show the stomach and spleen are in the RIGHT upper abdomen and liver is in the LEFT.
Findings: unlabeled
CT of the pelvis shows the cecum is in the LEFT lower quadrant.
Findings: unlabeled
Findings: labeled

Heart

Bronchiectasis/dilated airways with peribronchial wall thickening

Scattered areas of “tree-in-bud” nodular opacities
Diagnosis

Kartagener’s Syndrome with Situs Inversus and Recurrent Bronchiolitis/Bronchopneumonia
Case Discussion

• Initial Presentation
  • Recurrent infections of upper and lower respiratory tract in childhood
  • Common symptoms included: chronic cough, nasal congestion, mucopurulent sputum, recurrent sinusitis, and recurrent otitis media

• Imaging modalities
  • Plain CXR reveals situs inversus with dextrocardia, right-sided stomach bubble, and right aortic arch
  • CT scan of the chest reveals dextrocardia and bilateral bronchiectasis.
  • CT scan of abdomen reveals left-sided liver, right-sided stomach, and right-sided spleen.
Case Discussion

• Kartagener Syndrome is an autosomal recessive genetic disorder presented as primary ciliary dyskinesia with complete reversal of circulatory system and viscera (situs inversus totalis).

• Evidences of situs inversus totalis are seen with liver and cecum on the left, and spleen and stomach on the right of the body.

• Extensive bronchiectasis noted with bronchial dilation and peribronchial wall thickening, worst in the anatomic left middle lobe. Scattered bilateral “tree-in-bud” nodular opacities throughout the lung. These constellation of findings are consistent with bronchiolitis or bronchopneumonia.
Case Discussion

• Diagnosis of Kartagener Syndrome included triad of: situs inversus, chronic sinusitis, and bronchiectasis.
• The patient had repeat bacterial infections which caused an acute exacerbation of her bronchiectasis.
• Commonly, patients will have recurrent aspiration/pneumonia due to inability of cilia to clear airway secretion
• Recommendation of long term preventive antibiotic therapy such as macrolide.
• Vaccination against influenza and pneumococcus.
References:


• UptoDate. Primary ciliary dyskinesia (immotile-cilia syndrome). Accessed May 16 2019