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CLINICAL IMAGE

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Persistent cough and situs inversus in a middle-aged female

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Key message
Kartagener syndrome, a rare genetic disorder, can present in adults with persistent respiratory symptoms and radiological changes, such as bronchiectasis and situs inversus. Clinicians should maintain a high clinical suspicion, as early recognition and appropriate management are crucial for preserving pulmonary function.

KEYWORDS
bronchiectasis, Kartagener syndrome, persistent cough, situs inversus

CLINICAL IMAGE

A 54-year-old female presented with persistent productive cough and a history of recurrent pneumonia. The patient's remote location resulted in limited access to specialist care, leading to outpatient antibiotic treatment without a thorough evaluation. She also had chronic sinusitis and recurrent otitis. Computed tomography revealed situs inversus, bronchiectasis, centrilobular nodules and tree-in-bud pattern (Figure 1). During bronchoscopy, copious mucopurulent secretions were observed. Sputum cultures revealed pseudomonas aeruginosa in multiple sessions, and prompt antibiotic treatment and bronchial washing were initiated. Notably, the patient has undiagnosed infertility and has not had any children.

Primary ciliary dyskinesia (PCD) is a genetic disorder characterized by congenital impairment of mucociliary clearance. Most patients present in childhood, but some present in adulthood. Kartagener's syndrome is a subset of ciliary motility disorders and has a prevalence of around 1 in 20,000 to 40,000 individuals, and it compromises a triad of situs inversus, bronchiectasis, and sinusitis.[1] While the central characteristic of PCD involves an inherited cilia error, a universally accepted gold standard diagnostic test has yet to be established. A clinical suspicion for PCD can be raised by noting early-onset persistent cough, chronic rhinosinusitis, otitis, and laterality defects such as situs inversus. In adults, it is crucial to consider PCD in males with dyskinetic spermatozoa and respiratory symptoms and in females who are infertile or subfertile without other explanations, especially when accompanied by respiratory issues. Despite the lack of a gold standard diagnostic test for Kartagener syndrome, several tests can be used to validate the diagnosis. These include assessment of ciliary ultrastructure and motion, nasal nitric oxide measurement, cell culture, and genetic testing. Definitive diagnosis relies on identifying ciliary abnormalities through high-speed videomicroscopy analysis or transmission electron microscopy. These specialized tests necessitate nasal or bronchial biopsies and are only obtainable at expert centers experienced in performing them.[2]

AUTHOR CONTRIBUTIONS

All authors were involved in the preparation of the manuscript.

CONFLICT OF INTEREST STATEMENT

None declared.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author, [N.K], upon reasonable request.

ETHICS STATEMENT

The authors declare that appropriate written informed consent was obtained for the publication of this manuscript and accompanying images.

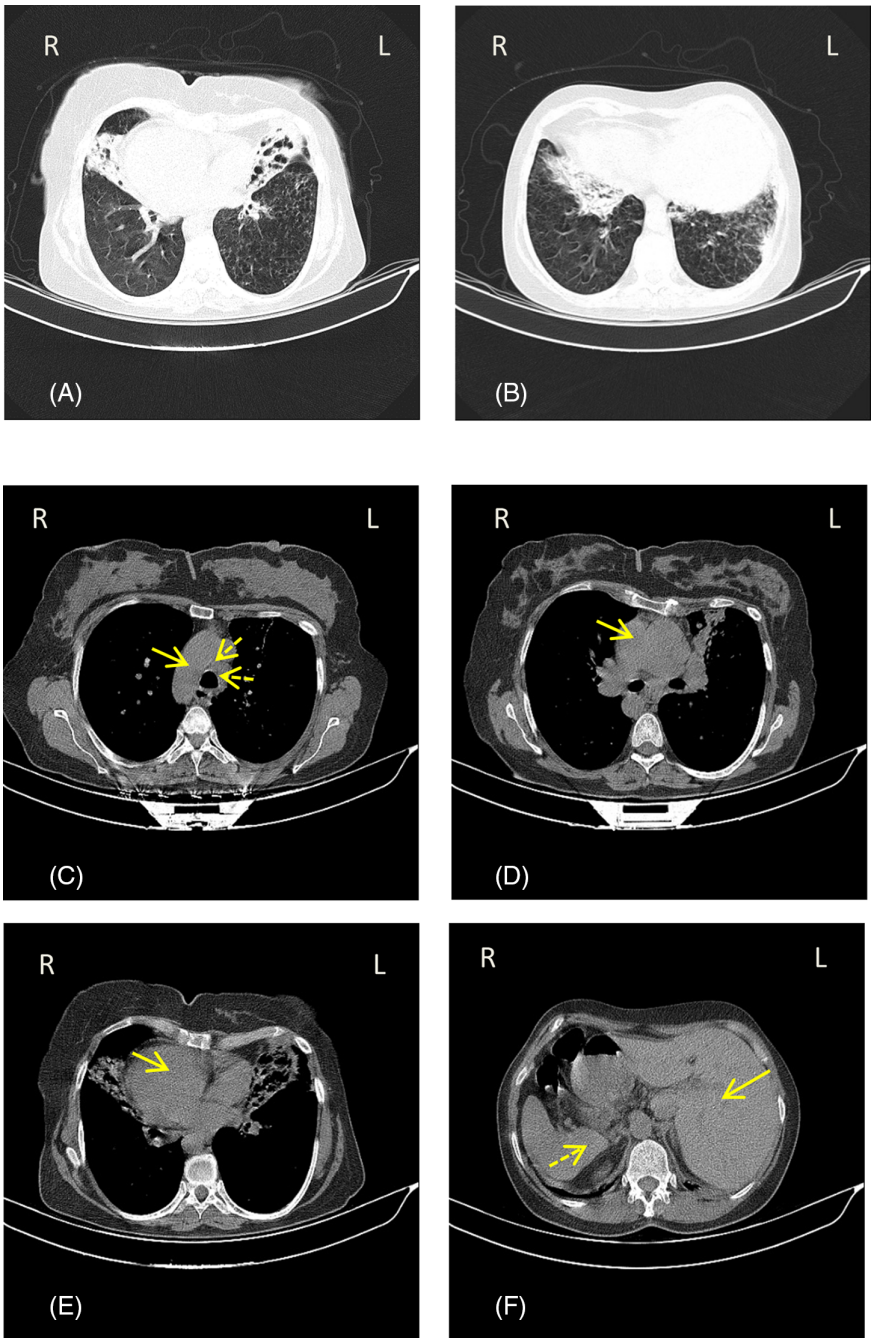

FIGURE 1 High-resolution computed tomography chest demonstrating, (A) situs inversus, tree in bud, bronchiectasis in the left middle lobe and right upper lobe, (B) tree in bud, hyperinflation, bronchiectasis and consolidation in the right lung, (C) Aortic arch in the right (arrow), azygos and brachiocephalic vein in the left (dashed arrows), (D) pulmonary artery trunk in the right (arrow), (E) dextrocardia (arrow), and (F) situs inversus, the liver in the left (arrow) and spleen in right (dashed arrow) part of the abdomen.

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