|| ISSN(online): 2589-8698 || ISSN(print): 2589-868X ||
International Journal of Medical and Biomedical Studies
Available Online at www.ijmbs.info
NLM (National Library of Medicine ID: 101738825)
Index Copernicus Value 2020: 79.44
Volume 6, Issue 06; June: 2022; Page No. 52-57

Case report

Cystic Chromophobe Renal Cell Carcinoma Presenting as a Cystic Lesion in 27year Old Female

Tonushree Purohit[1], Archana Shrivastava[2], Reeni Malik[3]

[1]PG Resident, Department of Pathology, Gandhi Medical College and Hamidia Hospital, Bhopal

[2]Associate Professor, Department of Pathology, Gandhi Medical College and Hamidia Hospital, Bhopal

[3]Professor and Head, Department of Pathology, Gandhi Medical College and Hamidia Hospital, Bhopal

Article Info: Received 24 May 2022; Accepted 30 June 2022
doi: https://doi.org/10.32553/ijmbs.v6i6.2557
Corresponding author: Archana Shrivastava
Conflict of interest: No conflict of interest.

Abstract

Chromophobe renal cell carcinoma (ChRCC) is an uncommon variant of renal cell carcinoma which accounts for 4% to 6% of RCC mainly diagnosed in the advance decades of life.[1] This entity must be identified since it has a much better prognosis than clear cell (conventional) and papillary renal cell carcinomas. The chromophobe renal cell carcinoma should be differentiated from oncocytoma and clear cell carcinoma. We report a case of a 27year-old female who presented with history of right sided flank pain and a palpable tender lump in the right lumbar region since one year. The likelihood of a right renal neoplasm was highlighted on radiography. A right total nephrectomy was done and histopathological diagnosis of cystic chromophobe renal cell carcinoma was given. As the prognosis and therapy procedures vary with different variants, we discuss this case because of its relative rarity of occurrence, difficulties encountered, and differential diagnoses to consider during diagnosis. Early identification and treatment is necessary as there is increased risk of metastasis mainly to liver.[2] Numerous studies have also confirmed that tumor size and the presence of sarcomatoid morphology indicates poor prognosis and increased risk of metastatic development of chromophobe RCC.[2-3]

Key words: chromophobe, oncocytoma, nephrectomy

Introduction

Chromophobe renal cell carcinoma (chrRCC) is a rare subtype of RCC which accounts for 4% to 6% of RCC subtypes which are: clear cell RCC, papillary RCC, hereditary cancer syndromes, multilocular cystic RCC, collecting duct carcinoma, medullary carcinoma, mucinous tubular and spindle cell carcinoma, neuroblastoma-associated RCC, Xp11.2 translocation–*TFE3* carcinoma, and unclassified lesions.[1]

ChrRCC arises from the cortical collecting duct, type B intercalated cells and is characterized histologically by its large polygonal pale cells, prominent cell membrane, reticulated cytoplasm, and peri-nuclear halos [4]. It has a favorable prognosis and an estimated 90% 5-year survival rate if not diagnosed early with time can lead to sarcomatous degeneration and necrosis thereby increasing unfavourable outcome.[5] There is no sex predilection male to female ratio is 1:1.

CD117 and CK-7 positivity is demonstrated by tumor cells in ChrRCC [6]. In our case report, we describe a rare presentation of a chrRCC in a 27year old female presented with right flank pain. [1].

Case report

A 27year female came with complaints of pain in right flank since one year with no pre-existing medical condition, no family history no prior surgical history, no history of addiction or trauma. On physical examination, scaphoid abdomen was observed with a lump palpable over right lumbar region with tenderness when lump hits the anterior wall. The initial laboratory assessment was done.

Investigations:

Table 1:

PARAMETER	VALUE
Hemoglobin	10.0g/dl
Hematocrit	30.3 %
Total leucocyte count	17000 cells/cumm
Neutrophils	76.7%
Lymphocytes	20.5%
Monocytes	2.1%
Eosinophils	2.2%
Basophils	0.5%
Platelet count	1.20 lakhs/cumm
Blood urea	50mg/dl
Serum creatinine	1.8 mg/dl

The patient underwent initial assessment by ultrasonographic scanning. It revealed an ill-defined hyperechoic mass of size measuring approximately 52x49mm involving mid-pole of right kidney showing central anechoic areas within the mass and vascularity on color flow. CT abdomen suggested a well-defined expansile iso-attenuating mass measuring 58x53x61mm lesion arising from interpole cortex of right kidney showing thick peripheral post contrast enhancement which is hypo-enhancing as compared to normal renal cortex showing central necrotic area within suggesting neoplastic etiology .The patient underwent a total right nephrectomy with an uneventful post-operative period.

Gross pathological examination of the nephrectomy specimen revealed kidney weighing 400 grams with its adhered fascia measures 8x5.5x3.5cm along with ureter and fibrofatty tissue. Outer surface was greyish brown, smooth with greyish white bosselated area on the anterolateral surface of the specimen measuring 4x4cm. On cut section through renal parenchyma, cortico-medullary differentiation could be identified at upper and lower pole. There was a well circumscribed greyish white growth occupying the middle pole extending upto the pelvis (pelvis could not be identified) measuring 3.5x3x3 cm was noted which showed greyish white friable areas with few greyish black areas. There was a unilocular cystic space present in the growth measuring approximately 2.5cm in

maximum diameter filled with serous material with the wall thickness measuring approximately 1mm.Attached ureter measured 4cm in length and 0.5cm in maximum diameter. Ureter showed invasion by tumor cells.

Histologically:

Multiple sections from nephrectomy specimen examined revealed tumor cells arranged in alveolar and papillary pattern separated from each other by fibrous septa. Cells were showing eosinophilic granular cytoplasm with centrally placed nuclei showing perinuclear halo. Multiple microcytic spaces filled with proteinaceous material was also observed Adjacent normal renal parenchyma showed evidence of tubular necrosis, tubulorhexxis. Few tumor cells were showing clear cytoplasm. Ureter showed invasion by tumor cells. These findings were consistent with the diagnosis of cystic chromophobe renal cell carcinoma.

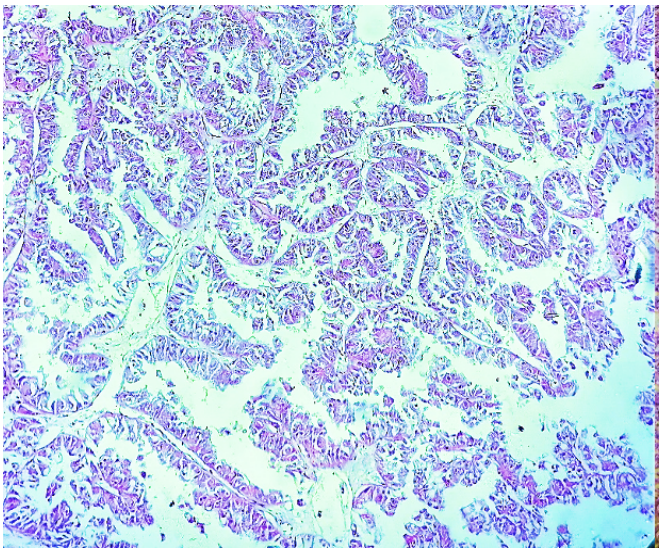

Fig.1:

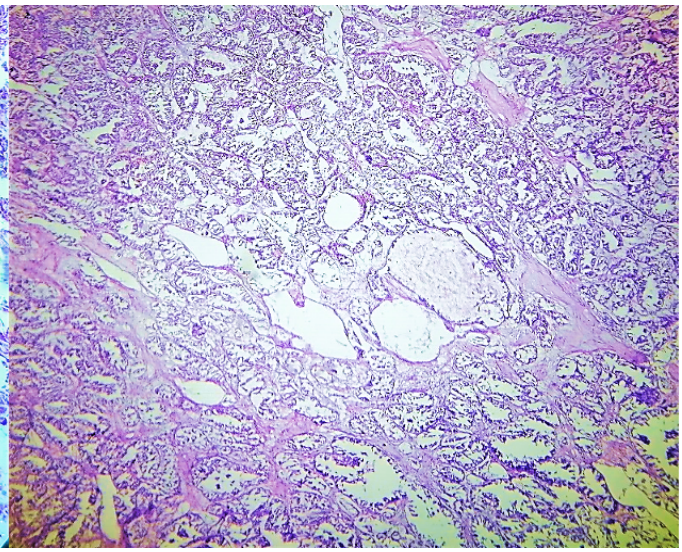

Fig.2:

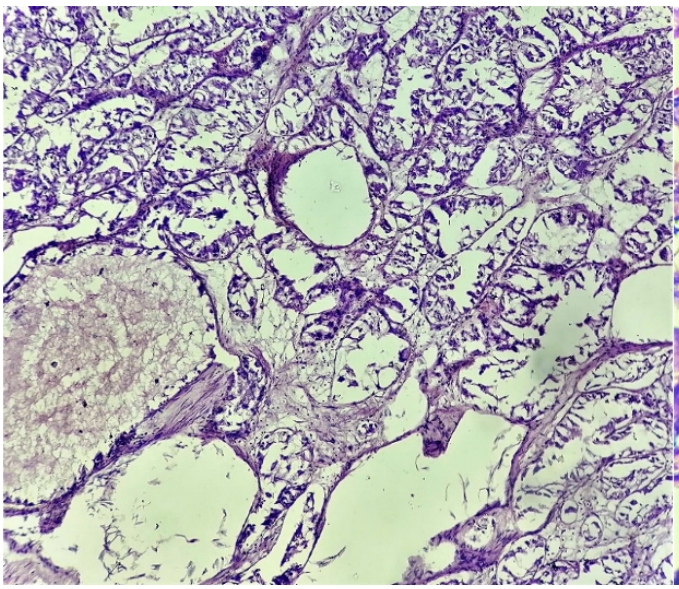

Fig.4:

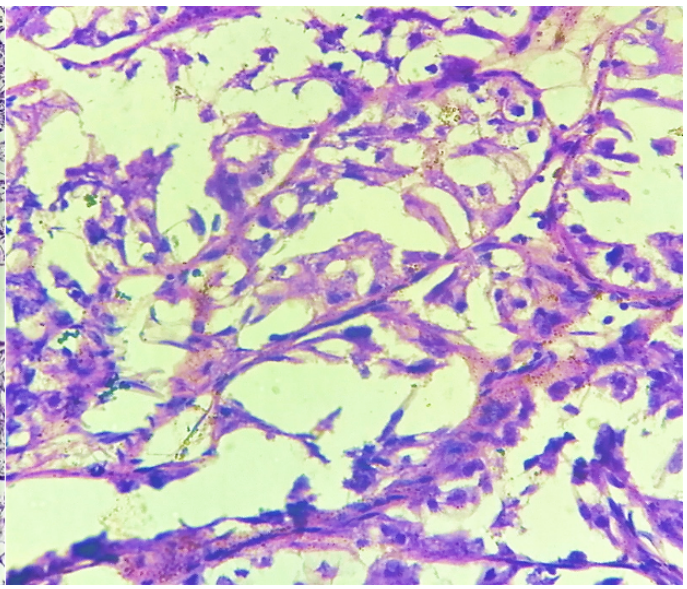

Fig.5:

Fig.5:

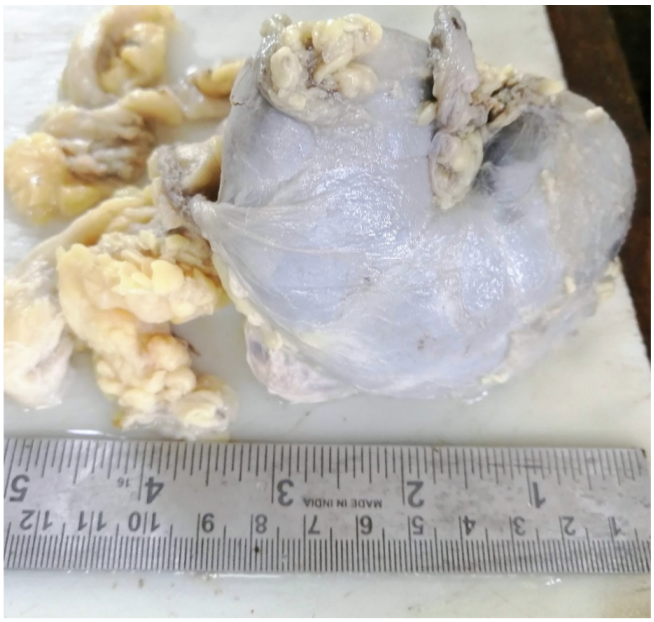

Fig.6:

Discussion:

Currently, under the classification World Health Organization (WHO) **[7]**, renal cell carcinomas are recognized to have several variants- clear cell type (70%), papillary type (10-15%), chromophobe type (4-6%), collecting duct type (about 1%) and unclassified RCC (4-5%). RCCs are more often incidentally diagnosed and usually remain asymptomatic. Chromophobe renal cell carcinoma presents in advance stages of life mean age being in the sixth decade [8] years whereas in our case our patient was in her late twenties.

The main symptoms form a classical triad of flank pain, gross hematuria, and a palpable abdominal mass. Flank pain has been reported as the most common presenting symptom **[9]** for patients with ChRCC which was similar in our case as our patient presented with flank pain and discomfort in abdomen. Several risk factors were identified that are potentially responsible for the increasing number of newly-diagnosed renal cell carcinomas, among them were smoking, hypertension, age, and male gender **[10]** but in our case patient was female with no addiction history, no previous medical history of hypertension. There are reports of osseous metaplasia **[11]** whereas there was no evidence of calcification histologically as well as radiologically in our case. Macroscopically chromophobe renal cell carcinoma presents as a solitary, circumscribed uncapsulated mass. The morphological parameters of chromophobe renal carcinoma are :100% solid, mostly solid with a small cystic component, equally cystic and solid, mostly cystic with a small solid component, or 100% cystic**.[13]** In our case there was predominantly cystic component presentation.

Microscopically, Chromophobe renal cell carcinoma is divided into: Type I (eosinophilic) tiny sized call with eosinophilic and granular cytoplasm. Other varieties, such as Type II (mixed) cells, have a close resemblance to eosinophilic variants but are larger in size and have a translucent peri nuclear zone. **[13]**In our situation, microscopic evidence indicated to a type II variation. Type III (classical) cells have well-defined, thick borders, wrinkled or 'raisinoid' nuclei, and pale, granular cytoplasm.

The benign renal oncocytoma is a differential diagnosis for ChRCC. The radiological and

cytological findings are similar **[14].** As a result, a definitive diagnosis of oncocytoma is required, which can be achieved through histological investigation. Oncocytoma is the most similar eosinophilic form of ChRCC. Sheet-like organisation, raisinoid nuclei, and the presence of type I, type II, and type III are all histologic features unique to the chromophobe variation of RCC. Oncocytomas, on the other hand, have a nested and tubular configuration with rounded hyperchromatic nuclei and degenerative atypia, which were both histologically and radiologically lacking in our patient, indicating chromophobe renal cell carcinoma. Immunohistochemistry can also be done to differentiate between chromophobe renal cell carcinoma and oncocytoma where the former shows positivity for CK7 and latter is negative**.[6]**

Conclusion

Cystic RCC is a rare occurrence, and chromophobe RCC that presents as a primarily cystic mass is extremely rarer. These tumours have a good prognosis with a 90% 5-year survival rate. There is no gender preference. The male to female ratio is one to one**.[1]**

We present this case report as it has relatively low prevalence and unique nature, and have a substantially better prognosis than other RCC variants, identifying and diagnosing this variant is important and helpful. In comparison to other subtypes of RCCs, ChRCCs are more frequently diagnosed at an earlier stage and are thus associated with a better prognosis and survival rates as the carcinoma may metastasize Patients with high grade tumours (sarcomatoid differentiation and/or tumour necrosis) should have a stringent follow-up because of their increased risk of metastasis. Favourable responses to VGF-tyrosine kinase inhibitor agent, mTOR inhibitors and immune checkpoint inhibitors have been reported. Sarcomatoid differentiation in chromophobe RCC is an indicator of limited response to systemic therapy and poor overall survival**.[3]**

References:

1. Alghamdi MH, Alshabyli NA, Alayed A. Chromophobe Renal Cell Carcinoma Presenting as a Cystic Renal Mass: Case Report and Review of the Literature. Am J Case Rep 2019; 20: 631-634.
2. Casuscelli J, Becerra MF, Seier K, Manley BJ, Benfante N, Redzematovic A, Stief CG, Hsieh JJ, Tickoo SK, Reuter VE, Coleman JA. Chromophobe renal cell carcinoma: results from a large single-institution series. Clinical genitourinary cancer. 2019 Oct 1;17(5):373-9.
3. Ged Y, Chen YB, Knezevic A, Casuscelli J, Redzematovic A, DiNatale RG, Carlo MI, Lee CH, Feldman DR, Patil S, Hakimi AA. Metastatic chromophobe renal cell carcinoma: Presence or absence of sarcomatoid differentiation determines clinical course and treatment outcomes. Clinical genitourinary cancer. 2019 Jun 1;17(3):e678-88.
4. Wader JV, Kumbhar SS, Huddedar AD, Khatib WG. Chromophobe renal cell carcinoma. Online Journal of Health and Allied Sciences. 2013 Apr 15;12(1 (15)).
5. Daga D, Dana R, Kothari N. Chromophobe renal cell carcinoma with sarcomatoid changes: case report and review of literature. Central European Journal of Urology. 2014;67(1):31.
6. Garje R, Elhag D, Yasin HA, Acharya L, Vaena D, Dahmoush L. Comprehensive review of chromophobe renal cell carcinoma. Critical reviews in oncology/hematology. 2021 Apr 1;160:103287.
7. Chandra A. Chromophobe renal cell carcinoma in 76 years old female: A case report. IAIM 2017; 4(8): 139-142
8. Saguema I, Charfia S, Kallel R, Maknia S, Fourati M, Ayadi L, Mhiri MN, Boudawara T. Chromophobe renal cell carcinoma in an 18-year-old female. African Journal of Urology. 2016 Sep 29;22(3):178-82.

9. Bhalsod HD, Khan IA. Chromophobe variant of renal cell carcinoma–A rare case report.
10. Wu SL, Fishman IJ, Shannon RL. Chromophobe renal cell carcinoma with extensive calcification and ossification. Annals of Diagnostic Pathology. 2002 Aug 1;6(4):244-7.
11. Kefeli M, Yildiz L, Aydin O, Kandemir B, Yilmaz AF. Chromophobe renal cell carcinoma with osseous metaplasia containing fatty bone marrow element: a case report. Pathology-Research and Practice. 2007 Oct 18;203(10):749-52.
12. Raman SP, Johnson PT, Allaf ME et al: Chromophobe renal cell carcinoma: Multiphase MDCT enhancement patterns and morphologic features. Am J Roentgenol, 2013; 201: 1268–76
13. Katzman PJ, Schwartz JI. Chromophobe Renal Cell Carcinoma in a child: Case Report and Review of the Literature. Pediatr Dev Pathol 2007; 10(2):125-128
14. Vera-Badillo FE, Conde E, Duran I. Chromophobe renal cell carcinoma: a review of an uncommon entity. International Journal of Urology. 2012 Oct;19(10):894-900.