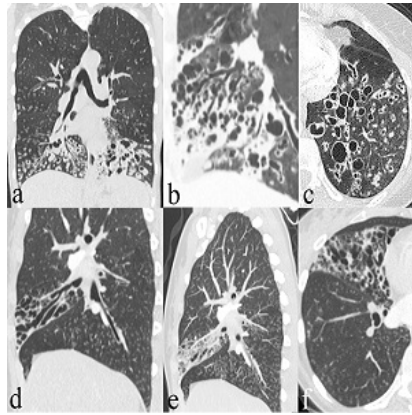




Typical features of primary ciliary dyskinesia on CT



Among patients with non-cystic fibrosis bronchiectasis, 1–18% have an underlying diagnosis of primary ciliary dyskinesia (PCD) and it is suspected that there is underrecognition of this disease. Researchers evaluating the specific features of PCD seen on computed tomography (CT) in the cohort of bronchiectasis found a predominance of bronchiectasis in the middle and lower lobes, severe tree in bud pattern, mucous plugging, and atelectasis in patients with PCD.

In addition, patients with other underlying diseases more often showed a predominance of bronchiectasis in the upper lobes as well as fibrotic and emphysematous changes of lung parenchyma.

"These findings may help practitioners to identify patients with bronchiectasis in whom further work-up for PCD is called for," the researchers explain.

PCD is an autosomal, recessive, inherited disorder resulting in ultrastructural defects of the ciliary apparatus with consequent abnormal or absent beating of cilia. This interferes with normal mucociliary clearance and causes repeated respiratory infections leading to airway damage and increasing the risk for the development of bronchiectasis. The diagnosis of PCD is complex and is based on suggestive clinical findings, e.g., a chronic wet cough and chronic rhinitis since infancy, a pathologic high-frequency video microscopy analysis (HVMA), an ultrastructural ciliary defect seen in transmission electronic microscopy (TEM) and/or immunofluorescence microscopy (IF). Results from these different tests are often inconclusive thus making the diagnosis difficult.

The study included 121 patients with bronchiectasis, 46 of them with PCD as the underlying disease. CTs performed in these patients were scored for the involvement, type, and lobar distribution of bronchiectasis, bronchial dilatation, and bronchial wall thickening. Later, associated findings such as mucous plugging, tree in bud, consolidations, ground glass opacities, interlobular thickening, intralobular lines, situs inversus, emphysema, mosaic attenuation, and atelectasis were registered. Patients with PCD ($n = 46$) were compared to patients with other underlying diseases ($n = 75$).

In patients with PCD, the extent and severity of the bronchiectasis and bronchial wall thickness were significantly lower in the upper lung lobes. The lobar distribution differed significantly with a predominance in the middle and lower lobes in patients with PCD. Significantly more common in patients with PCD were mucous plugging, tree in bud, atelectasis, and a history of resection of a middle or lower lobe. Less common were emphysematous and fibrotic changes. A situs inversus (Kartagener's Syndrome) was only seen in patients with PCD.

The research team said theirs "is the first study which shows that CT might be helpful, in addition to a patient's medical history, for identifying patients in the bronchiectasis cohort in which further work-up for PCD is warranted."

The major limitation of our study is the small number of patients who have undergone CT in PCD. "Therefore, further studies with more patients will be required," the researchers noted. "Nevertheless, PCD is a rare disease and this is the largest study addressing the use of CT for guiding the diagnosis in addition to a patient's medical history and clinical features to date."

Source: [PLOS One](#)

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