

Clinical and function manifestations of radiological bronchiectasis in ADPKD

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Background: Although increased prevalence of radiographic bronchiectasis in ADPKD has been reported, little is known about the clinical and functional manifestations of this association. We retrospectively explored the clinical and functional features of ADPKD-associated radiographic bronchiectasis in a selected cohort from a single large academic institution.

Materials and Methods: Clinical records of adult ADPKD patients evaluated between 2000 and 2010 at Mayo Clinic, Rochester, with chest computed tomography (CT), abdominal CT, and pulmonary function testing (PFT) were included in the study. Relevant clinical data were collected. Radiologic criteria for diagnosis of bronchiectasis included enlarged internal bronchial diameter, failure of airway tapering at least 2 cm beyond the last branch point, and airway presence within 1cm of the lung periphery. All CT scans were reviewed independently by two expert thoracic radiologists (RH and TH) and discordant findings settled by consensus. PFT data was categorized using standard criteria into one of six groups: obstructive, restrictive, mixed, non-specific, isolated decrease in DLCO and normal; by an experienced pulmonologist. Patients with known or suspected clinical causes of bronchiectasis (cystic fibrosis, ABPA, recurrent airway obstruction, etc.) were excluded. The Mayo Clinic IRB approved the study.

Results: Seventy one ADPKD patients with chest CT, abdominal CT, and pulmonary function testing were included in the study. Mean age was 60.2 ± 12.6 years (37 Female). The frequency of radiographic bronchiectasis was 19% (14 of 71) with a predominance of findings in the lower lobes bilaterally. The distribution of age, gender, eGFR and smoking history (current/former versus never) was not statistically different between ADPKD patients with and without bronchiectasis, 67 (range 32-84) versus 60 (22-88) years, $p=0.08$, 8 M/6F versus 26M/31F, $p=0.55$, 49 (42-74) versus 44 (19-63) mL/min/BSA, $p=0.18$, and 11/3 versus 30/27, $p=0.13$, respectively. PFT findings and clinical symptoms were similar between the two groups ($p=0.25$), with a predominance in both for normal and obstructive PFT results in both groups.

Conclusions: Our results suggest that despite the presence of radiologic bronchiectasis for some ADPKD patients, the structural abnormality does not correspond with increased respiratory symptoms, pulmonary diagnosis and PFT abnormalities. Future study with larger number of ADPKD patients is necessary to confirm our findings.

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