Atypical Renal Cyst (Clear Cell Type)

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Case Report

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Abstract

Background: Atypical cysts do not diagnostic criteria form classification as any of the variants of renal tumors, which have led to diagnostic difficulties. When these cysts have been encountered in surgical pathology practice it can be easily misdiagnosed as renal carcinoma and results in overtreatment.

Case presentation: A 55-year-old Chinese woman with a right renal cystic mass came to our hospital. The computed tomography revealed bilateral cysts in both kidneys. In histology, the right renal cystic mass was arranged by stratified epithelium with short papillary projections and tufting. The covered epithelial cells were characterized by clear cytoplasm and small nuclei. Immunohistochemistry, these epithelial cells were diffusely positive for CK7, CK8, CK18, Vimentin and EMA, weakly positive for RCC and PAX-8, while diffusely negative for AMACR, CD10 and TFE-3. And the Ki-67 positive rate was 3%. Based on the immunohistochemical profile and histological features, the final diagnosis is an atypical renal cyst (clear cell type). The patient underwent right nephrectomy. The patient is alive and without any sign of subsequent renal cancer during 11 months of follow-up.

Conclusion: Atypical renal cyst is extremely rare among the routine renal pathological diagnosis, we report this case to enhance the understanding and differential diagnosis of the atypical renal cyst and remind the pathologist of this precancerous lesion, and therefore, to avoid unnecessary or excessive treatment.

Background

Atypical renal cyst is extremely rare, and one central review and several scattered cases have described this disease in English-language literature, to our knowledge^[1–9]. These cystic lesions haven been called as renal cysts with papillary hyperplasia or renal cysts with atypical epithelial proliferation. However, there is a lack of standardized terminology of these cystic lesions. In the largest publication on atypical renal cysts, Matoso *et al.*^[1] divide them into three groups: ①clear cell, ②eosinophilic stratified; ③eosinophilic papillary. Atypical renal cysts usually occur in patients with complex syndromes (autosomal dominant polycystic kidney disease (APDK), von-Hippel-Lindau disease (VHLD), tuberous sclerosis complex (TSC)) or patients in chronic hemodialysis^[1, 10–12]. Rarely, atypical renal cysts occur in patients who do not suffer from end-stage renal disease. Still, they are associated with another solid renal cancer or represent a similar finding on imaging studies for other reason^[1]. There are no noticeable gender differences, and the age of onset varies from 17 to 80 years old. Multifocal is slightly more than unifocal, and the size of cyst varies from 0.8 cm to 10 cm. Per the definition, the clear cell group of atypical cysts are lined by stratified epithelium which is composed by cuboidal cells and characterized by clear cytoplasm and small nuclei (International Society of Urological Pathology (ISPU) nuclear grade 1 to 2). Occasionally, the cysts have an epithelial lining which was “piled up” or stratified and/or with short papillary projections or tufting, but lacking nests/cords of epithelial cells in the stroma or septa and without a solid mural nodule. By immunohistochemistry, the cuboidal lining cells are positive for CK7 and CAIX, and variably express AMACR and CD10. Cytogenetic abnormalities have been found till now include trisomy 7 and 17. These Cytogenetic features are similar to those of clear papillary renal cell carcinoma (CPRCC), which suggest that clear cell type of atypical cysts may be the precursor of CPRCC^[1]. In contrast to clear cell type, stratified eosinophilic and eosinophilic papillary type of atypical renal cysts are lined by cuboidal cells with scant eosinophilic cytoplasm. The immunophenotype and cytogenetic abnormalities are similar to that of the clear cell type. Besides, there is one case which found 3p deletion in the eosinophilic papillary type case. Although the clinical follow-up is limited in many cases, the great majority has no recurrence or metastasis, which suggests a relative inert prognosis. In the current case, we report a case of the atypical renal cyst (clear cell type).

Case Presentation

Clinical history

A 55-year-old Chinese woman was admitted to our hospital because a lump can be touched on the right waist along with pain. Physical examination found a round mass in the right area. The mass was soft and could not be pushed. No hematuria, no urinary frequency and urgency were found. The patient's past medical history is normal. Computed tomography (CT) scanning showed a cystic mass and exhibited with low-density shadow (8 x 9.2) in the inferior pole of the right kidney. The left kidney was normal in size, and two low-density shadows (1.1 x 0.7 and 0.7 x 0.5) were visible in the left renal parenchyma, which protruded from the outline of the kidney. Neither bilateral renal pelvis compression, kidney stone, nor enlargement of abdominal or retroperitoneal lymph nodes was found in CT scanning. Besides, there were no signs of ascites and no significant abnormalities in the bilateral adrenal glands (Fig. 1). The patient underwent right nephrectomy. The patients recovered well after the operation. CT recheck found no apparent abnormalities.

Methods

Immunohistochemical staining

Complete sampling of the double right renal cyst was performed according to conventional guidelines. All specimens were fixed in 10% formalin and embedded in paraffin. A series of thick sections were cut from each paraffin-embedded block. Immunohistochemistry studies were performed using the streptavidin-peroxidase system (Ultrasensitive; Mai Xin Inc., Fuzhou, China) according to the manufacturer's instructions. The primary antibodies used were the rabbit monoclonal anti- α -methylacyl-CoA racemase (AMACR), mouse monoclonal anti-cytokeratin (CK), mouse monoclonal anti-cytokeratin (CK)7, mouse monoclonal anti-CK8, mouse monoclonal anti-CK18, and mouse monoclonal anti-CD10 and directed against epithelial membrane antigen (EMA), Ki-67, Paired box 8 (PAX8), Vimentin, RCC and TFE-3. All antibodies were purchased from Mai Xin Inc., Fuzhou, China. The primary antibody was replaced with phosphate-buffered saline for negative controls.

Result

Morphological findings

All the right renal cystic mass was sampled to exclude the renal cancer accompanied with cystic change. No mural nodule was found under general or microscopy examination (Fig. 2A). The cyst is lined by cuboidal cells with clear cytoplasm and small nuclei (Fig. 2B-C). The lining epithelium was "piled up" or stratified and/or with short papillary projections or tufting, but lacking nests/cords of epithelial cells in the stroma or septa and without a solid mural nodule (Fig. 2D-E). The lining epithelial cells showed minimal atypia with small nuclei (ISUP grade 1) (Fig. 2F).

Immunohistochemical findings

The lining cells were positive for CK (Fig. 3A), CK7 (Fig. 3B), EMA (Fig. 3C), CK8 (Fig. 3D), CK18 (Fig. 3E), Vimentin (Fig. 3F), PAX-8 (Fig. 3J), but negative for CD10 (Fig. 3G), AMACR (Fig. 3H), RCC (Fig. 3I), TFE-3 (Fig. 3K), ki-67(3%)(Fig. 3L).

Final diagnosis is atypical renal cyst based on above findings.

Discussion

The renal cystic lesion is frequently encountered in clinical practice; it may include benign cysts, precancerous lesions, or malignant lesions^[13]. Among these lesions, the most common is simple renal cortical cysts, followed by renal cell carcinoma with the cystic change, and atypical renal cyst, which is much less encountered. By definition, atypical renal

cysts are those cystic masses of the kidney that bear an epithelial lining “piled up” or stratified and/or have short papillary projections or tufting. The epithelial cells are cytologically bland with small nuclei and a variable amount of clear or eosinophilic cytoplasm. The stroma or septa should be free of nests/cords of epithelial cells and often coexist with other diseases^[1]. A review of the English literature at PubMed (www.ncbi.nlm.nih.gov) shows that 14 papers with a total of 42 cases had relevant information and clinical records. Due to the rarity of this renal cyst, we discuss the case and make a literature review of previous cases (all cases were summarized in Table 1) to enhance the understanding and differential diagnosis of the atypical renal cyst to alert pathologists to the precancerous lesion and to avoid unnecessary or excessive treatment. The age of the patients was between 17 and 80 years old. There was no significant gender difference. Atypical renal cysts often occur in unilateral kidneys, and multilocular cysts are slightly more than unilocular cysts. The cysts vary in size with a diameter from 0.8cm to 10cm. The overall survival of atypical renal cysts is good, if there are no combined other primary diseases. The patient recovered well after surgery, without any sign of malignant progression. Among the reviewed cases, only one patient died of postoperative tumour recurrence. One patient died of metastatic lung cancer. Two patients died of sepsis, one of who was associated with respiratory failure and multiple metabolic disorders. One patient died of unknown causes. Atypical renal cysts usually coexist with other diseases, such as renal cell carcinoma (RCC) and VHLD. 34 of the 42 cases has accompanied other conditions, mainly of which are renal cell carcinoma (13/34), and seven cases of which are accompanied by VHLD (7/34), and others are ADPKD, angiomyolipoma (AML), tuberous sclerosis (TS) and so on.

Renal cystic lesions are relatively common, and the CT manifestations of the atypical renal cyst are complex and lack of signs of typical renal cyst, which may easily lead to misdiagnosis. Therefore, it is necessary to differentiate from other renal cystic diseases. The ultrasonography of simple renal cysts was smooth and without separation. They are common and require no clinical follow-up or treatment because they do not pose any risk for malignant transformation^[14]. Unless the cyst is large, it compresses other kidney tissue or surrounding organs. At this time, surgical treatment needs to be considered. We must admit the coexistence of renal cell carcinoma and atypical renal cysts do exist in clinical practice^[15, 16]. Ibrahim *et al.*^[6] found that atypical renal cysts exist in renal parenchyma close to the renal carcinomas. The characteristics of atypical cells lining the cysts are similar to those found in the close well-differentiated renal cell carcinomas^[17]. DNA quantitative studies show that both the renal cell carcinomas and the atypical cyst lining cells have the same DNA indices and are essentially DNA euploid^[6]. And it was reported that atypical renal cysts associated with end-stage renal disease had been considered to be precursors of malignant renal tumours^[9]. These descriptions suggest that atypical renal cysts may be associated with renal carcinoma. CT and ultrasound have a high diagnostic value for cystic renal carcinoma^[19, 20]. The Bosniak classification system, created in 1986^[21], established morphological CT criteria to differentiate between benign cysts and malignant cysts. Studies have shown that Bosniak criteria combined with ultrasound can greatly improve the diagnostic of benign and malignant renal cystic lesions^[22]. If the shape of the renal cyst found by ultrasound examination does not have the characteristics of a simple renal cyst and the boundary with the surrounding renal tissue is unclear, the inner wall is not smooth, or there are nodules and separation, which most likely cystic kidney cancer or a complex renal cyst. At this time, CT or enhanced CT examination is needed to eliminate the possibility of renal malignancy further. But exact diagnosis is difficult at times because the imaging features between CRCC and benign renal cystic diseases are similar^[23]. Therefore, if necessary, an ultrasound-guided biopsy should be performed. There are multiple cysts of different sizes in the cancer tissue, the cyst wall is lined with transparent cancer cells, and the transparent cancer cells in the capsule membrane are the main pathological features of cystic renal cell carcinoma. Multilocular cystic renal cell carcinoma (MCRCC), also known as multilocular clear cell renal cell carcinoma or multicystic clear cell carcinoma, is a rare type of renal cell carcinoma (RCC) with a relatively better outcome. And the diagnostic criteria for MCRCC are relatively strict, limited to RCCs with a malignant clear cell lining and small collections of cells without the formation of a malignant and expansive nodule. Histologic examination of MCRCC will reveal cysts lined by occasionally flattened cuboidal clear cells and septa containing aggregates of

epithelial cells with clear cytoplasm^[24]. But in the case, the morphological finding shows that the lining epithelium is “piled up” or stratified and/or with short papillary projections or tufting, and computed tomography (CT) scanning shows that there is lacking nests/cords of epithelial cells in the stroma or septa and without a solid mural nodule. Therefore, we excluded renal cancer accompanied by cystic change and diagnosed it as an atypical renal cyst. Also, to reduce the misdiagnosis of cystic renal carcinoma and improve the understanding of cystic renal carcinoma and atypical renal cyst, we should comprehensively analyze the patient's medical history, symptoms, signs and imaging examinations to make the diagnosis correct and provide reliable clinical support. For patients with atypical renal cysts, the possibility of cystic renal cancer should be considered before surgeries to avoid misdiagnosis.

Autosomal dominant polycystic kidney disease (ADPKD) is the most prevalent inherited nephropathy, which due to mutations in the PKD1 or PKD2 gene^[25, 26]. Autosomal dominant polycystic kidney disease (ADPKD) is characterized by progressive enlargement of kidney cysts leading to chronic kidney disease (CKD) and end-stage renal disease (ESRD)^[27–29]. Characteristic pathological changes and imaging tests are the primary diagnostic basis of ADPKD. Commonly used imaging examinations mainly include ultrasound and magnetic resonance imaging (MRI)^[30]. In this case, the patient had no typical pathological features and family genetic history. Therefore, the genetic polycystic kidney is not considered first.

Here, we reported a rare case of atypical renal cyst (clear cell type). Atypical renal cyst is extremely rare among the routine renal pathological biopsies. Based on its histopathologic features, it might be over diagnosed as renal cell carcinoma with cystic change and caused unnecessary treatment and excessive psychological burden.

Abbreviations

ADPKD: autosomal dominant polycystic kidney disease; VHLD: von Hippel-Lindau disease; TSC: tuberous sclerosis complex; ISUP: International Society of Urological Pathology; CPRCC: clear papillary renal cell carcinoma; CT: computed tomography; CK: cytokeratin; EMA: epithelial membrane antigen; PAX8: Paired box 8; AMACR: α-methylacyl-CoA racemase; RCC: renal cell carcinoma; TS: tuberous sclerosis; CRCC: cystic renal cell carcinoma; CAIX: Carbonic Anhydrase IX; CKD: chronic kidney disease; ESRD: end-stage renal disease; MCRCC: Multilocular cystic renal cell carcinoma

Declarations

Ethics approval and consent to participate

This study followed the principles of the Declaration of Helsinki.

Consent for publication

Written consent for publication was obtained from the patient discussed in this report.

Availability of data and materials

Not applicable.

Competing interests

The authors declare that they have no competing interest.

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Author contributions

Funding acquisition: YL, DZ.

Methodology: MS

Writing – original draft: MS, YL, NY.

Writing – review & editing: YL, DZ.

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Table

Table 1

Clinical data of reported atypical renal cyst.

Case	A/G	Size (cm)/Side	Multi/ Unifocal	Associated Lesions And Comorbidities	Follow up (mo)	CK7	AMACR	CD10	CAIX
1	72/F	7.0/R	M	Lithiasis	19/NED	+	+	-	+
2	64/F	1.5/R	M	PRCC and CRCC	7/ NED	+	-	-	+
3	70/F	2.5/R	M	DBT,on hemodialysis	3/ NED	+	-	-	+
4	34/F	NA/L	M	Lithiasis	110/ NED	+	-	-	+
5	56/M	2.5/L	M	None	120/ NED	+	+	-	-
6	60/M	2.8//L	M	None	NA	-	+	+	-
7	74/M	3.3/L	M	Hypertension	84/ NED	+	+	-	+
8	80/F	0.8/L	U	Oncocytoma	NA	+	-	-	+
9	37/F	1/L	M	PRCC	36/ NED	+	-	-	+
10	36/F	1/R	U	CRCC,Marfan Sd	20/Died	+	+	+	-
11	64/M	10/R	U	CRCC withsarcomatoid transformation	20/AWD	-	+	-	-
12	29/M	2/R	U	Organ donor	24/ NED	+	-	-	+
13	67/F	2.4/L	U	None	10/ NED	+	-	-	-
14	57/M	2.5/R	M	Hypertension	108/ NED	+	+	-	-
15	77/M	NA/L	U	CRCC, Chronic renal insufficiency	NA/Died	-	-	-	-
16	54/M	6.5/R	M	ADPKD	22/ NED	+	-	-	-
17	73/F	3/R	M	Chronic renal insufficiency in hemodialysis	5/ NED	+	+	-	-
18	67/M	1.5/L	U	AML	6/ NED	+	-	-	-
19	64/F	3/L	U	CRCC, Chronic renal insufficiency in hemodialysis	5/ NED	+	-	-	+
20	47/M	2/R	U	ADPKD	3/ NED	+	-	-	-
21	58/M	1.5/L	U	PRCC	10/ NED	+	-	+	-

PRCC, papillary renal cell carcinoma; CRCC, cystic renal cell carcinoma; ADPKD, autosomal dominant polycystic kidney disease; AML indicates angiomyolipoma;ARCD, acquired cystic renal disease; TS, tuberous sclerosis; VHLD, Von Hippel-Lindau disease; CCRCC, clear cell renal cell carcinoma; ESRD,end stage renal disease;RCC, renal cell carcinoma; MPNST, malignant peripheral nerve sheath tumor; AWD, alive with disease; DBT, diabetes mellitus; NA, not available; NED, no evidence of disease; ND, not described.

Case	A/G	Size (cm)/Side	Multi/ Unifocal	Associated Lesions And Comorbidities	Follow up (mo)	CK7	AMACR	CD10	CAIX
22	32/M	2/Bilat	M	ADPKD	3/ NED	+	-	-	-
23	47/F	1.5/L	U	None	NA	+	-	-	-
24	48/M	3.1/R	M	None	12/ NED	+	-	+	+
25	56/M	2.5/L	M	None	120/ NED	+	+	-	-
26	67/M	2.8/L	M	Lung cancer	NA/Died	+	-	-	-
27	69/M	3.2/R	U	None	NA	-	+	-	-
28	73/F	6.3/L	M	None	NA	-	+	+	+
29	51/M	2.8/R	M	CRCC	96/ NED	+	+	-	-
30	21/F	1/ Bilat	ND	ARCD	ND	ND	ND	ND	ND
31	17/F	ND/ Bilat	M	TSC	NED	ND	ND	ND	ND
32	26/F	1/ Bilat	M	VHLD CCRCC	NED	ND	ND	ND	ND
33	28/M	0.8-2/ Bilat	M	VHLD CCRCC	NED	ND	ND	ND	ND
34	31/M	ND/L	M	VHLD	ND/Died	ND	ND	ND	ND
35	30/M	ND	ND	ESRD	NED	ND	ND	ND	ND
36	42/F	ND	ND	VHLD RCC	NED	ND	ND	ND	ND
37	36/M	ND	ND	VHLD	NED	ND	ND	ND	ND
38	34/F	ND	ND	VHLD RCC	NED	ND	ND	ND	ND
39	76/F	ND	ND	ND	NED	ND	ND	ND	ND
40	62/F	ND	M	AML	NED	ND	ND	ND	ND
41	70/F	8.0/L	M	MPNST	10/ MPNST	ND	ND	ND	ND
42	43/M	ND/R	ND	VHLD,RCC,endocrine pancreatic head cancer	3/NED	ND	ND	ND	ND
42	56/M	6.0/L	ND	multicystic nephroma	NED	ND	ND	ND	ND
43(current case)	55/F	6.0/R	M	multiple renal cyst	11END	+	-	-	-
PRCC, papillary renal cell carcinoma; CRCC, cystic renal cell carcinoma; ADPKD, autosomal dominant polycystic kidney disease; AML indicates angiomyolipoma;ARCD, acquired cystic renal disease; TS, tuberous sclerosis; VHLD, Von Hippel-Lindau disease; CCRCC, clear cell renal cell carcinoma; ESRD,end stage renal disease;RCC, renal cell carcinoma; MPNST, malignant peripheral nerve sheath tumor; AWD, alive with disease; DBT, diabetes mellitus; NA, not available; NED, no evidence of disease; ND, not described.									

Figures

Figure 1

CT results. CT scanning showed multiple renal cyst, the biggest one was in the inferior pole of right kidney (8 cm x 9.2 cm). Two smaller cysts were in the left renal parenchyma. The arrows indicated the renal cysts.

Figure 2

Histologic features (A) No mural nodule was found under general or microscopy examination. (B-C) The cyst is lined by cuboidal cells with clear cytoplasm and small nuclei (B, magnification $\times 40$; C, magnification $\times 100$). (D-E) The lining epithelium was "piled up" or stratified and/or with short papillary projections or tufting, yet lacking nests/cords of epithelial cells in the stroma or septa and without a solid mural nodule (magnification $\times 200$). (F) The lining epithelial cells showed minimal atypia with small nuclei (ISUP grade 1)(magnification $\times 400$).

Figure 3

Immunohistochemical staining results (A-F,J) The lining cells were diffusely positive for CK (A), CK7 (B), EMA (C), CK8 (D), CK18 (E), Vimentin (F), PAX-8 (J); (G-I,K,L) The lining cells were negative for CD10 (G), AMACR (H), RCC (I), TFE-3 (K), Ki-67(3%) (L).

Supplementary Files

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