
Mucinous Tubular and Spindle Cell Renal Cell Carcinoma with Associated Renal Pelvic Stone**Dr. Sunirmal Choudhury¹, Dr. Susanta Kumar Das², Dr. Suchandra Ray³, Dr Dilip Kumar Pal⁴**¹Assistant Professor, Dept. of Urology, IPGME & R, Kolkata, India²MCh Urology Resident, Dept. of Urology, IPGME & R, Kolkata, India³Associate Professor, Dept. of Pathology, IPGME & R, Kolkata, India⁴Professor & HOD, Dept. of Urology, IPGME & R, Kolkata, India***Corresponding author**

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Abstract: We report a case of 40 years old gentleman with mucinous tubular and spindle cell renal carcinoma (MTSCRC) with associated renal pelvic calculus, a rare clinical entity of the kidney. Initial presentation was that of right side flank pain. Radiological evaluation revealed a right renal mass along with a pelvic calculus. After subsequent evaluation he underwent right radical nephrectomy. The histopathological examination was suggestive of mucinous tubular and spindle cell renal cell carcinoma. This is a very rare case and treatment protocol is yet to be established in such a scenario.**Keywords:** renal carcinoma, mucinous tubular, spindle cell

INTRODUCTION

Mucinous tubular and spindle cell renal carcinoma (MTSCRC) is a relatively rare variant of kidney tumor. In literature, it is documented as a tumor of low malignant potential and may metastasize to local lymph nodes and is occasionally associated with nephrolithiasis. However, there is no definite treatment protocol in published data. Ours is a case of MTSCRC with associated nephrolithiasis.

CASE REPORT

Forty years of age gentleman presented with episodic right sided dull aching pain for six months along with one episode of gross hematuria. He was initially treated conservatively at a local hospital with suspicion of infective etiology. However his pain persisted for which he visited our hospital. He had no history of loss of weight or appetite, of fever or constitutional symptoms. He was non-diabetic, normotensive and a chronic smoker. His physical

examination was unremarkable. Pallor was absent with no palpable lymphadenopathy. Abdominal examination did not reveal any abnormality. The renal angle was non-tender.

At presentation in our hospital, complete blood count showed a white cell count of 8,000/mm³ with 60% neutrophils, 28% lymphocytes and 8 % monocytes, hemoglobin of 10.5 gm./dl and platelet count of 200,000/mm³. Blood biochemistry reports revealed serum urea of 36mg/dl and a creatinine of 1.0 mg/dl, calcium of 9.1mg/dl, albumin of 3.2mg/dl, alkaline phosphatase of 67 IU/L, SGOT of 23U/L and SGPT 22U/L, fasting glucose of 102mg/dl. Urine routine showed 6 red blood cells/ HPF and pus cells of 2-3/HPF. Urine culture was sterile.

Ultrasonography revealed a 19×10 mm mass lesion in the renal pelvis of the right kidney (Figure 1).



Fig-1: Ultrasonography of the right kidney showing the right renal pelvis mass

Contrast enhanced CT scan revealed a small right kidney with mild hydronephrosis along with

21×25×15 mm soft tissue density mass lesion occupying the interpolar region (Figure 2).

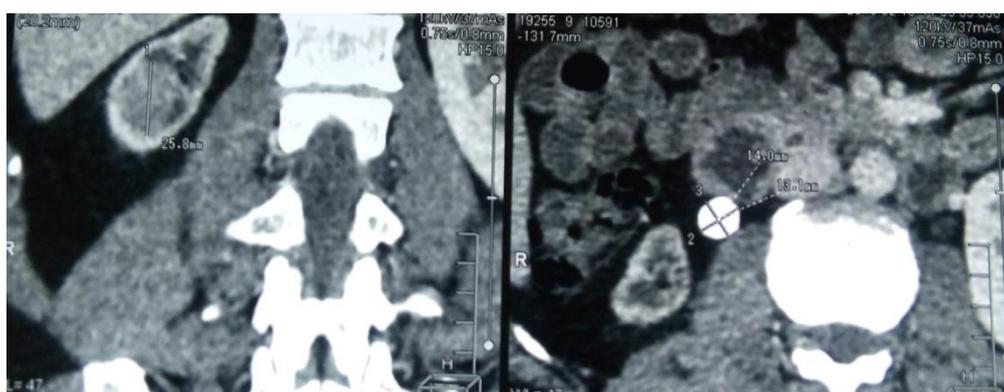


Fig-2: Contrast enhanced CT scan of the abdomen showing the right renal space occupying lesion in the pelvis along with a pelvic renal calculus

The mass showed contrast enhancement of >15 Hounsfield unit in the arterial phase. There was a 15 mm calculus also seen in the pelviureteric junction of the right kidney. The left kidney was normal. In view of the above mentioned clinical and radiographic picture, a probable diagnosis of right renal mass lesion was diagnosed. The contracted right kidney with central location of the tumor prompted us to offer him with the option of right radical nephrectomy.

He underwent right radical nephrectomy. The post-operative course was uneventful. He was discharged after five days. A detailed pathological examination of the specimen was done. Grossly, on cut-section there was a brownish growth measuring 2x1.5 cm noted in the medullary region of the kidney specimen (Figure 3).



Fig-3: Gross cut-section of the right renal tumour showing centrally located tumor brownish cut surface measuring 2x1.5 cm in size

Multilocular cystic spaces were found along with one calculus in the renal pelvis. Histologically, there were elongated interconnected tubules with slit like lumen. The lining cells were low cuboidal with small amount of eosinophilic cytoplasm and bland

nuclei. Mucinous stroma was noted focally. An inflammatory infiltrate consisting of lymphocytes, plasma cells and neutrophils were also present in the stroma (Figure 4).

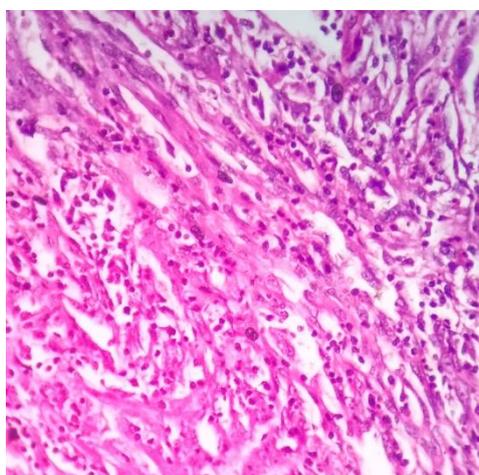


Fig-4: Histopathological (H&E, 400X) photograph of the MTSCRC showing mucinous stroma with inflammatory infiltrate with low cuboidal lining cells

Sections from the other areas of the kidney showed features of chronic pyelonephritis. Immunohistochemical studies revealed neoplastic cells of tubular and spindle cell origin. The cells stained positive for PAX2, cytokeratin CK7 and alphas-methylacyl-CoA racemase (AMACR). The whole picture was that of a mucinous tubular and spindle cell carcinoma.

Post-operative course was uneventful. On six months follow up there was no recurrence seen on imaging study.

DISCUSSION

Mucinous tubular and spindle cell carcinoma of the kidney is a relatively recently identified denomination under the low grade renal epithelial neoplasm of the kidney. This specific entity was first recognized in the World Health Organization Consensus conference on the classification of renal neoplasm in December 2002. In 2004 World Health organization tumor classification recognized this as a distinct entity [1]. Currently this terminology is descriptive that reflects three salient histological

features – tubules, mucinous stroma and spindle cell areas.

Although ours was a male patients, this tumor has a female predominance [male: female = 1:4]. The age at presentation of these tumors ranges between 21 to 81 years. The right kidney is more commonly involved. These tumors usually present with an asymptomatic mass detected in ultrasound. Often flank pain may be present along with hematuria [1-3]. In 80% cases, these tumors are generally confined to the kidney (pT1 or pT2).

In literature, there has been an association with nephrolithiasis as was seen in our patient who had a concomitant renal pelvic calculus [4]. Association with other renal abnormality like simple renal cyst, papillary adenoma, synchronous renal cell carcinoma [3] or angiomyolipoma [5] has been reported. There is a single case report of its coincidental endemic association with tuberculosis [6].

Gross pathological examination of these tumors generally show a centrally located tumour that has a uniform homogenous tan, gray cut surface with occasional areas of necrosis. Usually these tumors are small, 2-4 cm in width; however size may range between sub-centimeters to as large as 18 to 20 cm in diameters.

Histologically, there are elongated curvilinear and stretched tubules which are separated by mucinous stroma of variable amount. These parallel and collapsed tubules give a spindle cell appearance. When this pattern predominates it may mimic a tumour of mesenchymal origin. They may have foam cells, lymphocytic infiltrates with occasional appearance of psammoma bodies. The nuclei are uniform with low nuclear grade. Occasionally, neuroendocrine differentiation [7] and sarcomatoid differentiation has also been reported [8].

Epithelial markers, especially AMACR, CK7, EMA and Vimentin, have been reported to be positive in 80-100% of cases [9]. Fluorescent in situ hybridization (FISH) and comparative genomic hybridization (CGH) was used first to demonstrate the characteristic multichromosomal loss ((-1, -4, -6, -8, -9, -13, -14, -15 and -22) which were further reported indifferent studies [9,10]. In addition, gains of chromosomes (+7, +16, +17, +20) have also been reported.[9,10]

Mucinous tubular and spindle cell carcinoma of the kidney is very distinct entity and should not have a diagnostic problem. Certain variations in the histopathology, such as paucity of mucin and abundance of spindle cells may create a diagnostic dilemma. Differential diagnosis of sarcomatoid variant of renal cell carcinoma may be considered in case of

dominance of spindle cells, however in mucinous tubular and spindle cell carcinoma the spindle cells are uniform in their architectural pattern with low nuclear grade and lack pleomorphic nuclei or increased mitotic activity or sheets of necrosis as seen in the sarcomatoid renal cell carcinoma. The spindle cell predominance may also be confused with neoplasms of mesenchymal origin or that of inflammatory myofibroblastic tumor. Sometimes the morphological features may overlap with papillary renal cell carcinoma and diagnosis may be difficult.

Classical mucinous tubular and spindle cell sarcoma has a favorable prognosis. Complete surgical excision in the form of radical nephrectomy is the best treatment option. However because of usual low pathological tumour stage, partial nephrectomy may be a therapeutic option [1]. There are reports of few post-surgical excision local recurrence, occasional involvement of the loco-regional lymph nodes. Few cases of distant metastasis and tumour associated deaths have been reported [4, 8, 9, 11]. Usually, metastasis in these tumors, are associated with atypical histologic features like the sarcomatoid differentiation or higher nuclear grade. There is a case report, in which the patient presented with extensive regional and distant metastases, but both primary and metastatic tumour showed the typical histology of bland cuboidal or spindle cells lacking pleomorphism, mitoses, and necrosis [11]. Hence, it is of utmost importance for regular follow up of these patients with clinical examination and cross-sectional imaging. No treatment protocols for metastatic disease had been published till date. Larkin et al reported the first index case of the use of sunitinib, the tyrosine kinase inhibitor, in metastatic mucinous tubular and spindle cell carcinoma of kidney with both symptomatic and radiological response [12].

CONCLUSION

Mucinous tubular and spindle cell renal carcinoma (MTSCRC) with associated renal pelvic calculus is a rare clinical entity of the kidney. Treatment protocol is still undefined and further studies are needed to establish proper management and follow-up plan.

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