1. Clinical Key Message

The incidence of renal cell carcinoma (RCC) in patients with polycystic kidney disease (PCKD) has shown to be higher compared to the general population [1, 4]. However the incidence of RCC in young individuals (<40 years old) is extremely rare compared to older patients (>40 years old) with the genetic disease. A far more common complication of PCKD is renal cyst infections [1]. The clinical presentation is often ambiguous and radiological imaging can lead to misdiagnosis. A biopsy of the lesion is required for definitive diagnosis and targeted treatment.

2. Key words
Polycystic kidney disease; Renal cell carcinoma; Renal biopsy; Renal cyst infection

3. Case

A 24-year-old African American man with history of polycystic kidney disease, autism and schizophrenia was admitted to our hospital for nausea, vomiting and one episode of mild hematuria for 1 week. He was afebrile at presentation. Urinalysis was negative. There was no leukocytosis on admission. His renal function tests were within normal limits. CT scan (with contrast) of the abdomen was acquired showing innumerable bilateral renal cysts (Figure 1). One of which measured 4.5 x 5.2 cm as a heterogeneous mass with epicenter in the superior pole of the left kidney showing extension into renal hilum. This was most concerning for RCC.

The patients was discharged after overnight resolution of hematuria with fluids and scheduled for outpatient follow up CT (with contrast) scan and biopsy. Two months later the patient followed up with urology. At the time, he was afebrile and did not report any symptoms. Prior to biopsy, re-imaging of the cyst demonstrated decrease in size from 4.5 x 5.2 cm to 2.5 x 2.8 cm. On re-evaluation by the urologist, the decrease in size of the cyst warranted further monitoring. The biopsy was postponed to monitor the cyst size. The patient continued to follow up with urology and 6 months later biopsy resulted was negative for RCC.

4. Discussion

Renal cyst infection is a common complication in patients with PCKD. Presentation tends to include abdominal pain and fever, however some patients are asymptomatic [2]. A diagnosis of cyst infection includes positive culture of cyst fluid and/or presence of neutrophil debris. In a clinical setting, a definitive diagnosis is difficult to obtain, physicians mostly rely on clinical features (abdomen pain, fever, hematuria, and positive urine cultures) [3]. Although imaging is obtained, it rarely is reliable. One study found ultrasound, CT and MRI failed to detect cyst infections on average of 78% [2]. PET (positron emission tomography) was found to be the most reliable method of detecting infection within renal cysts [2]. Infected cysts account for significant morbidity in PCKD; therefore biopsy is necessary for identification of the infection [3].

Renal cyst infections are largely due to ascending infection caused by gram-negative bacteria (most commonly E. coli). The standard of care is treatment with antimicrobials such as fluoroquinolones and third generation cephalosporin [3]. Larger cysts (>5 cm) may require surgical drainage in addition to antibiotics [3].

*Corresponding Author(s): Sonya Patel, Department of Surgery, Memorial Health University Medical Center, Savannah, GA, USA, E-mail: sonyakanti@me.com

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The role of biopsy is consisting determined by the size of renal mass. Surgical data demonstrates evaluation of renal mass that deceases in size is more likely benign than malignant. Therefore biopsy may be postponed in selected patients in order for “watchful waiting” [5]. High clinical suspicion is necessary in patients with PCKD presenting with abnormal renal mass for adequate management.

References

Figure 1. CT of abdomen without contrast exhibits 4.5 x 5.2 cm heterogeneous mass with epicenter in the superior pole of the left kidney with extension into renal hilum.

Figure 2. Follow up CT with contrast displays decrease in cyst size to 2.5 x 2.8 cm.