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A Case Report on Multilocular Cystic Renal Neoplasm of Low Malignant Potential

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Abstract: ***Multilocular cystic renal neoplasm of low malignant potential (MCRNLMP) is a rare variant of clear cell RCC with good prognosis and often present as multilocular lesions. Due to the similarities in morphological appearance these two entities often pose a diagnostic dilemma to the pathologists. With the help of immunohistochemistry and morphological criterias provided by the WHO, a diagnosis can be made without much hassle.***

Keywords: cystic, immunohistochemistry, multilocular, renal

1. Introduction

Multilocular cystic renal neoplasm of low malignant potential (MCRNLMP) are cystic tumors which were previously termed as multicystic RCC due to its pathological similarity with clear cell RCCbut termed so now due to its low malignant potential and excellent prognosis. [1] Here we discuss about a case which was detected incidentally after nephrectomy.

2. Case report

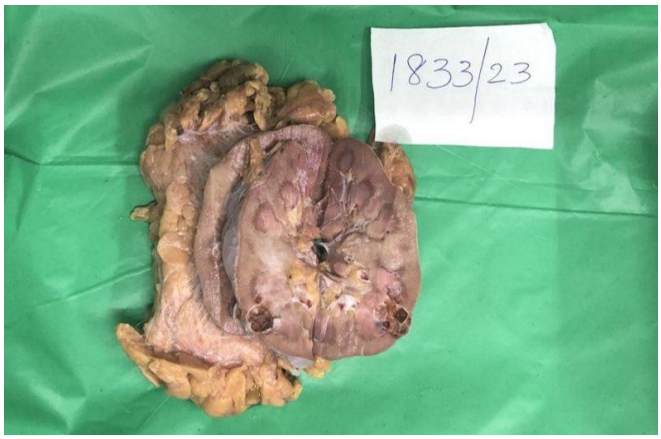
A 37year old gentleman with no other comorbidities presented with complaints of lower abdominal pain.

USG – identified a vesico ureteric junction calculus and a complex cyst with thick internal septations measuring 1.5x1.7 cm in the lower pole of right kidney. No solid component.

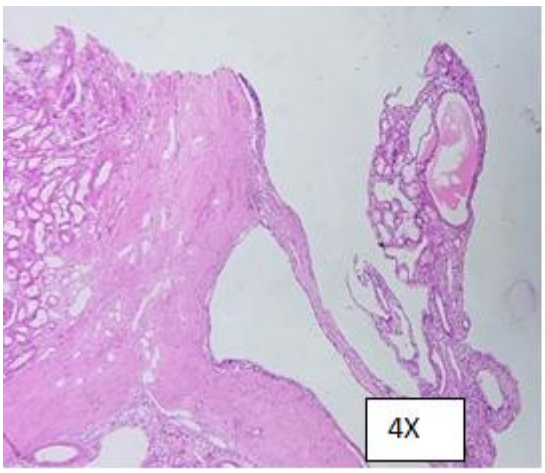
CT urogram – cyst measuring 17 x25 x22 mm with multiple enhancing septa with measured enhancement in interpolar region [Bosniak III].

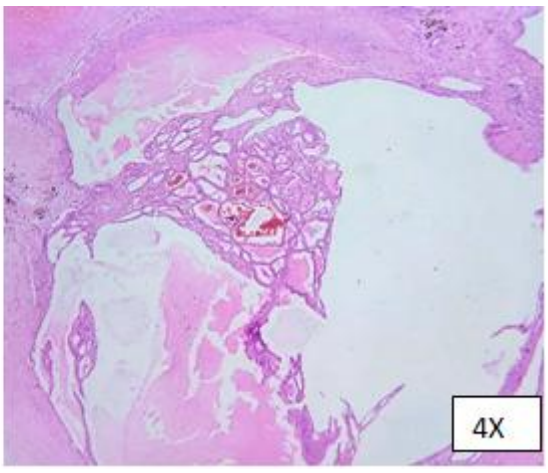
Patient underwent nephrectomy.

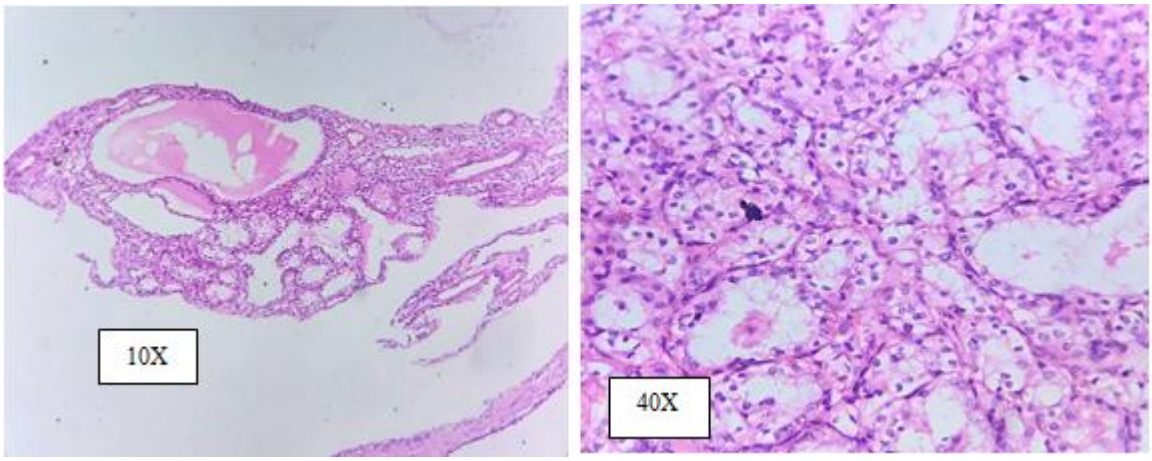
Macroscopy: right radical nephrectomy specimen with portion of attached ureter whole 150 g, measuring 15.5×7.5×6.5 cm. Surface shows a bosselationnear the lateral border of kidney measuring 0.7 cm in greatest dimension. Cut section identified a cortical multiloculated cystic lesion measuring 3 ×1.2×2.5 cm.



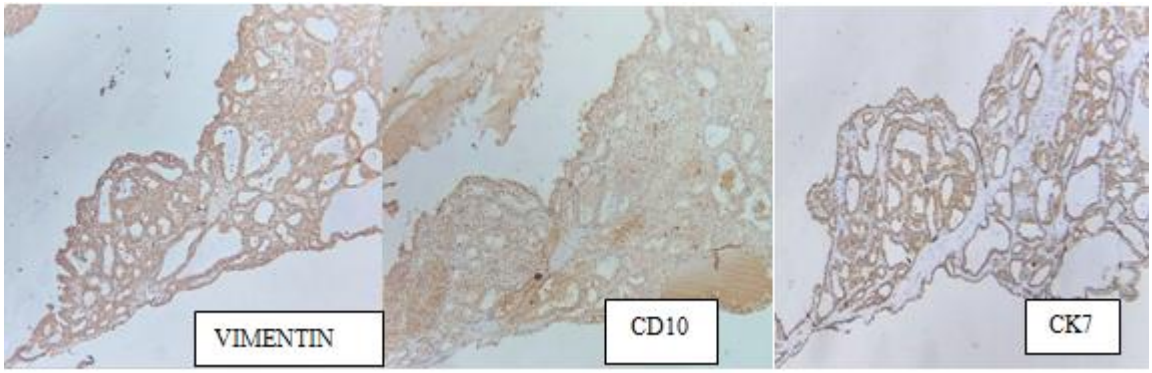
Microscopy: cystic neoplasm composed of cells arranged in acinar pattern and short papillaroid pattern with polygonal cells with clear cytoplasm which also lines the fibrous septae.







Immunohistochemistry: positive for vimentin and CK7, negative for CD10 with background positivity.



Diagnosis – multilocular cystic renal neoplasm of low malignant potential.
Follow up - Post surgery the patient is doing well.

3. Discussion

MCRNLMP is a cystic multiloculated tumor lined by cells with low grade morphology and with indolent behaviour. They were earlier classified under Clear cell RCC but now (2016 WHO tumors of classification) reclassified as a separate one.

They are usually asymptomatic and detected incidentally radiologically (as in our case). USG often reveals well defined multilocular cysts. Bosniak grading is done according to the structure of the cystic lesion, the number of septae present, the thickness and regularity of the septa and wall, the presence of contrast enhancement in the septa, and the presence of calcifications or nodules. [2] Patients can also present with hypertension, hematuria, palpable mass, flank pain.

MCRNLMP show VHL gene mutations as in clear cell RCC. They develop cysts due to defective microtubule formation and degeneration of primary cilia.

Macroscopically they are usually solitary well demarcated masses with fibrous pseudocapsule composed of variably sized cysts ranging from 6mm to 150mm. They are filled with clear, serous, gelatinous or hemorrhagic fluid. There is no solid nodules or necrosis. [3, 4]

Microscopy – shows cysts lining consists of one to a few layers of cells which are bland looking and optically clear. When clusters of cells are present within fibrous septae or septations, they should not alter their contours by expansile growth and should not exceed 1mm in diameter. There should not be any necrosis, atypical mitoses, lymphovascular invasion or rhabdoid or sarcomatoid differentiation. [3, 4]

The differentials to be considered are clear cell RCC with cystic change, Papillary renal cell RCC, tubulocystic RCC
Immunohistochemistry – it is similar to Clear cell RCC that is they show diffuse reactivity for CA IX, EMA, PAX8. But in contrast, they less frequently express CD10 and more frequently express (diffuse) CK7. [3, 5]

These tumors are with excellent prognosis mostly requiring nephron sparing surgery. [2, 6]

4. Conclusion

Multilocular cystic renal neoplasm of low malignant potential is a low grade tumor, presenting as solitary cystic masses with indolent behaviour and excellent prognosis. They can be diagnosed with the help of immunoprofile and morphological criteria which distinguishes them from other cystic renal tumors.

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