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Radiological Imaging Diagnosis of Adult Renal Cystic Diseases and Management

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Abstract

Background Renal cysts are a heterogonous group of cystic renal diseases that present in approximately 40% of the patients undergoing imaging.

Objective To describe the role of the radiological imaging methods for the diagnosis of adult renal cystic diseases and their management.

Methods A descriptive study carried out for 218 patients of renal cystic diseases in adults in two Private Hospitals in Aden Province during the period from February 2006 to December 2014, diagnosed by ultrasonography, contrasted urography and computerized tomography determined the age, gender, frequency, and complications and type of treatment.

Results Autosomal dominant polycystic kidney disease (ADPKD) presented in 96 patients (44%), simple renal cysts 90 (41%), medullary sponge kidney 25 (11%), localized cystic renal disease (LCRD) six (3%) and adult multilocular cystic nephroma one (1%). Ultrasound correctly interpreted 179 patients (82%), nonvisualized 25 (11%), and inconclusive 13 (7%) contrasted urography correctly interpreted 176 (76%), nonvisualized 13 (6%) and inconclusive 19 (9%) and computerized tomography (CT) scan 199 patients (91%), six (3%) and 13 (6%) respectively. Complications occurred in 65 patients (68%) of ADPKD, 23 (25%) of simple renal cysts, 15 (60%) of medullary sponge kidney (MSK) and three patients (50%) of LCRD. Conservative treatment performed in 171 (78%) and surgical intervention in 47 patients (22%).

Conclusion The commonest renal cystic disease is adult dominant polycystic kidney. Ultrasound is inconclusive for renal cystic masses. CT scan is the effective imaging study for localized and multifocal renal cystic diseases, complications and associated pathology. The frequent complications occur in adult dominant polycystic kidney disease and medullary sponge kidney.

Keywords Adults, renal cystic diseases, imaging, management

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List of abbreviations: ADPKD = Autosomal dominant polycystic kidney disease, CT = Computerized tomography, LCRD = Localized cystic renal disease, US = Ultrasonography, UTI = Urinary tract infection

Introduction

Renal cystic disease of the kidney in adults comprises a wide spectrum of hereditary, acquired, developmental and acquired conditions; they are a common cause of high morbidity and mortality worldwide [(1)]. Inherited cystic diseases in adults are linked to various genes involved in the formation and functioning of the primary cilia of the renal tubular epithelium. Dysfunction of primary tubular cilia leads to increased tubular epithelial proliferation and excessive fluid secretion. Nonhereditary renal cystic diseases are secondary to obstructive, stromal-epithelial mal-inductive and neoplastic mechanisms [(2-4)]. They may complicate by hemorrhage, infection

and ischemia, which produce a difficulty to be differentiated from renal cell carcinoma [5,6]. Clinically present with flank, back and abdominal pain, hematuria, renal swellings and hypertension [7]. Adult dominant polycystic kidney is the most common inherited cause of end-stage renal failure with more than 45% of patients requiring dialysis or transplantation by the age of 60 years. Implementation of genetic, cellular, and molecular techniques made accurate diagnosis, and carrier detection [8]. Actually, the use of various imaging diagnostic studies plays an important role in the diagnosis and management, detects their complications, disease progression and the response for treatment [9]. The use of Bosniak classification by computerized tomography (CT) scan findings differentiated by their attenuation, contrast enhancement, presence of calcification and septation differentiates benign from malignant cystic lesions, guides patient management and determine nonsurgical from surgical lesions (10). The management of renal cysts is best done with a multidisciplinary team that includes a urologist, nephrologist, geneticist and an internist with the assistance of a nurse practitioner or physician assistant facilities their management [11].

The main objective is to describe the importance of radiological diagnosis of adult cystic renal diseases and their complications.

Methods

A descriptive retrospective study carried out for a total of 218 adult patients with congenital and developmental cystic renal diseases including 117 men (54%) and 101 women (46.3%), during the period from February 2006 to December 2014 in two Private Hospitals in Aden Province named "Al Naquib and Al Mansorah". Age groups range from 18 to 70 years (mean age 52 years). A detailed history and thorough physical examination and abdominal ultrasonography, contrasted urography and CT scan. Clinically presented with abdominal, flank and back pain in 105 patients (48%), hematuria 33 (15%), recurrent urinary tract infection (UTI) 53 (24%) and abdominal palpable masses 27 (12%) respectively, determining the frequency of each adult renal cystic diseases, excluding the children and acquired renal cystic disease and those treated by endoscopic procedures, recognize the associated renal anomalies, complications and type of treatment whether medical or surgical.

Data retrieved from medical outpatient records and Hospitals registries. Ethical clearance was obtained and patients identifiers were not included and non-exposure of organs. Imaging Figures only to be used only in the scientific field. Data was processing using SPSS 21 version. Independent samples t-test was performed to determine the significance among variables.

Results

Congenital and developmental renal cystic diseases in adults presented in 117 men (54%) and 101 women (46%). Age group were as follows (18-30 years) eight patients (4%), (31-40 years) 33 (15%), (41-50 years) 48 (22%), (51-60 years) 63 patients (29%) and (61-70 years) 66 (30%). Frequency of renal cystic diseases included adult dominant polycystic kidney disease (ADPKD) 96 patients (44%), simple renal cyst 90 (41%), medullary sponge kidney (MSK) 25 (11.5%), localized cystic renal disease (LCRD) six (3%) and one patient 1% with adult multilocular cystic nephroma (AMCN) (Table 1).

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Table 1. Adult renal cystic diseases regarding to age, gender and frequency in two Private Hospitals in Aden Province

Age group (years)	Males No. (%)	Females No. (%)	Total No. (%)	Frequency	No. (%)
18-30	5 (62)	3 (37)	8 (4)	ADPKD	96 (44)
31-40	15 (46)	18 (55)	33 (15)	SRC	90 (41)
41-50	27 (56)	21 (44)	48 (22)	MSK	25 (11)
51-60	33 (52)	30 (48)	63 (29)	LCRD	6 (3)
60-70	37 (56)	29 (44)	66 (30)	AMCN	1 (1)
Total	117 (54)	101 (46)	218 (100)	Total	218 (100)

ADPKD = Adult dominant polycystic kidney disease, SRC = Simple renal cyst, MSK = Medullary sponge kidney, AMCN = Adult multilocular cystic nephroma

Complications in ADPKD including hypertension 27 (41%), recurrent UTI 21 (32%), stones seven (11%), renal failure seven (11%) and renal cell carcinoma three (5%) with a total of 65 patients (68%). Simple renal cysts hypertension presented in seven patients (30%), UTI nine (39%), stones four (17%), malignant tumors two (9%), and renal failure one (4%) with a total of 23 patients (24%). Medullary sponge kidney hypertension found in two patients (13%), UTI seven (47%), stones five (33%), and renal failure one (7%) for a total of 15 patients (60%). Localized cystic renal disease, hypertension presented in one patient (33%), UTI one (33%), tumors one (33%) with a total of three (50%) of patients (Table 2).

Table 2. Patients with adult renal cystic diseases regarding to complications in two Private Hospitals

Cystic renal diseases	Hypertn-sion No. (%)	Recurrent UTI No. (%)	Stones No. (%)	Tumors No. (%)	Renal failure No. (%)	Total No. (%)
ADPKD	27 (41)	21 (32)	7 (11)	3 (5)	7 (11)	65 (68)
SRC	7 (30)	9 (39)	4 (17)	2 (9)	1 (4)	23 (25)
MSK	2 (13)	7 (47)	5 (33)	0 (0)	1 (7)	15 (60)
LCRD	1 (33)	1 (33)	0 (0)	1 (33)	0 (0)	3 (50)

ADPKD = Adult dominant polycystic kidney disease, SRC = Simple renal cyst, MSK = Medullary sponge kidney, AMCN = Adult multilocular cystic nephroma

Ultrasound carried in all patients, correctly interpreted renal cystic diseases 179 patients (82%), inconclusive 25 (11%) and non-visualized 13 (6%). Contrasted urography carried out in 209 patients, correctly interpreted 176 (82%), nonvisualized, 13 (6%), and inconclusive 29 (14%). CT scans, correctly interpreted in 199 (91%), nonvisualized six (3%) and inconclusive in 13 (6%) (Table 3). CT scan was the most effective imaging method for the diagnosis of renal cystic diseases in adults. Conservative treatment carried out in 171 patients (78%) and surgically intervened 47 (22%).

Table 3. Adult congenital cystic renal diseases according to results of imaging in two Private Hospitals in Aden Province

Imaging study	Correctly interpreted No. (%)	Nonvisualized No. (%)	Inconclusive No. (%)	Total No. (%)
Ultrasound	179 (82)	25 (11)	13 (7)	218 (100)
Contrasted urography	176 (76)	13 (6)	19 (9)	209 (100)
Contrasted CT scan	199 (91)	6 (3)	13 (6)	218 (100)

Discussion

The most common adult congenital and developmental renal cystic diseases included ADPK, followed by simple renal cysts (SRCs), medullary sponge kidney (MSK) and localized cystic renal disease (LCRD) respectively and adult multicystic nephroma was the rarest one consisting with reported [12-14].

Complications were more common in ADPKD and MSK. ADPKD was the most common cause of hypertension and end-stage renal failure. MSK was complicated by urinary stones and recurrent UTI Cystic dilatation of collecting tubules producing urinary stasis, which contribute to UTI and stones [15-17].

This study presented unilateral ADPKD associated with renal agenesis seen in three patients; one of them was an adult woman with right renal agenesis associated with incomplete duplications of left kidney with upper pole normal function, and lower pole hydronephrosis (Figure 1).

Figure 1. IVU shows right renal agenesis and double left pelvicalyceal system and lower pole hydronephrosis in ADOKD

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LCRD presented in six patients. It is a rare benign non-progressive, non-surgical unilateral multiple renal cyst characterized by conglomerate mass of multiple simple renal cysts replacing a variable portion or an entire kidney separated by normal or atrophic renal tissue. Relatively few cases described in literature [18-23].

Multilocular cystic nephroma in adult diagnosed in one patient. Multlocular cystic nephroma is a rare finding and few cases reported in literature. In CT scan typically appears as a well-circumscribed encapsulated multicystic hypodense cyst of variable size which is separated by hyperechogenic septae (Figure 2). It is benign nonhereditary unilateral hypoechoic renal cyst lined by an epithelium and fibrous hyperechogenic septae contain mature tubules [24-26].

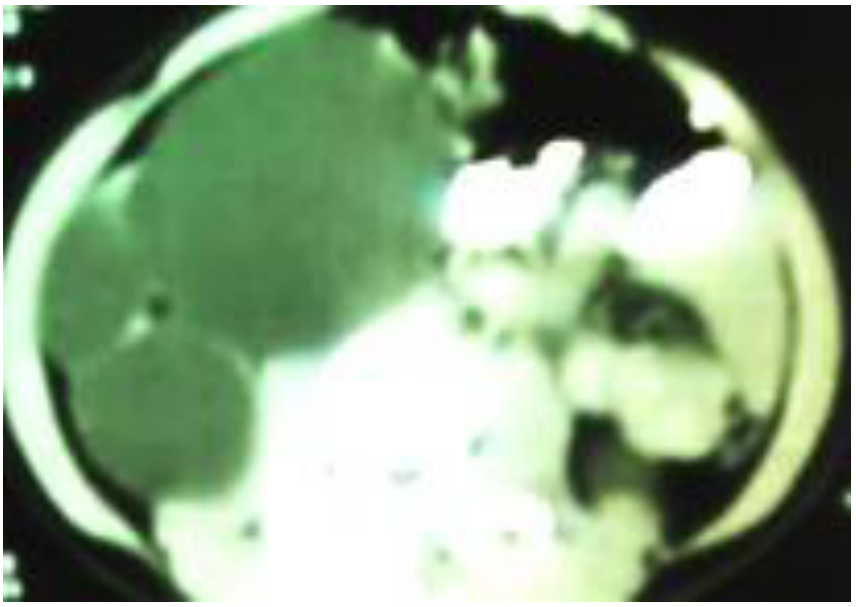

Figure 2. CT scan reveals hypoechoic renal cysts of different sizes separate by hyperechogenic septae diagnostic of adult multilocuar cystic nephroma

Ultrasonography (US) is the first study for assessment of cysts location and size of both kidneys but is inconclusive for cystic renal masses evaluation and ectopic kidneys. It is an excellent study for determination of cysts in other organs and follow-up. Pei et al. in 2015 [17] considered US as the gold standard for the diagnosis of ADPKD. Simple renal cyst is seen as a round or oval hypoechoic structure surrounds by a thin and smooth wall, while multiple bilateral renal cysts at the age of 20-30 years with large kidneys and extrarenal cysts most commonly seen in the liver confirms ADPKD.

Intravenous urography is an effective imaging method for diagnosis of medullary sponge kidney and ADPKD, which seen with thin prolonged calyces described as a spider-leg deformity; it determines the obstruction level and renal excretion [11,17,27].

CT scan is very sensitive imaging study for the diagnosis and characterizing of simple renal cysts and complex cystic mass with solid components or thick septae with enhancement suspected of renal cell carcinoma, detection of complications and associated abnormalities consistent with reported by Weibi et al. in 2017 [28].

Non-contrasted CT urography and US images detects non-radiopaque stones and used in renal failure [29].

MSK was diagnosed by contrasted urography; it shows a paint brush like appearance produced

by medullary collecting ducts. Koraishi et al. in 2014 [30] considered CT urography as the best diagnostic study for MSK.

Sporadic angiomolipoma is an extremely rare clinical entity; this study presented one patient with bilateral multiple simple cysts [11,31].

Medical treatment was the most frequent type of treatment and surgical intervention reserved for complicated patients and for renal cystic tumors [32]. The association of renal cysts with malignant renal cystic tumors is infrequent finding [33]. This study diagnosed four patients with malignant cystic lesions in adult renal cystic diseases, one of them seen in the left kidney (Figure 3) and other one in the right kidney, presented with thick calcification (Figure 4) and another one with cystic hypoechoic structure in horseshoe kidneys (Figure 5) and the later seen bilaterally in an adult dominant polycystic kidney disease (Figure 6).

This study concluded that:

1. Ultrasound is inconclusive for cystic renal masses.
2. CT scan is the most effective study for localized and multifocal cystic renal diseases, complications, and associated pathology.

Figure 3. CT scan demonstrates malignant cystic mass in the left kidney

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Figure 4. CT scan reveals right kidney enlargement and hypoechoic cystic tumor with hyperdense calcification

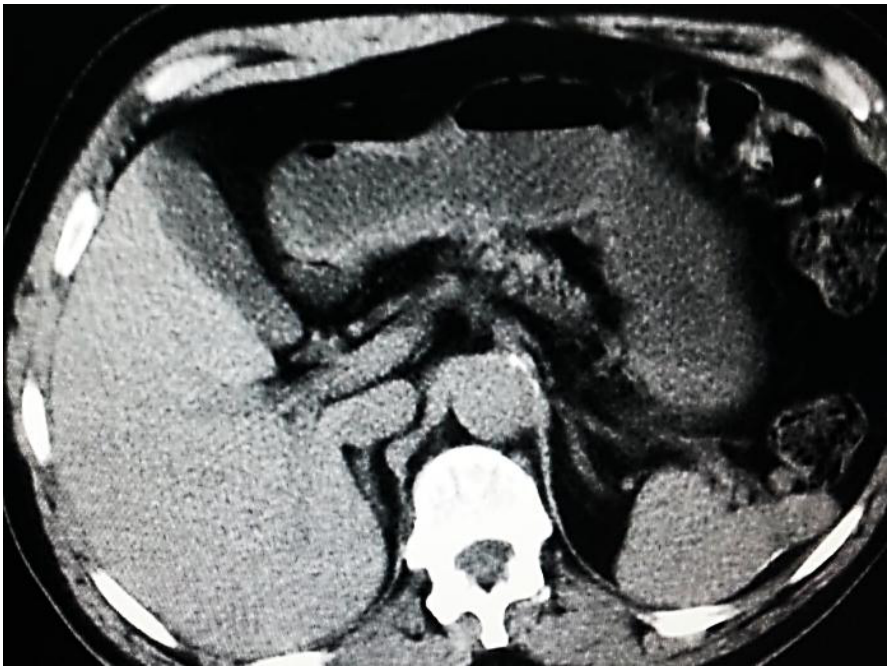

Figure 5. CT shows a hypoechoic cystic lesion in horseshoe kidney

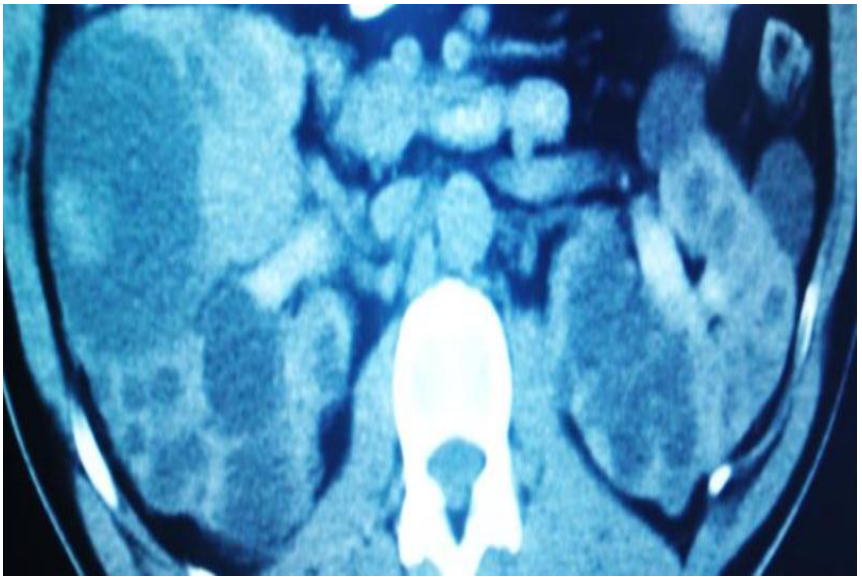

Figure 6. Contrasted CT scan reveals bilateral malignant cystic lesions in ADPKD

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Author contribution

Dr. Akares did the case selection and prepared the manuscript; Dr. Bahomil reviewed the radiological tests and prepared the manuscript.

Conflict of interest

The authors declare no conflicts of interest regarding the publication of this paper.

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