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Case Report

Multilocular cystic renal neoplasm: a rare tumor in kidney

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ABSTRACT

Multilocular cystic renal cell carcinoma is a rare cystic tumour of kidney with excellent outcome. It is usually included in the group of tumours of undetermined malignant potential with low nuclear grade. We are presenting a case of 61 year. old female came with history of giddiness for 1 week and weight loss of 20 kg in 2 years. Imaging was suggestive of cortical based tumour arising from upper pole of right kidney. Right side partial nephrectomy was done. On histopathologic examination it was found to be MCRCC, stage 1 with Fuhrman nuclear grade 1. Immunohistochemistry with epithelial membrane antigen and cytokeratin-7 confirmed the diagnosis.

Keywords: Cystic tumour. Multilocular cystic renal cell carcinoma, Nephrectomy

INTRODUCTION

Multilocular cystic renal cell carcinoma is a rare subtype of renal neoplasm with good prognosis and composed of thin fibrous septa lining multiple cystic spaces.[1] MCRNLMP is described as a tumour completely composed of multiple cysts, with septa containing clear cells without expansive growth, and morphologically indistinguishable from low-grade CCRCC.[2] According to FISH analysis, most MCRCCs had chromosome 3p deletions, and 25% of them had VHL gene mutations.[3] Preoperatively distinction between tumours with acquired cystic lesions and cystic changes is difficult by imaging. When cystic renal masses appear on radiographs as per Bosniak classification, enhanced computed tomography (CT) and magnetic resonance imaging are useful for identifying malignancy.[4] Confirmatory tests for MLCRCC based on IHC with Carbonic anhydrase IX, Epithelial membrane antigen (EMA), Cytokeratin (CK7) and CD10. We present a case of MCRCC diagnosed in 61-year-old female patient. EMA and CK-7 are positive in our case.

CASE REPORT

A 61 year. old female came to urology OPD with complains of giddiness for 1 week and history of weight loss 20 kg in the last 2 year. USG (A+P) suggestive of a well-defined partly exophytic heterogenous predominantly isoechoic lesion measuring 1.9x1.4 cm noted in the upper pole of right kidney. On colour doppler it shows vascularity. No evidence of calcification. Renal function test and pre-operative investigations were within normal limits. CECT section of a small heterogeneously enhancing cortical based tumour measuring 3x1.8x1.8 cm noted in upper pole of right kidney She underwent right partial nephrectomy under GA. Intra operative findings suggestive of 3x2 cm tumour in upper pole in posterolateral aspect of right kidney.

Macroscopy

A grey-white to grey-brown globular mass with perinephric fibro fatty tissue measuring 7x3x1. 5cm.External surface shows nodular growth with a cyst

measuring 3x3x1.5 cm. Cut surface shows well circumcised nodule with grey black areas and cystic component measuring 3x2 cm.

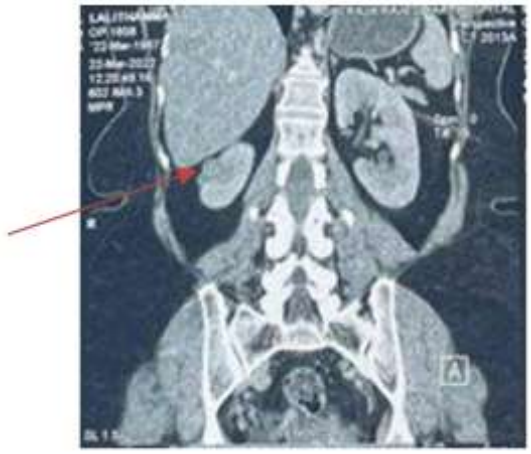

Figure 1: CECT.

Figure 2: Intra-operative image of renal mass.

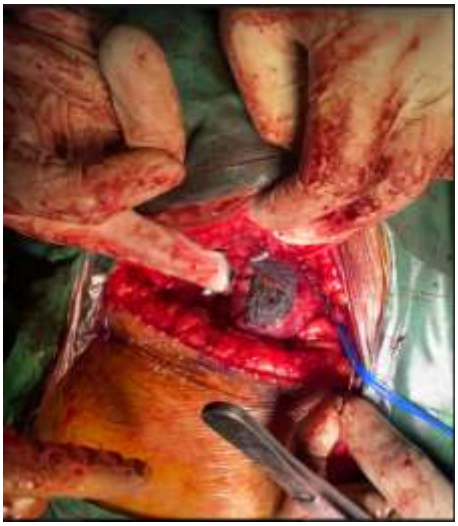

Figure 3: Post partial nephrectomy.

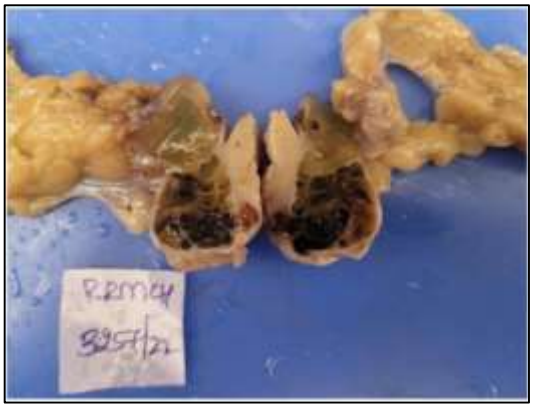

Figure 4: Gross photograph of cut section showing blackish brown cystic areas.

Microscopy

Multiple sections study shows structures of kidney with renal parenchyma and a well circumcised partially encapsulated metacystic mass comprising of one to several layers of tumors cells lining delicate fibromuscular trabeculae. The individual tumour cells are large with abundant clear cytoplasm and Fuhrman grade 1.

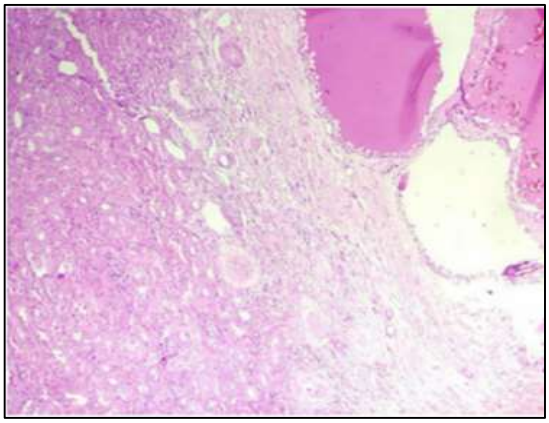

Figure 5: Photo microscopy showing normal renal parenchyma on the right and cystic changes on left.

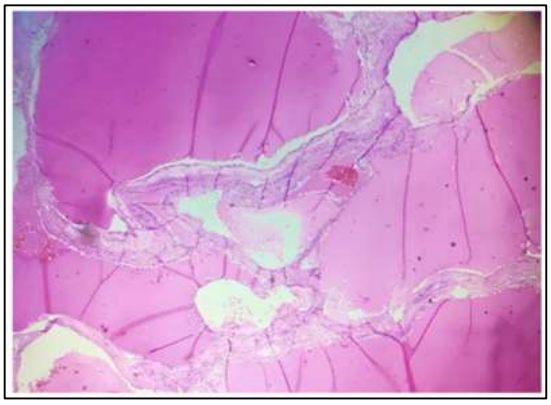

Figure 6: Photo microscopy showing multiple cysts separated by thin septa 4x H and E.

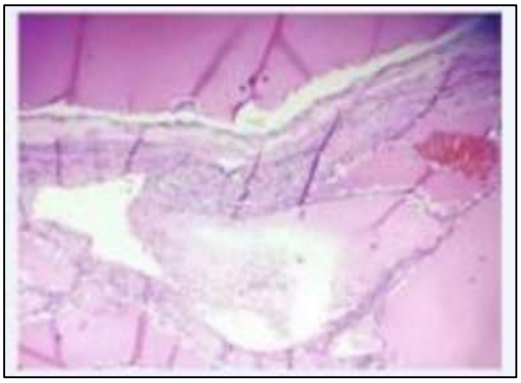

Figure 7: Photo microscopy shows cyst containing proteinaceous fluid lined by tumor cells 10x H and E.

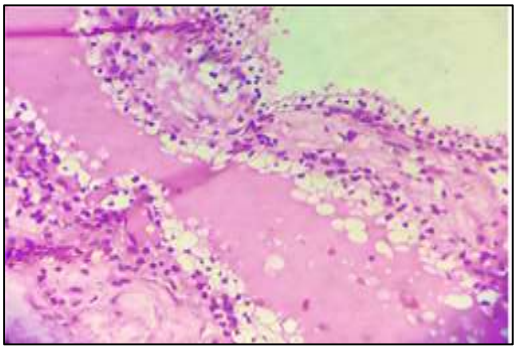

Figure 8: Photo microscopy shows neoplastic cells with clear cytoplasm arranged in layer Fuhrman grade 1 40x H and E.

The cystic space contains proteinaceous eosinophilic fluid and hemorrhage along with cyst macrophages and hemosiderin laden macrophages. The cyst wall at places shows lining by cuboidal to flat epithelium and by foamy macrophages. Small collection of tumour cells is seen in the intervening fibrous septa.

DISCUSSION

Among the categories of cancers of the urinary tract and male genital organs included in the 2016 WHO classification, multilocular cystic renal cell carcinoma became multilocular cystic renal neoplasm of low malignant potential.[2] This rare neoplasm affects mostly middle-aged adults with slight male pre-dominance. In the differential diagnosis of MCNLMP, cystic lesions include benign renal cortical cysts (absence of clear cells within the wall of cyst), ccRCC with cystic and/or regressive changes (cysts filled with hemorrhage, necrosis and hemosiderin deposits; keyboard like arrangement of nuclei), cystic nephroma (cysts lined by bland nonclear cells with presence of ovarian stroma), cystic clear cell papillary RCC (contains clear cells with low-grade nuclei, but differentiating feature is the presence of papillary architecture), MiT family of tumours (high grade tumours with focal solid areas), and tubulocystic carcinoma (cells have eosinophilic cytoplasm with high-grade nuclei instead of clear cells).[5] Most of the cases are asymptomatic and incidentally detected. Chromosomal abnormalities like 3p deletion (74% cases) and VHL gene mutation (25% cases) were associated.[3] According to the Bosniak classification defining cystic tumours of the kidney on computed tomography (CT), MCRNLMP belongs to category 2, 2F or 3.[6] On histology they appear as well defined multilocular cysts filled with serous, gelatinous, haemorrhagic, or mixed fluids. The cysts are lined by single or multiple layers of tumour cells with abundant clear or granular cytoplasm with low grade nuclei.[3] The tumour cells show positive staining for cytokeratin 7 and epithelial membrane antigen (EMA).[7]

Follow up

Patient is on regular follow up as per NCCN guidelines, till now there is no evidence of recurrence or metastasis.

CONCLUSION

Because of rare nature and variable diagnostic criteria exact incidence of MLCRCC still unknown. < 1% of renal cell neoplasm accounts for MLCRCC.[8-12] Multilocular cystic renal cell carcinoma of low malignant potential has similar genetic profile and histopathological characteristics to that of renal cell carcinoma but different prognosis and metastasis. This makes it important to differentiate it from other variants of RCC. No expansive growth, solid nodular growth of the clear tumor cells, necrosis, vascular invasion or sarcomatous changes differentiate it from other variants of RCC. It is cystic lesion of the kidney with excellent prognosis and no metastasis so minimally invasive nephron sparing surgery is performed.

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Ethical approval: Not required

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