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CASE REPORT

A Rare and Unusual Renal Tumour: A Case Report

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ABSTRACT

INTRODUCTION:

Multilocular cystic renal cell neoplasm of low malignant potential is a rare histologically identified form of clear cell renal cell carcinoma included in 2016 Renal tumour classification of WHO. It is a low risk neoplasm with clear cells that have less expansile proliferation than other clear cell variants of renal cell carcinoma with no recurrence and metastasis with few exceptional cases. The incidence of multilocular cystic renal cell carcinoma is estimated to be 1-2 percent.[6,12] Although the WHO classification of multilocular cystic renal cell carcinoma with low malignant potential was reported in 2004. The WHO 2016 categorization of renal cell tumour has been assessed as a special entity with no documented cases of metastasizing and recurrent tumour development.[4]

We present a rare clinical case of multilocular cystic renal cell neoplasm with low malignant potential, which should not be mistaken with other differential diagnoses such as cystic necrosis in conventional renal cell carcinoma, tubulo cystic renal cell carcinoma, multilocular cystic nephroma, clear cell variant of renal cell carcinoma and other benign multilocular renal tumors.

KEYWORDS | Multilocular cystic neoplasm of low-gradepotential; CD 10; Cytokeratin 7 (CK 7); PAX8; Carbonic anhydrase IX and EMA.

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INTRODUCTION

Multilocular cystic renal cell neoplasm of low malignant potential was described in classification of WHO 2016 as neoplasm composed entirely of numerus cyst with septa. The lining cells are single layer of clear cells occasionally with stratification. There is no infiltrative growth and this does not metastasize or reoccur again. This neoplasm accounts for 1-2% of renal neoplasm.[6,12] It is usually unilateral. Radiological diagnosis of this tumor can be done. VHL gene mutation and Chromosome 3p deletion has been identified with these tumors.[8]

Histopathologically multiple cysts of varying size are seen separated by thin septa and filled with clear, gelatinous or serous fluid. Sometimes the cystic spaces also contain hemorrhagic debris. There are cysts are lined by a layer of clear cells with small nuclei and no nucleoli[4,13] and here we present one such case.

CASE REPORT

A 36 year old male came with complaints of abdominal pain for past 5 years. USG abdomen done 4.4x3.6x4.6 cms cystic lesion in left perinephric space was diagnosed andas?

Cystic renal cell carcinoma. CT chest done was normal. Intra operative findings 4.5 cm exo and endophytic tumour in the left upper pole of kidney. Partial nephrectomy done and specimen sent for histopathological study.

We received a left partial nephrectomy specimen measuring 5.5x7x3cms with a separate multiple fatty tissue bits measuring 1x0.5x0.2cms. Cysts were visualised on the surface. Cut surface showed multiple cysts extending from the pole to 2 mm above the resected end, size of neoplasm measuring 4.4x2.8cms. On cutting through extruded pale-yellow fluid drained out with multiple cystic spaces. The large cyst was measuring 1.5cms and smallest one measured 0.3cms in diameter.

Histopathology, sections studied showed fully encapsulated cystic neoplasm composed of numerous variably sized cystic spaces lined by single layer of clear cells, stratification seen in few places. Nuclei are small and does not show pleomorphism. The cavities are filled with eosinophilic material. Intervening stroma composed of loose oedematous fibrocollagenous tissue with thin walled blood vessels and sparse inflammatory cells No necrosis or hemorrhage noted. The renal parenchyma surrounding the tumor showed normal glomerulus, tubules and thick walled blood vessels. No other lesion or neoplasia identified in surgically resected margin. Immunohistochemistry was done with CK 7 and CD10, which showed positivity for CK7 and was negative for CD10.

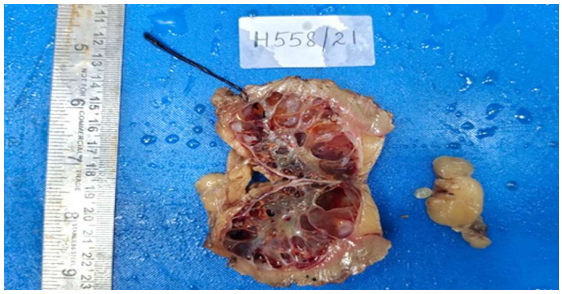

Fig. 1: Gross picture of multilocular renal neoplasm of low grade potential.

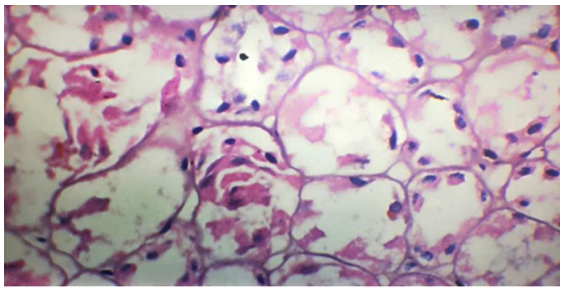

Fig. 2: Clear cell in multilocular cystic neoplasm of low grade potential with tiny nucleus.

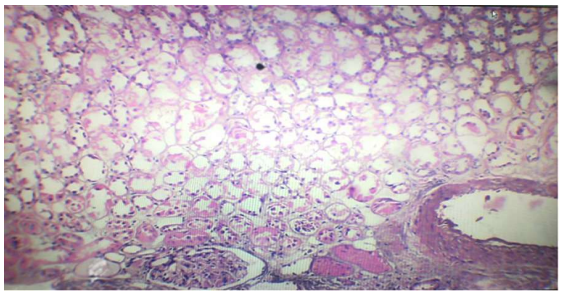

Fig. 3: Multilocular cystic neoplasm of low grade potential showing clear cells.

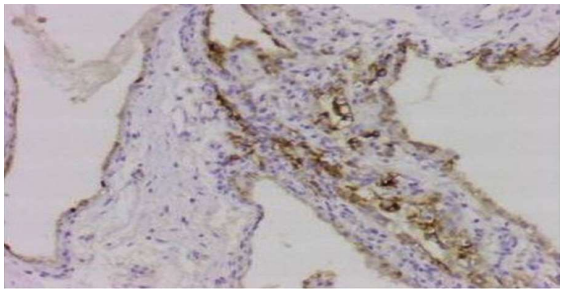

Fig. 4: CK7 positivity seen in epitleial clear cells of multilocular renal neoplasm of low grade potential.

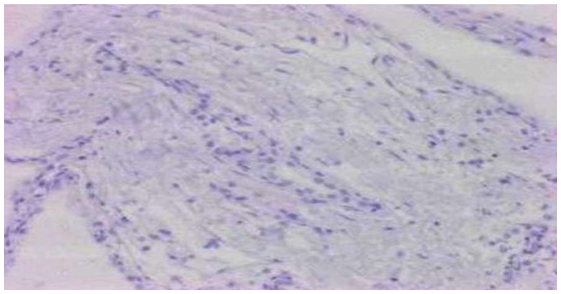

Fig. 5: Multilocular renal neoplasm of low grade potential negative for CD10 in neoplasm seen.

DISCUSSION

Multilocular cystic renal cell neoplasm of Low grade potential is a rare variant of renal neoplasm and distinct subtype of clear cell renal tumour with favourable outcome.[9] Rarely they do extend into perinephric fat and has been reported. The diagnostic criteria have been well described in WHO of classification

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renal tumor 2016. There are three factors which describe this tumour: These tumours present as unilateral, solitary lesion, macroscopically consisting of numerous fluid filled separated cysts of variable size. Rarely the symptoms typically associated with renal symptoms like flank pain, hematuria may be present. In our case, patient presented as abdominal pain.[2]

A series of 76 cases on multilocular cystic renal neoplasm of low malignant potential reported the mean age diagnosis as 46.7+ or -10.5 years. In our case, the age of patient is 36 years. The nuclear grade is low and unrelated to tumour size and TNM stage, suggesting that the current stage criteria may not be suitable for lesion.[2]

In another study of 19 cases which included Immunohistochemistry and were compared with cystic clear cell renal cell carcinoma and cystic tubulepapillary renal cell carcinoma, In Multilocular cystic nephroma and Multicystic renal neoplasm, the pathological study in these study shows 1.5 to 7 cms in maximum diameter. The cyst contains cyst fluid rarely hemorrhagic or turbid fluid. Solid areas are discernible neural nodules were absent. In our study, we could not identify any hemorrhage only clear fluid and only solid areas. Histologically single layer of cuboidal epithelial tumour cells lined the cyst. The cells have clear cytoplasm, small nucleus, no nucleoli, Furrham nuclear grade 1 or 2. In our study, the nucleus was small and no nucleoli was observed. The tumor cells are usually positive for pan Ck-100%, EMA and Ck 7 in more than 80% of cases. CA IX in more than 80% of cases. CD10 is showing lower positivity.[5] We observed CK 7 more than 90% positive in the study sections and negative for CD10.

CD10 positive 63% multilocular cystic renal cell carcinoma and 96% of Renal cell carcinoma, CK 7 seen in 92% of multilocular cystic renal cell carcinoma and 38% of Renal cell carcinoma. Co expression of CK 7 and CA IX may represent Renal cell carcinoma.[7] Molecular methodologies may be required for further typing of renal cell neoplasm. No patient developed recurrence or metastasis. Monosomy 3 is seen in both Multilocular renal neoplasm in 3/3 cases, papillary renal cell carcinoma was seen in 6/7 cases.[1]

Eble et al. proposed the characteristic features of 1. Fibrous wall with increase in size of tumour mass, 2. Inside of tumour has numerous cysts and septa but expandible solid nodules absent, and 3. Septal mass has epithelial clear cells.[14] Further, it was introduced into WHO histological classification of renal tumour in 2004. The first two descriptions were published in the pathologic literature in 1928 and 1957, respectively.[11,14]

Eble et al. originally defined multilocular cystic renal cell neoplasm of low grade potential as a tumour composed of many cysts, the septa of which contained tiny groups of clear cell indistinguishable from grade I clear cell carcinoma.[11,14]

Li. T et al., Murad et al. and Nassir et al. postulated that Multilocular cystic renal neoplasm of low grade potential was a benign entity and has no recurrence or metastatic potential. Therefore, the WHO classification was redefined on 2016.[15,16,2] This stringent application of pathological definition of Solid Renal clear cell carcinoma undergoing necrosis (Furham 3 or 4) has been differentiated from multilocular cystic renal neoplasm of low malignant potential which was (Furham 1 or 2).[10]

Histologically, Grade 2 clear cells seen in solid Clear cell renal cell carcinoma (positive for CD10 and vimentin) when compared to multinodular cystic renal neoplasm of low grade potential which has Grade 1 clear cells positive for CK 7, PAX 8, Carbonic anhydrase IX and EMA.[10]

Another entity called multilocular cystic nephroma which was first described by Edmund. V in his literature following study of 200 cases in 1892 only 13 cases have been reported as cystic adenoma of the kidney with malignant potential.[16] In 1951, T. Powel later classified it into cystic nephroma and cystic partly differentiated nephroma based on the presence or lack of blastemal element, respectively.[17]

Bosniak classification, an improved version published in 2019 for renal cell tumours based

on CT imaging features such as wall, septa, high attenuation, calcifications, and wall/septum enhancement, has classified Multilocular cystic renal cell neoplasm of low grade potential as Category IIF, where most renal masses are benign, but later considered a Category III lesion requiring histological confirmation.[3]

What was once known as "multilocular cystic renal cell carcinoma" is now known as "multilocular cystic renal neoplasm of low malignant potential." The prognosis is good for multilocular cystic neoplasm of low-grade potential following resection as there is no recurrence and no metastasis.[4]

CONCLUSION

We, here present a interestingly rare case of Multilocular cystic renal cell neoplasm of low grade malignant potential which was confirmed histologically. The partial nephrectomy which was done had good prognosis for this patient and was followed up.

Conflict of Interest:
The authors declare that there are no conflicts of interests
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All data associated with this study are presented in the paper.

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