



UNIVERSITY of  
**ROCHESTER**  
MEDICAL CENTER

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DEPARTMENT OF IMAGING SCIENCES

## Imaging Sciences Interesting Cases

### CASE 52

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**CLINICAL PRESENTATION:** A 69-year-old woman presented to the hospital with dyspnea.

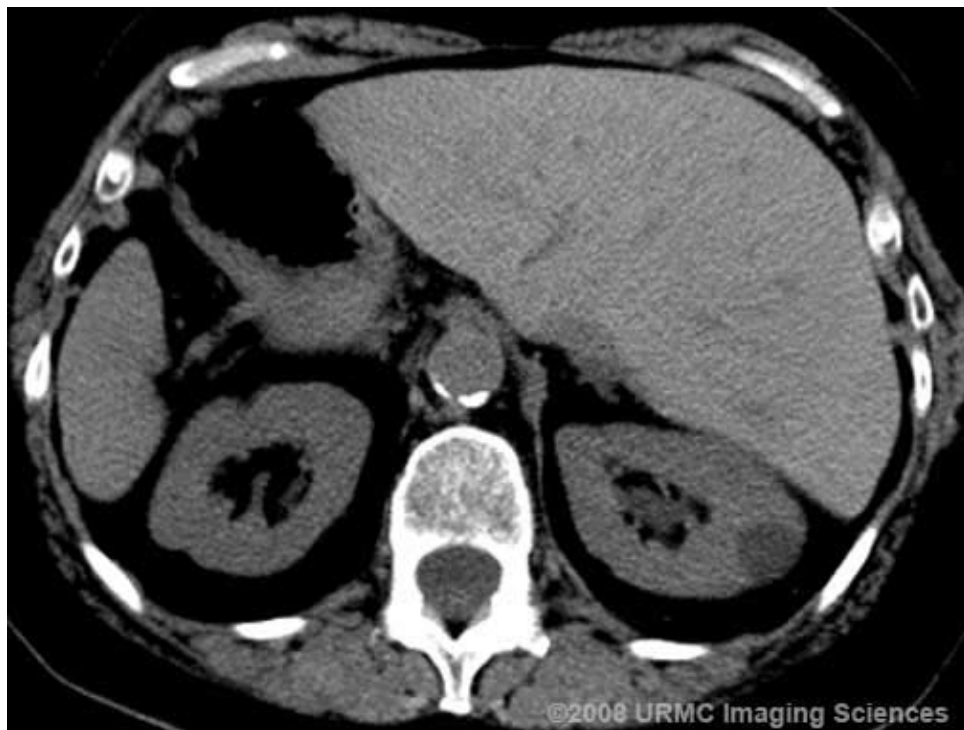
**IMAGING FINDINGS:**



**Figure 1:** There is bilateral basilar scarring, fibrosis and bronchiectasis. Hyperinflation of the lungs and bilateral interstitial thickening are also noted.



**Figure 2:** Extensive, severe, cystic bronchiectasis with bronchial air-fluid levels. Mucus filled bronchi in the lower lobes.



**Figure 3:** Situs inversus.

**DIAGNOSIS: Kartagener syndrome**

**DISCUSSION:** Kartagener syndrome or dyskinetic cilia syndrome (DCS) is an autosomal recessive disease

that produces an abnormality in ciliary structure which results in abnormal ciliary motion characterized by the clinical triad of chronic sinusitis, bronchiectasis, and situs inversus. Male infertility can also occur. The incidence of this genetical disorder is estimated about 1/25,000.

The defective cilia lining the respiratory tract are unable to clear the secretions and pathogenic bacteria, resulting in mucus retention and chronic or recurrent respiratory tract infection leading to damage to airways walls. Patients with Kartagener syndrome typically present in childhood with symptoms related to bronchitis, sinusitis and rhinitis. Bronchiectasis usually develops in childhood and young adulthood and it is associated with recurrent pneumonia with a predilection for the right middle lobe and both lower lobes. For fertility, most men with this syndrome have living but immotile spermatozoa and are infertile. Women likewise have decreased fertility, with fewer than 50% successfully completing pregnancy. Situs inversus is found in 50% of the patients with this syndrome.

Radiographic findings include:

- \* dextrocardia and situs inversus
- \* paranasal sinusitis
- \* bronchiectasis
- \* bronchial wall thickening
- \* segmental atelectasis
- \* hyperinflation, and
- \* recurrent pneumonia

CT findings include:

- \* bronchial wall thickening
- \* signet ring sign: dilatation of the bronchi
- \* variable severity of the bronchiectasis: cylindrical, varicose, cystic bronchiectasis
- \* bronchiolectasis: Tree-in-bud, V and Y shaped peripheral centrilobular opacities
- \* peribronchial or confluent airspace opacities representing pneumonia
- \* area of decreased attenuation suggest small airway disease
- \* diffuse centrilobular small nodules up to 2 mm in diameter, and
- \* situs inversus

#### **REFERENCES:**

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3. Afzelius BA, Bergström S-E. Primary ciliary dyskinesia (immotile-cilia syndrome). Uptodate.