Case Report

Multilocular cystic renal cell carcinoma in a 23 year old female

Shivakumarswamy Udasimath, J. Niranjan[1], R. Puruhotham, K. R. Nagesha

Department of Pathology, Hassan Institute of Medical Sciences, Hassan, Karnataka, India, [1]Consultant Urologist, Karnataka, India

Abstract

Multilocular cystic renal cell carcinoma (MCRCC) has been identified as a separate subtype of renal cell carcinoma (RCC) in the 2004 World Health Organization classification of adult renal tumors. MCRCC represents a rare variant of clear cell RCC. The common age group for this tumor is between 40 and 60 years. In our case, MCRCC occurred at the age of 23 years in a female patient. We report this case because of its rarity in this age group, sex, good prognosis, and also to avoid a misdiagnosis as conventional clear cell RCC. However, before making a diagnosis of MCRCC, it has to be differentiated from tubulocystic carcinoma, cystic nephroma, cystic clear cell carcinoma, and clear cell papillary RCC.

Key Words: Clear cell renal cell carcinoma, kidney, multilocular cystic renal cell carcinoma

Address for correspondence:
Associate Prof. Shivakumarswamy Udasimath, Department of Pathology, Hassan Institute of Medical Sciences, Hassan - 573 201, Karnataka, India.
E-mail: udasimath@gmail.com

Received: 21.02.2016, **Accepted:** 05.05.2016

INTRODUCTION

Multilocular cystic renal cell carcinoma (MCRCC) represents a rare variant of clear cell renal cell carcinoma (RCC). MCRCC is also known as multilocular clear cell RCC and multicystic clear cell carcinoma.[1] MCRCC has been identified as a separate subtype of RCC in the 2004 World Health Organization (WHO) classification of adult renal tumors. The 2004 WHO classification of kidney tumors recognizes MCRCC as a rare variant of clear cell carcinoma with a good prognosis.[2] These tumors comprise approximately 1–2% of all renal tumors.[3] It is important to differentiate this entity from the clear cell RCC since it is associated with very good prognosis. There are other entities from which MCRCC has to be differentiated such as tubulocystic carcinoma cystic nephroma, cystic clear cell carcinoma, and clear cell papillary RCC. We report this MCRCC a rare entity in a 23-year-old female patient.

Access this article online	
Quick Response Code:	Website: www.urologyannals.com
	DOI: 10.4103/0974-7796.192095

CASE REPORT

A 23-year-old female presented with dull pain in the left lumbar region and dysuria since 2 months. Ultrasonography revealed a cystic mass in the cortex of the upper part of the left kidney. Contrast-enhanced computed tomography showed a heterogeneously enhancing, well-defined, encapsulated multicystic lesion in the upper part of the left kidney: Left partial nephrectomy was performed. Grossly, the left partial nephrectomy along with cystic mass specimen measured

How to cite this article: Udasimath S, Niranjan J, Puruhotham R, Nagesha KR. Multilocular cystic renal cell carcinoma in a 23 year old female. Urol Ann 2016;8:506-8.

7.4 cm × 6.2 cm × 5.4 cm and included a small portion of the left kidney. The cut surface of the left kidney showed a well-circumscribed, well-encapsulated cystic growth measuring 6.5 cm × 5.2 cm × 4.8 cm situated in the cortex in the upper pole of the kidney. The cut surface of the growth was having multiloculated cystic spaces. Cystic spaces were of varying sizes with thin septa in between the cysts. The cysts were filled with pale yellow colored thin, gelatinous material [Figure 1]. Microscopic examination of the cystic growth revealed a complex cystic tumor comprising of variably sized noncommunicating cysts separated predominantly by thin septa [Figure 2]. Septa were lined by sheets and nests of tumor cells showing uniform, hyperchromatic nucleus, inconspicuous nucleoli (Fuhrman nuclear Grade 1), and abundant clear cytoplasm with distinct cell borders [Figure 3]. Mitotic figures were scarce. Lumen of the cysts contained eosinophilic secretions. Septa between the cysts contain cords and clusters of similar tumor cells. Adjacent renal parenchyma showed degenerative changes in the form of focal glomerular hyalinization and sclerosis, tubular casts, interstitial fibrosis, and chronic inflammatory cell infiltrate [Figure 4]. The histomorphology was compatible with MCRCC, Fuhrman nuclear Grade 1. Now, the patient is well without any recurrence.

DISCUSSION

RCC account for 80–85% of primary malignant neoplasms of the kidney. Commonly, RCC presents as a solid mass; however, in 10–22% of cases, RCC appears as a unilocular or multilocular cystic mass on imaging studies.[4] MCRCC is also known as multilocular clear cell RCC and multicystic clear cell carcinoma.[1] These tumors comprise approximately 1–2% of all renal tumors.[3] The 2004 WHO classification recognizes MCRCC as a rare variant of clear cell RCC with a good prognosis.[1]

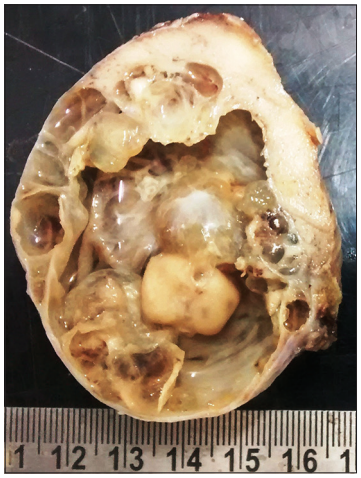

Figure 1: The left partial nephrectomy specimen showing an encapsulated growth with multiloculated, cystic growth. The cysts are filled with gelatinous material and are separated by thin septa

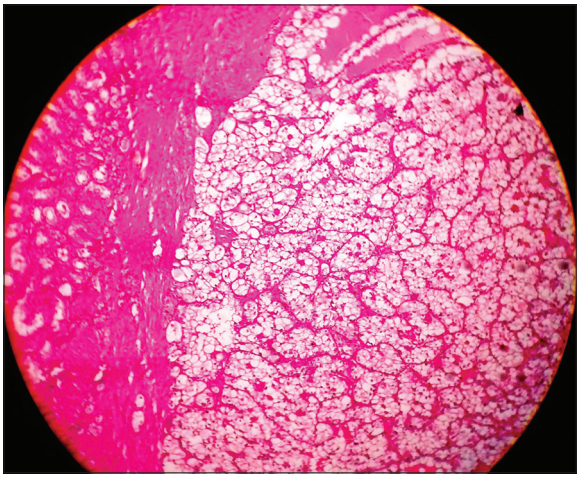

Figure 2: Cyst wall lined by clear cells with uniform hyperchromatic nuclei, inconspicuous nucleoli, clear cytoplasm, and well-defined cell borders (H and E, ×40)

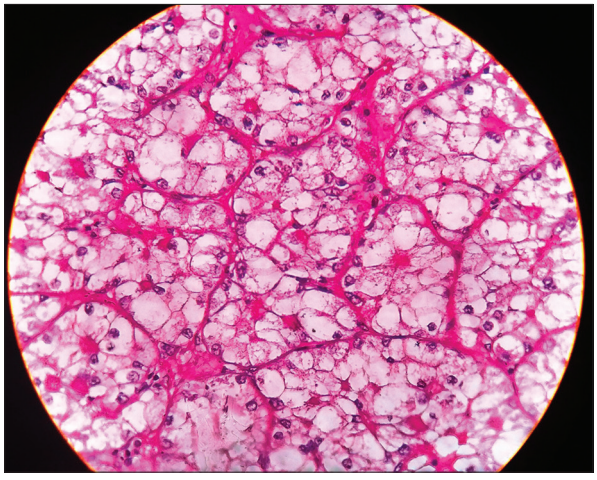

Figure 3: Sheets of tumor cells showing uniform, hyperchromatic nucleus, inconspicuous nucleoli (Fuhrman nuclear Grade 1), and abundant clear cytoplasm with distinct cell borders (H and E, ×40)

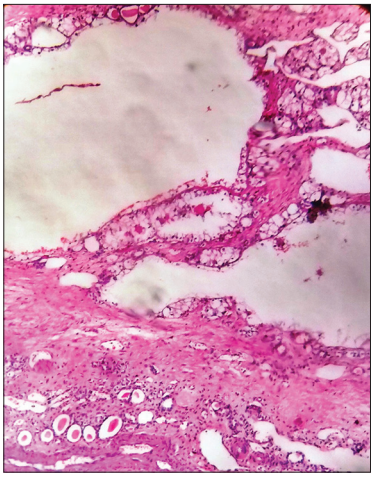

Figure 4: Adjacent renal parenchyma of multilocular cystic renal cell carcinoma showing degenerative changes, thyroidization of tubules, and interstitial fibrosis

While cysts are common in clear cell RCC, only rarely is the tumor entirely composed of the cyst. The term MCRCC should be used exclusively to identify cystic RCC with a small volume (25% or less) of neoplastic clear cells in the cyst walls.[1]

Described mechanisms to account for RCC with cystic features are (1) intrinsic unilocular cystic growth (papillary cystic adenocarcinoma), (2) intrinsic multilocular cystic growth, (3) tumor necrosis resulting in cyst formation (pseudocyst), and (4) tumor arising in a preexisting simple renal cyst.[4]

The male to female ratio for MCRCC is 3:1. The mean age of incidence is 51 years.[5] Diagnostic criteria for MCRCC were defined by the 2004 WHO classification of kidney tumors[6] Criteria were based on the previous reports and suggestions of Ebin and Bonsib.[7] These histological criteria for MCRCC are the presence of (a) circumscribed, noncommunicating, expansile nodule composed entirely of cysts and septa. (b) Cysts should be lined by single layer of low-grade clear cells. (c) No papillary growth should be identified. (d) The cysts should be separated by fibrous septa. (e) The septa may have groups of low-grade clear cells. These groups must not be expansile nodules and must not show infiltrative growth. (f) Nuclei should be of low Grade (1 or 2). Before making a diagnosis of MCRCC, the tumor should be extensively sampled.[5] The presence of any expansile nodules of clear cells in the septa between the cysts indicates an ordinary, fully malignant, clear cell carcinoma with cystic change.

The differential diagnosis of MCRCC consists of other cystic lesions of kidney, mainly tubulocystic carcinoma, primarily cystic nephroma, extensively cystic clear cell RCC, and clear cell papillary RCC.

Tubulocystic carcinoma differs from MCRCC in that the cystic spaces are lined by flat cuboidal and sometimes hobnail-type cells with eosinophilic cytoplasm and variable nuclear atypia, typically with nucleolar prominence in the range of Fuhrman Grade 2 or 3, a feature incompatible with MCRCC. In addition, the septal structures of tubulocystic carcinoma do not harbor clusters of clear cells.[6]

While cystic nephroma may have at least some clear cells lining the septa, the lining clear cells tend to be focally rather than diffusely distributed, and there are no clusters of clear cells in the walls. The ovarian-like stroma in cystic nephroma, if present, distinguishes it from multilocular cystic RCC by the fact that solid areas may be evident grossly, or expansile nodules of clear cells are observed microscopically. Clear cell papillary RCC is usually cystic and the cyst walls are lined by clear cells; however, much of the tumor exhibits papillary architecture, a feature not found in MCRCC.[7]

Halat *et al*. conducted a study in 2010 in which they performed fluorescence *in situ* hybridization analysis for 3p deletion on samples from MCRCC and compared the findings with those from a population of similarly low-grade conventional clear cell RCC. Deletion of 3p was observed in 74% of MCRCC and in 89% of clear cell RCC with no statistically significant difference in the incidence of 3p deletion between the groups.[8]

Hence, proving that MCRCC is a subtype of clear cell RCC. To date, this tumor has never been shown to metastasize.[9] However, Walsh *et al*. in 2010 have reported the first case of MCRCC metastasizing to one out of seven intra-aortocaval lymph nodes.[10]

We report this case because of its rarity in this age group and sex with good prognosis and also to avoid a misdiagnosis as conventional clear cell RCC.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Nassir A, Jollimore J, Gupta R, Bell D, Norman R. Multilocular cystic renal cell carcinoma: A series of 12 cases and review of the literature. Urology 2002;60:421-7.
2. Eble JN. Multilocular cystic renal cell carcinoma. Pathology and genetics of tumors of urinary system and male genital organs. In: Eble JN, Sauter G, Epstein JI, Sesterhenn IA, editors. World Health Organization Classification of Tumors. Lyon, France: IARC Press; 2004.
3. Han KR, Janzen NK, McWhorter VC, Kim HL, Pantuck AJ, Zisman A, *et al*. Cystic renal cell carcinoma: Biology and clinical behavior. Urol Oncol 2004;22:410-4.
4. Murphy WM, Grignon DJ, Perlman EJ. Tumors of the Kidney, Bladder, and Related Urinary Structures. AFIP Atlas of Tumor Pathology. Fourth Series, Fascicle 1. Washington, DC: American Registry of Pathology; 2004. p. 121-3.
5. Eble JN, Bonsib SM. Extensively cystic renal neoplasms: Cystic nephroma, cystic partially differentiated nephroblastoma, multilocular cystic renal cell carcinoma, and cystic hamartoma of renal pelvis. Semin Diagn Pathol 1998;15:2-20.
6. Amin MB, MacLennan GT, Gupta R, Grignon D, Paraf F, Vieillefond A, *et al*. Tubulocystic carcinoma of the kidney: Clinicopathologic analysis of 31 cases of a distinctive rare subtype of renal cell carcinoma. Am J Surg Pathol 2009;33:384-92.
7. Gobbo S, Eble JN, Grignon DJ, Martignoni G, MacLennan GT, Shah RB, *et al*. Clear cell papillary renal cell carcinoma: A distinct histopathologic and molecular genetic entity. Am J Surg Pathol 2008;32:1239-45.
8. Halat S, Eble JN, Grignon DJ, Lopez-Beltran A, Montironi R, Tan PH, *et al*. Multilocular cystic renal cell carcinoma is a subtype of clear cell renal cell carcinoma. Mod Pathol 2010;23:931-6.
9. Suzigan S, López-Beltrán A, Montironi R, Drut R, Romero A, Hayashi T, *et al*. Multilocular cystic renal cell carcinoma: A report of 45 cases of a kidney tumor of low malignant potential. Am J Clin Pathol 2006;125:217-22.
10. Walsh TJ, Naik P, Sweeney P, Fitzgibbon J, Murphy M. Metastatic multilocular cystic renal cell carcinoma – Malignant cells clearly a treatment dilemma. J Lab Physicians 2014;6:50-2. doi: 10.4103/0974-2727.129093 .