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On the Cover

What's the diagnosis?

A 70-year-old woman from Nepal presents for initial evaluation of chronic kidney disease. She reported being diagnosed with "hereditary" kidney disease years earlier in Nepal despite no family history. Past medical history was otherwise notable for pulmonary hypertension, atrial fibrillation, and pre-diabetes. On examination, blood pressure was 115/71 mm Hg; there was no CVA tenderness or nephromegaly. Serum creatinine was stable at 1.7 mg/dl over several measurements. Urinalysis was bland with 0.5 g of non-albumin proteinuria.

Image Description:

Computed tomography with renal mass protocol was performed to further evaluate the presence of renal cysts noted on the screening ultrasound (left, coronal and right, axial). The left kidney was noted to be near-completely replaced by large confluent cysts, with some of them showing milk-of-calcium deposition. There was no communication with the collecting system, which appeared distorted and slightly dilated, especially in the left upper collecting system. There was no obstructing calculus or hydronephrosis. The right kidney was noted to be normal. There were no other cysts noted in the abdomen.

Teaching Points:

Localized cystic disease is an uncommon presentation of renal cystic disease and is sometimes confused with autosomal polycystic kidney disease, multilocular cystic nephroma, and cystic neoplasm. It is characterized by extensive unilateral cystic disease, is a benign condition, and usually does not lead to clinically apparent kidney disease because the contralateral kidney is normal. Unlike the more common bilateral cystic condition, autosomal dominant polycystic disease, localized cystic kidney disease is not heritable and does not affect other areas of the body outside of the kidney. The pathogenesis of localized cystic kidney disease is unknown and may represent an acquired condition.

(Images and text provided by Sarah Struthers, Scott Bieber, and Achille Mileto of University of Washington, Seattle, Washington)