European Journal of Molecular & Clinical Medicine

ISSN 2515-8260 Volume 08, Issue 03, 2021

Adult Renal Cystic Disease: A Comprehensive Literature Review

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Abstract

Renal cystic disease occurs as a result of cysts of varying sized localized in an enlarged kidney. It can be classified into different forms such as autosomal-dominant polycystic kidney disease, autosomal-recessivepolycystic kidney disease, unilateral renal cystic disease, renal simple cysts, multicystic dysplastic kidney,pluricystic kidney of the multiple malformation syndromes and medullary cystic disease. Recent studies showed that there is a relation between inherited cystic disease and changes in a group of genes involved in the formation and function of bothprimary cilia of the embryonic node and cilia in epithelial renaltubes. On the other hand, cysts in acquired conditions occur as a result ofobstructive, stromal–epithelial malinductiveand neoplastic mechanisms. Moreover, several genetic and acquired renal cystic diseases were found to be linked to the development of renal cellcarcinoma. There are different approaches to diagnose various types of renal cysts, while ultrasound and slice imaging are the main tools for the diagnosis of simple renal cysts, Bosniak classification was selected as the basic method for the diagnosis of renal cystic masses larger than 1 cm. In the current review we will discuss the various types of renal cystic diseases and the main diagnostic approaches.

Keywords*: Kidney; Renal cyst; Renal carcinoma; Diagnosis*

1. Classification of renal cystic diseases

1.1. Autosomal-dominant polycystic kidney disease

Autosomal-dominant polycystic kidney disease (ADPKD) is classified as the most common genetically basedrenal cystic disease, having the rate of occurrence of1 to 2 cases per 1000[1]. Two genes were found to be incorporated in ADPKD these genes arePKD1 and PKD2. PKD1 gene is sited on chromosome 16and codes for polycystin-1, a protein that localized the primary cilia of renal epithelial cells and take partin cell-cycle regulation in addition to intracellular calcium transport. Thesecond gene, PKD2gene is sited on chromosome 4 and codes for polycystin-2, a protein that also localized the primary cilia of renal epithelial cells and recognized as a member of the family of voltage-activated Ca^{+2} channels[2].

Regarding age distribution, ADPKD might affect people at any age, but mostly during the4thand 5th decades, therefore it was classified as an adult type of hereditary polycystic kidney whereany

portion of the nephrons orcollecting ducts can be influenced[3]. For the percentage of affected nephrons or collecting ducts, it was reported that although every cell of these partscarries the PKD1 or PKD2germline gene mutation, only one to two percentage of the nephrons orcollecting ducts are affected where nephrons with disruptionof a 2nd allele experienced cystic enlargement. Cystogenesis induction occurs through a second hit, somatic mutation,incorporating eitherthe PKD1 or PKD2 gene, apart from the type ofgermline mutation [4].ADPKD (figure.1) manifests with innumerable,round cysts with a various size from only 0.1 cm to severalcentimeters, and their distribution was even through the renal cortex and medulla [5].

Attention must be attributed to the difference between polycystic and multicystic as the former refers to ADPKD while the later refers to a non-inherited cystic kidney condition with the presence of cartilage tissue and abnormal mesenchyme [6].

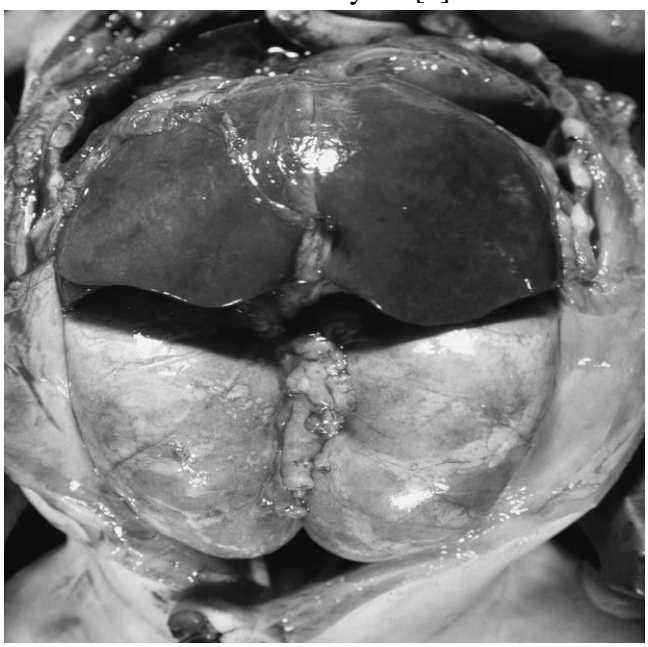

Figure1. Striking abdominal enlargement due to autosomal dominant polycystic kidney

1.2. Unilateral renal cystic disease

Unilateral renal cystic disease (URCD)(figure 2) is a rare renaldisorder showing no progression and was demonstrated to have no association with otherorgans cystsin addition to having no relation with other genetic cystic diseases. Moreover, pathogenesis of this diseaseis obscure where most caseswere diagnosed during adulthood[7].Common reported clinical manifestations of URCDare abdominal pain, a palpable mass, gross hematuria in addition to hypertension with well-preserved renal function[8].The process of URCD management is conservative where follow up with functional studies and imaging surveillance is the followed approach [9]. Analyzing the relation between URCD and renal carcinoma showed no correlation between both events [10].

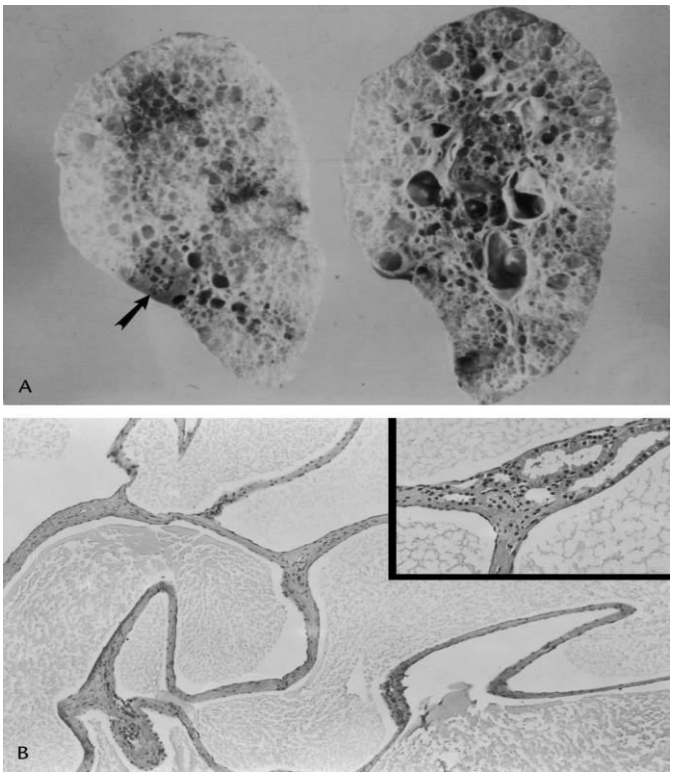

Figure2. Unilateral renal cystic disease; A- Two frontal views of the same half of the kidney, B- Cystically dilated tubules.

1.3. Renal simple cysts

Renal simple cyst either in mono (figure 3) or multiple form is consideredthe most frequent cystic disease in adult and elderly patients. The cortexis considered the most common location for this type of cysts where it lines the cortex epithelial cells. The major difference between simple and acquired cysts is that the simple one occurs in non-diseased kidney while the later one occurs to patients suffering from chronic renal problems[11].Regarding its general and age distribution, this kind of cysts affects about five percentage of population who performs abdominal ultrasound examination for whatever complicationsand this number reaches about twenty percentage for cases having 40 years old and again the percentage increases to thirty three percentage for cases older than 60 years old.With patient age progression, simple cysts shows an increase in number and size therefore accurate diagnosis is essential to differentiate it from malignancies [12].

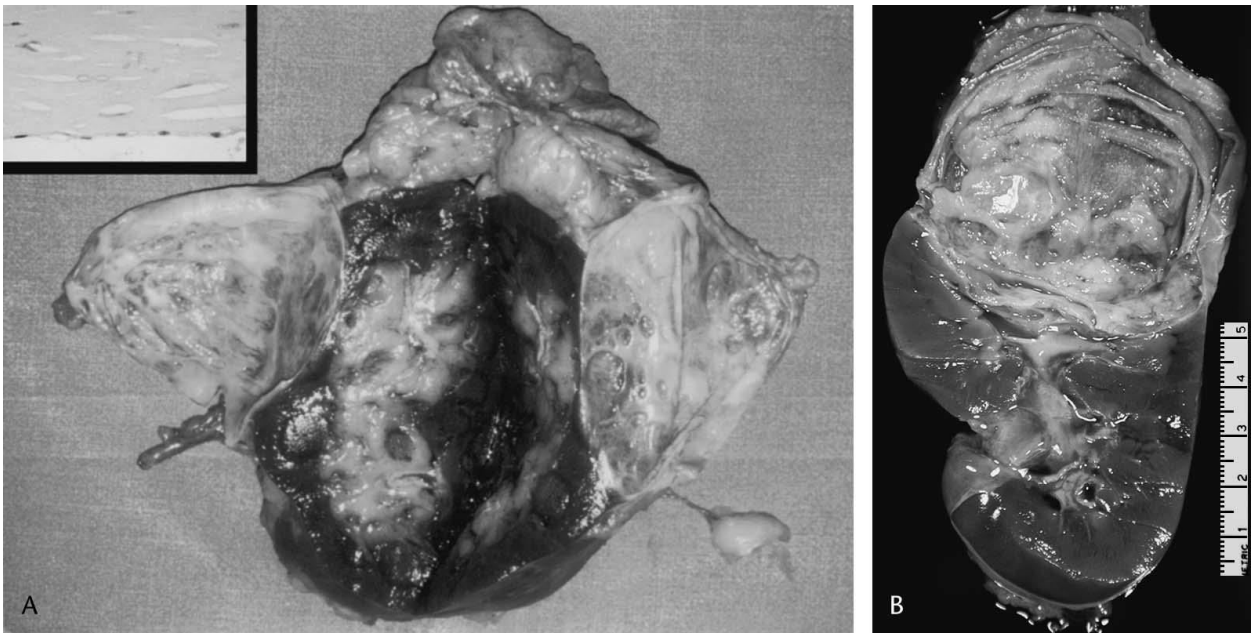

Figure3. Solitary unilocular renal simple cyst

1.4. Pluricystic kidney

This term was given to the cases with multiplerenal cysts that can be either heritable or non-heritable extra-renalsyndromal anomalies.This kind of disease has different macroscopical and microscopical manifestations in various syndromes[13].

2. Renal cystic diseases and renal neoplasms

Renal cell carcinomas (RCCs) ranges between 2 and 3 percentage of total tumor count in the United States where men suffer from this kind of tumor in a rate more than 1.6 times against women. Regarding geographical distribution, people in industrialized countries are more susceptible to RCCs as risk factors like smoking and industrial compounds are more common in these countries[14].

Moving to the point of RCCs and renal cystic diseases, researches and studies through the past few years demonstrated a positive correlation between RCCs and kidney cysts where cases with end-stage kidney disease and acquired cystic kidneydisease were found to possess various types of renal carcinoma (table 1). Close understanding of the molecular basis of kidney cysts and the progression of various types of renal carcinoma would put our hand on novel approaches for therapy rather than surgical and other traditional solutions [15].

Table1. Cystic renal diseases with risk of RCCs

Cystic disease	Mutated gene	Cancer risk (%)
ADPKD	*PKD1,PKD2*	Not known
von Hippel-Lindau disease	*VHL*	60%
Tuberous sclerosis complex	*TCS1,TCS2*	2-3%
acquired cystic kidneydisease	Diverse	5-10%

3. Classification system for renal cystic masses and their risk of malignancy

Ultrasound is considered the basic diagnostic procedure for simple renal cysts. Benign simple renal cyst has certain criteria regarding size and progression and at the same time other diagnosed cysts that do not meet these criteria can be classified as atypical complex. During the past few years, the Bosniak classification (table 2) was considered as the basic approach for diagnosing cystic lesions larger than 1 cm. This classificationrelied onthe outcome of computed tomography (CT) protocol[16].

Table2. Bosniak classification of kidney cysts

Category	Diagnosis	Risk of malignancy (%)
Bosniak I	Simple cyst	0
Bosniak II	Complicated cyst	0
Bosniak IIF	Complicated cyst	5-15
Bosniak III	Complicated cyst	50-60
Bosniak IV	Cystic cancer	90-100

4. Conclusion

In the current study we gave a brief review on the basic types of renal cysts that affect adults with a presentation of their causes, manifestations and management. Moreover, we showed the positive correlation between various types of renal carcinoma and renal cysts as a major causative agent. Finally, the review showed the basic diagnostic approaches followed for renal cyst.

5. References

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