



A Rare Finding of Clear Cell Papillary Renal Cell Carcinoma in a Complex Renal Cyst

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Received 13 September 2019;

Accepted 29 September 2019;

Published 05 October 2019

Abstract

Multilocular cystic renal cell carcinomas (RCC's), Unilocular cystic RCC's and RCC's with extensive cystic necrosis and unilocular cysts with mural tumor nodules forms a wide category of RCC's. Cystic RCC's represent upto 3-14% of all RCC's.^[1] Here we present a case of 68 year old female who is a known case of DM, hypothyroidism, bronchial asthma presenting with complains of pain abdomen since 1 year. She has been a case of renal TB and on ATT 30 years ago. Further the patient was evaluated and found to have a small partly exophytic cortical cyst measuring 13x12mm noted in the inter polar region posteriorly along with multiple well defined hyper dense non enhancing cortical and exophytic cysts notes in upper / lower and inter polar regions, largest measuring 1.8x1.7cms in upper pole suggestive of hemorrhagic cysts. A diagnosis of left renal complex cyst was made. Alaparoscopic left partial cystectomy was done and sent for histopathological examination. Microscopy revealed Cystic clear cell fgv Renal cell carcinoma, Fuhman Grade 1(Low grade) within a 0.1cm thickening which was further confirmed with IHC expressing strongly for CK7,PAX8 and CAIX and confirmed the diagnosis as Clear cell Papillary Renal cell carcinoma. Patient is being followed up and no recurrence / metastasis has been reported. This provides us an opportunity to consider an unusual finding of Clear Cell Papillary RCC in a complex renal cyst and to look into the cystic lesion with more importance helping us in avoiding missing a carcinoma as found in the present case.

Keywords: *Complex cysts, Cystic papillary clear cell RCC, Thickened cyst wall, Fuhrman Grade 1.*

Introduction

Cystic clear cell papillary renal cell carcinoma is a very unusual low grade RCC in a multilocular cyst. It represents upto 3-14% of all the RCC's reported^[1]. The prognosis of multilocular cystic RCC's is found to be excellent with an unusual recurrence when compared with other RCC's presenting with both solid and cystic component within^[2]. Cystic renal cell carcinomas shares a similar radiological finding as well as clinical presentation and hence its misdiagnosed as benign renal cyst^[3]. This case report helps us in meticulous analysis of such complex multilocular renal cysts.

Case Details

We came across a 68 year old lady presenting with pain abdomen since 1 year. She was a known case of Diabetes mellitus, Hypothyroidism and Bronchial asthma. She was also a case of Renal tuberculosis who was on ATT 30 years ago. She had also undergone anal fissurectomy and cholecystectomy 12 years ago and thyroidectomy 10 years ago. The vitals were stable at the time of presentation. Further sonological findings showed a cystic lesion

in the middle pole of left kidney consisting of a single vein and double renal artery with early branching. Under radiological examination a small partly exophytic cortical cyst measuring 13x12mm was noted in interpolar region posteriorly. It also showed a small enhancing focus along posterior aspect. Also noted few multiple well defined hyperdense non enhancing cortical and exophytic cysts in the upper / lower and interpar regions largest measuring 1.8x1.7cms in the upper pole suggesting hemorrhagic cysts. Radiological report was finalised as a Left complex renal cyst. The patient was taken up for a left complex partial cyst excision to rule out malignancy. The excised cyst was sent for histopathological analysis. Gross examination showed a cyst measuring 2x1x0.5cms. Cut section of both inner and outer surface appeared smooth except for a focal area of 0.1cms exhibiting a thickened area. The cyst was filled with hemorrhagic fluid. Under microscopy the cyst revealed a cuboidal lining epithelium. The area within the cyst exhibiting with thickening showed cells arranged in tubular and acinar pattern with round to oval shaped cells exhibiting abundant clear cytoplasm. A distinct cytoplasmic membrane was noted. Few cells showed prominent nuclei. A peculiar papillary projection was noted at the edge of the lesion.

The cells within the papillary projections revealed eosinophilic cytoplasm. Hemosiderin laden macrophages was also seen. The rest of the area revealed a benign tissue where in glomeruli showed sclerosis at few areas and tubules with focal thyroidisation. Interstitium with mild chronic inflammatory cell infiltrate. Histopathological diagnosis of Cystic Clear cell Renal cell carcinoma with papillary areas, Fuhrman Grade 1(Low grade) was given(Figure 1) and further subjected the tissue for immunohistochemical analysis for confirmation. The neoplastic cells expressed a strong positivity for CK 7, PAX 8 and CA IX. Negative for AMACR and CD10 (Figure 2,3,4,5 and 5a).A final diagnosis of Clear Cell Papillary Renal cell Carcinoma was given. Further the patient was followed up and found to have no evidence of recurrence or metastasis.

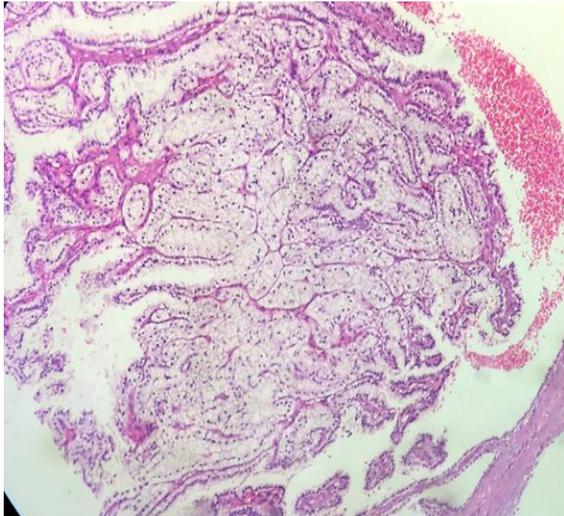


Figure1: Papillary clear cell features within cystic lesion. (H&E 40x)

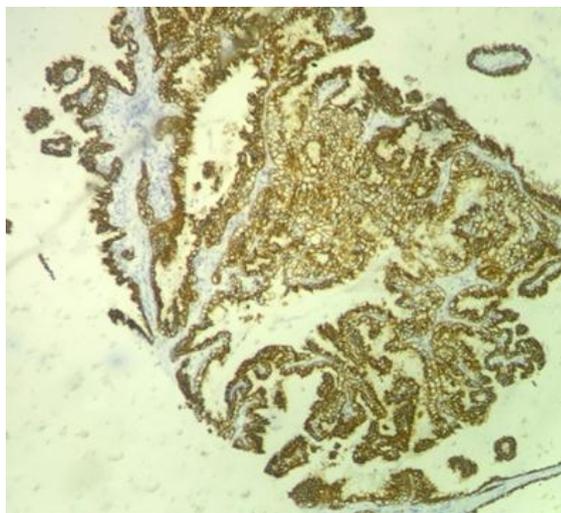


Figure 2:CK7 Positive

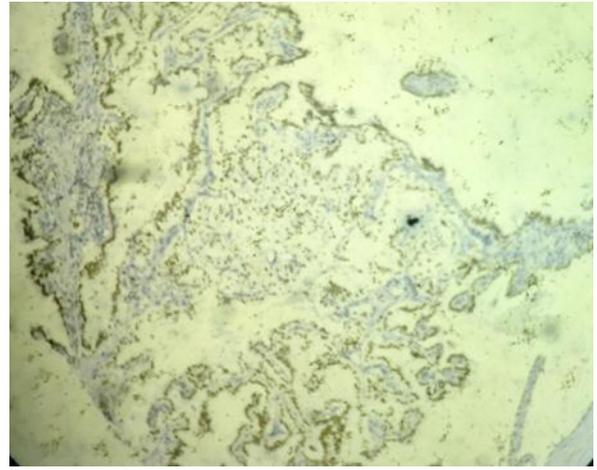


Figure 3:PAX8 Positive

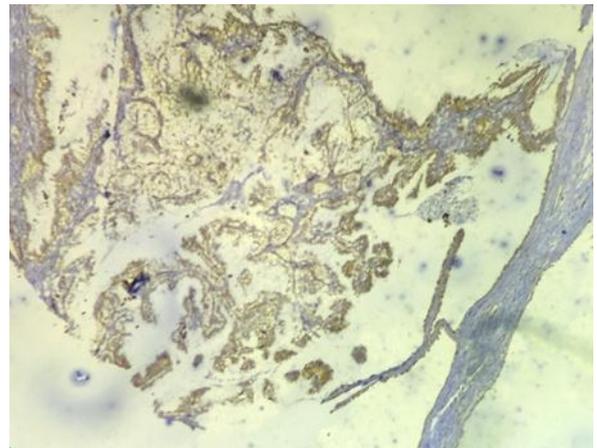


Figure 4: Vimentin positive

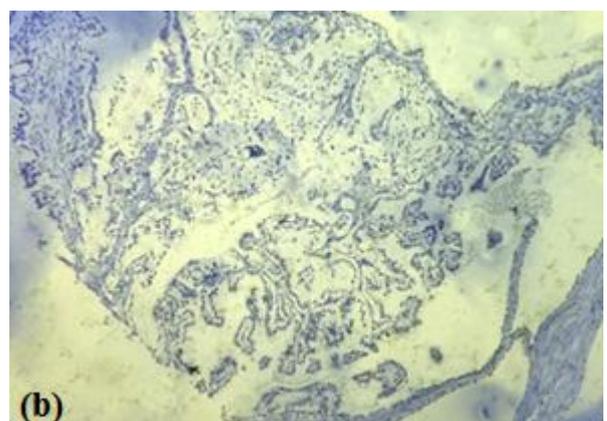
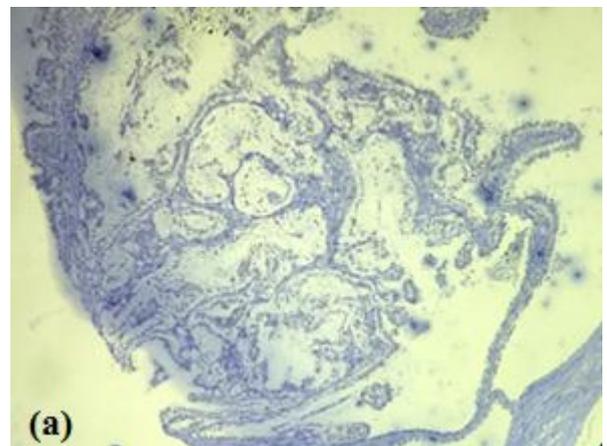


Figure 5: (a) & (b) CD10 & AMACR Negative

Discussion

Cystic change within the kidney is a very usual finding. The differential diagnosis for a Cystic renal cell carcinoma include RCC with cystic change, hereditary leiomyomatosis presenting with cystic lesions, cystic nephroma, clear cell papillary RCC and few others^[3]. The mechanisms considered for cystic RCC include intrinsic unilocular cystic growth, intrinsic multilocular cystic growth, tumor necrosis resulting in cyst formation and tumor origination in pre existing simple renal cyst. Clear cell papillary RCC is a cystic lesion with cyst wall lined by clear cells and much of the tumor exhibiting papillary architecture^[3].

The imaging and management protocols is followed by considering The Bosnaik system. Under this system cysts coming under category 1 and 2 require only follow up. Where as the risk of malignancy increases with further categories^[4].

The cystic RCC's carry a favourable prognosis because of the absence of local invasiveness such as vascular or lymph node or into healthy renal parenchyma^[5]. Cystic RCC's present with a variable diversity of lesions such as multilocularcystic RCC. Papillary cystic RCC presents with a loculated cyst with hemorrhagic fluid within which surrounded by a thick pseudo capsule^[1]. Further under microscopy Suzigen et al and Han et al have considered Furhrman nuclear grading as an important prognostic indicator where in a favourable outcome was found in patients with grade 1 and 2 in terms of pre operative growth and survival. A nephron sparing surgery can be considered in such cases presenting with small size of cystic RCC with low nuclear grades^[2].

Conclusion

Cystic clear cell RCC carries an excellent prognosis with no pre surgical increase or post surgical metastasis or recurrence of tumor. A possibility of diagnosis needs to be considered in benign cysts and should be not be misdiagnosed although both benign cysts and Clear cell RCC share similar imaging and clinical manifestations.

Conflicts of Interest

“The authors declares that there is no conflict of interest regarding the publication of this paper.”

Funding Statement

Self funding

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