AMSER Case of the Month
December 2022

HPI 23-year-old female with history of pneumonia presenting with chronic dyspnea

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Patient Presentation

HPI: 23-year old female presenting with chronic dyspnea
PMH: prior pneumonias throughout childhood
Physical Exam: Bilateral wheezing
Labs: Noncontributory
What Imaging Should We Order?
Select the applicable ACR Appropriateness Criteria

This imaging modality was ordered by the ER physician

<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>CT chest without IV contrast</td>
<td>May Be Appropriate (Disagreement)</td>
<td>⭐⭐⭐⭐⭐</td>
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<tr>
<td>CT chest with IV contrast</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>
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<tr>
<td>FDG-PET/CT skull base to mid-thigh</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>
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<tr>
<td>MRI chest without IV contrast</td>
<td>Usually Not Appropriate</td>
<td>⭐⭐⭐⭐⭐⭐</td>
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<tr>
<td>US chest</td>
<td>Usually Not Appropriate</td>
<td>⭐⭐⭐⭐⭐⭐</td>
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</table>
CXR Findings: (labeled)

- Dextrocardia
- Bronchiectasis with surrounding fibrosis
CT Chest Findings (unlabeled)
CT Chest Findings (labeled)

Severe bronchiectasis with surrounding fibrosis
Final Dx:

Bronchiectasis secondary to Kartagener Syndrome
Case Discussion

- **Patho-Physiology:** Gene mutation of dynein protein that leads to dysmotility of cilia

- **Clinical Presentation**
  - 50% present with dextrocardia
  - Frequent sinopulmonary infections due to decreased muco-ciliary clearance
  - Bronchiectasis
  - Infertility in men and women due to immotile sperm and decreased ciliary activity in fallopian tubes

- **Diagnosis can be made through**
  - Low levels of nasal nitrous oxide
  - Bronchoscopy + Electron microscope to visualize dysmotility of cilia
  - Genetic testing of DNAL1 gene Axonemal Dynein Intermediate chain gene
References:


