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Histopathology

MULTILOCULAR CYSTIC RENAL NEOPLASM OF LOW MALIGNANT POTENTIAL:A CASE REPORT

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ABSTRACT Multilocular Cystic Renal Neoplasm of Low Malignant Potential (MCRNLMP) accounts for <2% of renal tumours.It is an indolent neoplasm with an excellent prognosis.Nephron sparing surgery is the preferred treatment.Distinguishing this tumour from the clear cell renal cell Carcinoma (CC-RCC) with cystic degeneration and other cystic tumours of kidney may be challenging. We report herewith a case of MCRNLMP of right kidney in a 75 year old male.

KEYWORDS : Multilocular cystic renal neoplasm of low malignant potential ,clear cell renal cell carcinoma

INTRODUCTION

- Multilocular cystic renal neoplasm of low malignant potential was previously known as multilocular cystic renal cell carcinoma, but was reclassified as a low malignant potential tumour in the 2016 classification due to its excellent prognosis [1]
- It accounts for 0.5-2.5% of all renal tumours and is most prevalent in 6th decade of life[2]
- Distinguishing MCRN-LMP from Clear cell renal cell carcinoma with extensive cystic change or regressive cystic appearance may be challenging
- MCRN-LMP is a very slow progressing disease and if possible,nephron sparing surgery is the preferred treatment [3]

CASE REPORT

A 75 year old male presented to opd with complaints of pain in the right lumbar region and swelling of legs since 2 months .No rbcs were seen in the urine microscopic examination.

Ultrasonography revealed a multiloculated exophytic cystic lesion in the right kidney measuring 9.9x5.4 cm arising from the interpolar and lower polar region having septations.

CECT of right kidney showed large exophytic (>50% exophytic) multicystic lesion in the mid pole and lower polar region measuring7 x6.2x5.8 cm with multiple enhancing septations.The mass was seen abutting the D2 segment of duodenum and right renal vein.Right nephrectomy was performed.

Grossly,the right nephrectomy specimen measured 12x7x5 cm and included right kidney measuring 8.5x4x3 cm and perinephric fat ,ureter measuring 6cm in length.Cut section showed irregular multilocular cystic tumour in the interpolar and lower polar region measuring 7x6.5x6 cm pushing into the renal hilum .The cysts ranging in size from 4cm to 0.5 cm , filled with gelatinous fluid.No expansile growth or areas of necrosis were present [Figure 1]

Microscopic examination of the tumour revealed thin,fibrous septa lined by clear cells with low grade nuclei without prominent nucleoli (WHO/ISUP grade 1-2).Focal multilayering and calcification was seen at few places.No solid areas,no necrosis,no atypical mitotic figures seen.No lymphovascular/fat invasion was seen.No rhabdoid /sarcomatous areas seen. Thus the diagnosis of multilocular cystic renal neoplasm of low malignant potential (WHO/ISUP grade1-2) was made.[Figures2 -6]

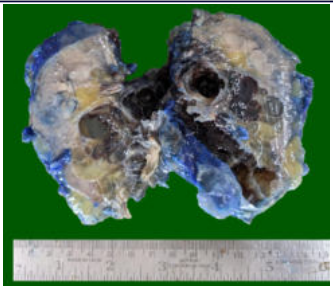


Figure 1: Cut section of the Right nephrectomy shows irregular multilocular cystic tumour with thin septae filled with gelatinous fluid.No necrosis/solid growth present

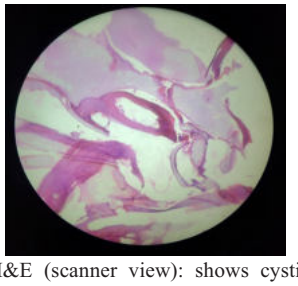


Figure 2: H&E (scanner view): shows cystic architecture with variable sized cysts filled with proteinaceous fluid

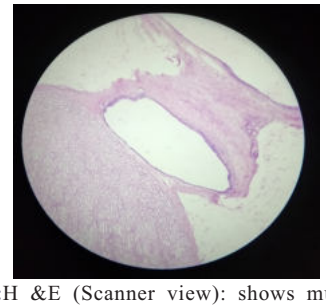


Figure 3:H &E (Scanner view): shows multiple cysts with surrounding normal renal parenchyma

Figure 4:H&E (low power) showing multiloculated cysts lined by single layer of cells

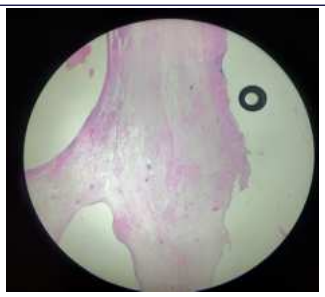


Figure 5: H&E (high power) showing the cyst lined by clear cells with low nuclear grade

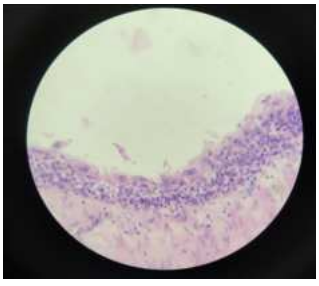
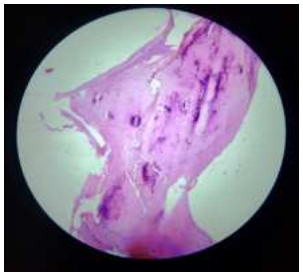


Figure 6:H&E (high power):Septa showing foci of calcification



DISCUSSION

Multicystic renal neoplasm with low grade malignant potential (MCRNLMP) is an indolent neoplasm .It was previously known as multilocular cystic renal cell carcinoma but the WHO 2016 classification of kidney tumours reclassified it as a low malignant potential tumour[1]. It accounts for 0.5-2.5% of all renal tumours and 10% of all cystic renal neoplasms[2][4]It can occur in the age group of 18-84 years but most prevalent in the sixth decade of life.Males more commonly affected than females in the ratio of 2:1[5]. In majority (95%) of the patients it is unilateral and solitary ;whereas in the rest 5% it is bilateral and multifocal. Upto 75% of cases have a chromosome 3p deletion, and upto25%have von hippel Lindau syndrome. Clinically, 80% of cases are asymptomatic and are discovered incidentally. Whereas the remaining patients present with symptoms such as hypertension, hematuria, palpable mass, flank pain(but not the classic triad of Renal cell carcinoma i.e gross hematuria, pain and palpable mass[6].Radiological studies suggest Bosniak IIF-III cysts [7]. Multilocular, moderately complex cyst with no solid areas/mural nodules.

Grossly,it is well circumscribed with fibrous pesudocapsule , size usually is <5cm ,with variable sized cysts with thin septa.The cysts are filled with clear,serous,gelatinous or hemorrhagic fluid.No mural solid nodules or areas of necrosis are seen[8]

Microscopically,the cysts consist of thin fibrous septa lined by clear cells .It consists of low grade nuclei without prominent nucleoli (WHO/ISUPgrade1-2).The septa may contain foci of calcification or ossification.The tumour does not exhibit necrosis, sarcomatoid or high grade foci. The cells are morphologically similar to those of low grade RCC(renal cell carcinoma) but there is no evidence of expansive growth.[9]

Distinguishing MRCP-LMP from CC-RCC with extensive cystic change or regressive cystic appearance may be challenging.Other entities to be considered in the differentials are benign renal cortical cysts, clear cell papillary renal cell tumour,cystic nephroma, tubulocystic renal cell carcinoma and TFE3 translocation tumours.

PAX8,CAIX and CK7 stain positively,while AMACR staining is negative and KI67 is very low.

Molecular characteristics are similar to those of CC-RCC (clear cell renal cell carcinoma).Upto75% of cases have a chromosome 3p deletion and upto 25% have VHL gene mutations.NGS study have identified 6 genes SETD2,FGFR3,BCR,GIGYF2,KMT2C and TSC2 for differentiating MCRNLMP from CCRCC with cystic change[10] Since MCRN-LMP is a very slow progressing disease;resection i.e. nephron sparing surgery is the preferred treatment[3]

CONCLUSION

MCRNLMP is a rare tumour of kidney with an excellent prognosis with nephron sparing surgery.Thus, differentiating it from Clear cell Renal cell carcinoma with cystic change and other cystic tumours of kidney is important.

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