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Case Report

Coexisting cystic lung disease as a rare extra-renal manifestation of autosomal dominant polycystic kidney disease

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ABSTRACT

Autosomal dominant polycystic kidney disease (ADPKD) classically presents with multiple bilateral renal cysts and ultimately progresses to end stage renal disease. While many of the extra-renal manifestations of ADPKD are well-documented, associated pulmonary findings are particularly rare, having only been recently been reported in a handful of studies to date. A 69-year-old female with ADPKD presented to our hospital with respiratory complaints. High resolution computed tomography revealed bronchiectasis, cystic lung disease, and interstitial fibrosis. The patient did not have concurrent risk factors or coexisting disease processes to explain the etiology of her airway and cystic lung disease, which we suggest are manifestations of ADPKD. We have not found a previous report of interstitial lung disease in this setting.

1. Introduction

Autosomal dominant polycystic kidney disease (ADPKD) is a systemic disease affecting approximately 500,000 people and accounts for 5–10% of the dialysis population in the United States alone [1,2]. Predominantly characterized by multiple large bilateral renal cysts, ADPKD typically presents in the third or fourth decade of life. As the renal cysts grow and distort the normal renal parenchyma, progression to end stage renal disease results with an incidence of about 50% by 60 years of age [1,2].

Extra-renal manifestations of ADPKD are prevalent, including cysts affecting the liver, pancreas, central nervous system, and genitourinary tract [1–4]. While many of these extra-renal manifestations have been well documented, until recently, there has been little mention of coexisting pulmonary pathologies [3–8]. In the few reported cases of synchronous lung pathology, the most common pulmonary manifestation of ADPKD has been bronchiectasis with even fewer case reports describing associated pulmonary cysts [4,6,8]. To date, concurrent interstitial lung disease has not been linked to ADPKD nor has it been reported in the literature. We herein report a rare case of airway and cystic lung disease as the pulmonary manifestation of ADPKD as well as the first reported case of coexisting interstitial lung disease in this setting.

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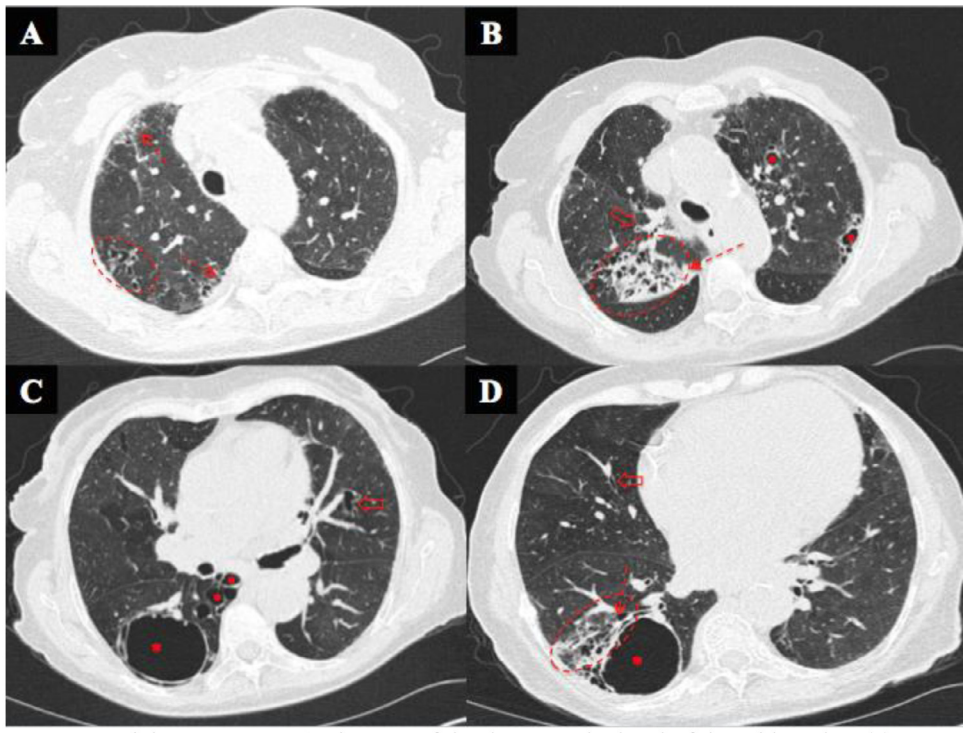

Fig. 1 – Axial non-contrast computed tomography images of the thorax at the level of the mid trachea (a), distal trachea (b), mainstem bronchi (c), and in the lower lung zones (d) show scattered areas of interstitial fibrosis (dashed arrows), honeycombing (dashed ovals), cylindrical and cystic bronchiectasis (open arrows), and pulmonary cysts (asterisks).

2. Case report

A 69-year-old woman with a history of ADPKD, end stage renal disease on dialysis, hypertension, and diabetes mellitus presented to our outpatient lung center with complaints of dyspnea, wheezing, and a worsening cough with yellow sputum production for several weeks. The patient denied any smoking history, inhaled drug use, or inhaled allergen exposure. There was no known history of pulmonary infection. At the time of admission, aside from an elevated blood pressure of 160/69, the patient's vital signs were within normal limits. Physical examination demonstrated crackles throughout both lungs with bronchial breath sounds noted at the right lung base. Laboratory data were unremarkable. Spirometry demonstrated an FVC of 1.56 L (62% of predicted), FEV_1 of 1.14 L (59% of predicted), FEV_1/FVC of 73% of predicted, none of which were significantly changed with bronchodilator treatment, and a TLC of 3.44 L (78% of predicted), overall in keeping with a very mild restrictive pattern.

Non-contrast high resolution computed tomography imaging of the chest was performed and demonstrated numerous cysts of varying sizes throughout both lungs (Figs. 1 and 2). Findings at the lung bases appeared grossly unchanged when compared to the prior computed tomography (CT) abdomen 9 years prior, but no comparative imaging was available for the remainder of the pulmonary parenchyma (Fig. 3). The largest cyst was irregularly shaped and located in the right lower lobe, measuring approximately 5.7 × 5.6 × 8.0 (AP x TV x CC) without air–fluid levels (Figs. 1 and 2). Several areas of cylindrical and cystic bronchiectasis without a lobar distribution with coexisting bronchial wall thickening and air trapping were identified (Figs. 1 and 2). In addition, predominantly subpleural reticulation, traction bronchiectasis, and honeycombing were found in the dependent portions of both lungs, consistent with interstitial fibrosis (Figs. 1 and 2). There was no significant change in the imaging appearance of this patient's pulmonary findings when compared to the study from several years prior. Within the visualized upper abdomen, numerous peripherally calcified hepatic and renal cysts were identified in keeping with the patient's known diagnosis of ADPKD (Fig. 4).

The patient was subsequently started on a short course of oral antibiotics and was recommended to continue her inhaled bronchodilators. An acute pulmonary infectious etiology was considered unlikely given the clinical, laboratory and imaging findings; therefore imaging findings were attributed to pulmonary sequela of ADPKD.

3. Discussion

ADPKD is the most common form of polycystic kidney disease, with an incidence between 1:500 to 1:1000, and classically presents in the third and fourth decades of life. Renal manifestations of ADPKD have been well characterized by the development of large bilateral renal cysts. As the cysts grow and distort the normal healthy renal parenchyma, pa-

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