

A Rare Renal Neoplasm with Dimorphic Histology and Mucinous Stroma

Sunitha Ramachandra*, Lakshmi Rao, Masoud Al Kindi

Department of pathology, Armed Forces Hospital, Muscat, Sultanate of Oman

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ABSTRACT

Mucinous tubular and spindle cell carcinoma (MTSCC) is a rare and recently described histologic variant of renal cell carcinoma (RCC). Usually considered as a tumor of low malignant potential it is important to appreciate the characteristic histologic features to arrive at the correct diagnosis. We report a case of MTSCC in a female aged 54 years who presented with a long standing vague loin pain. Diagnostic work-up showed a renal mass suggestive of RCC. With the preoperative diagnosis of RCC followed by nephrectomy showed a completely different histology of a rare tumour with tubules, spindle cells and mucinous stroma consistent with MTSCC, a low grade tumour with good prognosis.

***Corresponding author:**

Dr. Sunitha Ramachandra, PO Box 726, Seeb PC 111, Muscat, Oman.

Phone: +91- 96824331357

E-mail: sunithamanjunath98@yahoo.co.in



Introduction

Mucinous tubular and spindle cell carcinoma (MTSCC) is a rare subtype of renal cell carcinoma (RCC), a new entity incorporated in the 2004 WHO classification previously described as unclassified RCC.^[1] It is believed to be a low grade myxoid tumour with spindle cell change and distal nephron differentiation.^[2] Considered to behave in a benign fashion with a good patient prognosis, limited nephrectomy is emphasized.^[3] Hence the need to be aware of this unusual neoplasm thus differentiating it from its close mimics of a higher grade RCC.

Case report

A 54-year-old female patient had been suffering with chronic left loin pain since a few years. She had no other complaints and insignificant past history. She presented at our hospital (a tertiary referral centre) with detailed reports of her investigations done elsewhere. CT abdomen revealed a massive heterogenous left renal mass arising from the mid and upper pole with internal echoes compressing the renal parenchyma and involving the renal pelvis. Renal vessels were not involved. This large mass displaced the jejunal loops, descending colon and pancreatic tail. Right kidney, suprarenals, ureters and urinary bladder appeared normal. A radiological diagnosis of a malignant neoplasm with a differential of RCC or TCC (transitional cell carcinoma) was suggested. With the above findings and the possibility of a malignant tumour she was treated with left nephrectomy.

Grossly the specimen weighed 1905 grams consisting of a large lobulated soft mass difficult to orient in its anatomic position with a ureteric stump. Cut surface showed a necrotic, solid pale to golden brown tumour replacing the upper and the middle renal parenchyma measuring 16.5 x 14.5 x 12.5 cm involving the renal calices but not the renal pelvis or ureter (Figure 1). The lower pole showed unremarkable renal parenchyma. Suprarenal gland and Gerota's fascia were not seen.

Microscopy showed a well differentiated dimorphic (epithelial and spindle cells) tumour with a pseudocapsule (Figure 2). First component is composed of tightly packed small elongated parallel tubules lined by cells with round nucleus, fine granular chromatin, prominent nucleoli, mild anisonucleosis, eosinophilic cytoplasm and occasional mitoses (Figure 3). Second component is composed of whorls and fascicles of spindle shaped cells with similar morphology. Stroma showed a focus of foam cells, eosinophils, lymphocytes, perivascular plasma cells and thick walled blood vessels. Cystic degeneration with haemorrhage and cholesterol clefts

were noted. Renal pelvis and ureter were free of tumour. IHC (immunohistochemistry) showed epithelial and spindle cells positive for CK 7, Cam 5.2, CK 18, Pan CK and negative for CD 10 and CD 15 (Figure 4). EMA was positive in epithelial cells only. Vimentin showed weak positivity in spindle cells and negative in epithelial cells. The above microscopic and IHC findings were consistent with MTSCC.

Discussion

MTSCC is a rare renal malignant neoplasm recently included in the WHO classification previously called as unclassified renal carcinoma.^[1] This is a low grade polymorphic neoplasm composed of bland tubules set in a mucinous stroma with spindle cell areas.^[4] The tumour shows a female predominance with 1:4 male-female ratio and wide age distribution ranging between 17 to 82 years with a mean age of 52 years.^[1,3] One hundred cases of MTSCC have been reported till-date.^[3] Most of them are incidentally diagnosed while being investigated for some other indications.^[1] They are usually asymptomatic. A few large tumours present with flank pain, hematuria and anaemia.^[1,3] Due to its massive size, our case presented with loin pain and routine blood tests showed low haemoglobin 9.5 gm/dL and reduced red blood cell count ($3.6 \times 10^{12}/L$). Alternately called as low grade collecting duct carcinoma, low grade tubular-mucinous renal neoplasm and low grade myxoid renal epithelial neoplasm with distal nephron differentiation.^[5] Majority of these tumours are

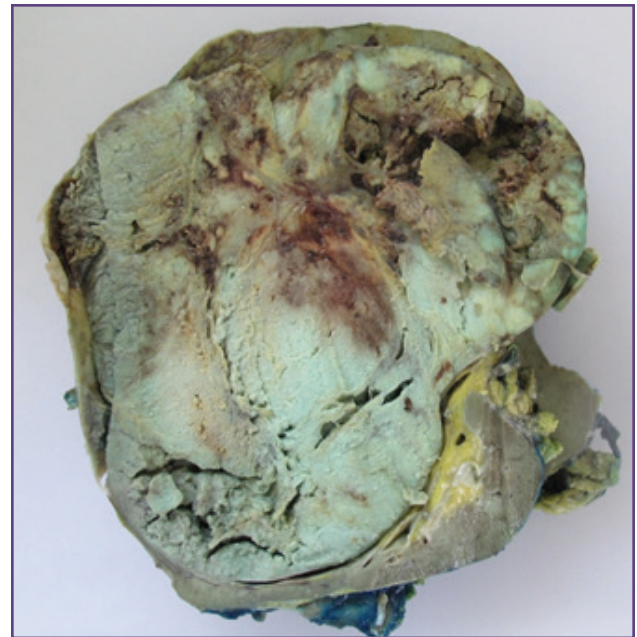


Fig. 1: Gross picture - golden brown tumour with necrosis and normal compressed kidney.

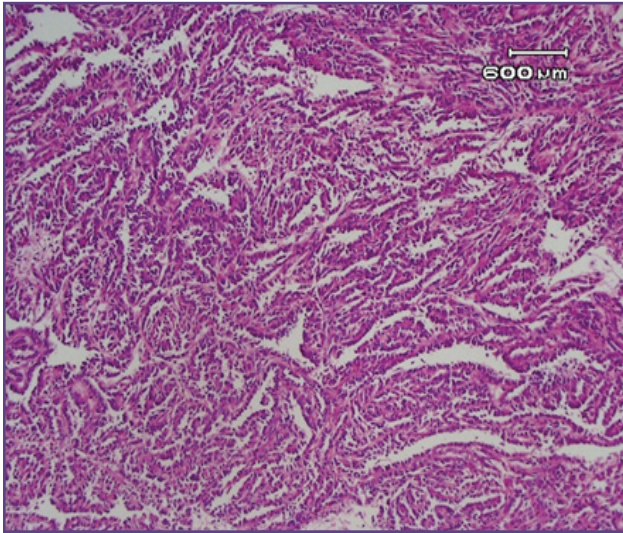


Fig. 2: Dimorphic tumour with tubules and spindle cells with myxoid stroma (H&E, 10x)

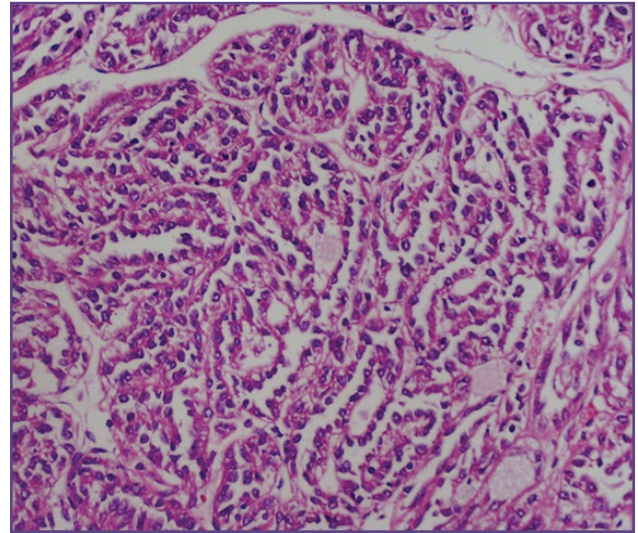


Fig. 3: Tubules lined by cuboidal cells (H&E, 40 x)

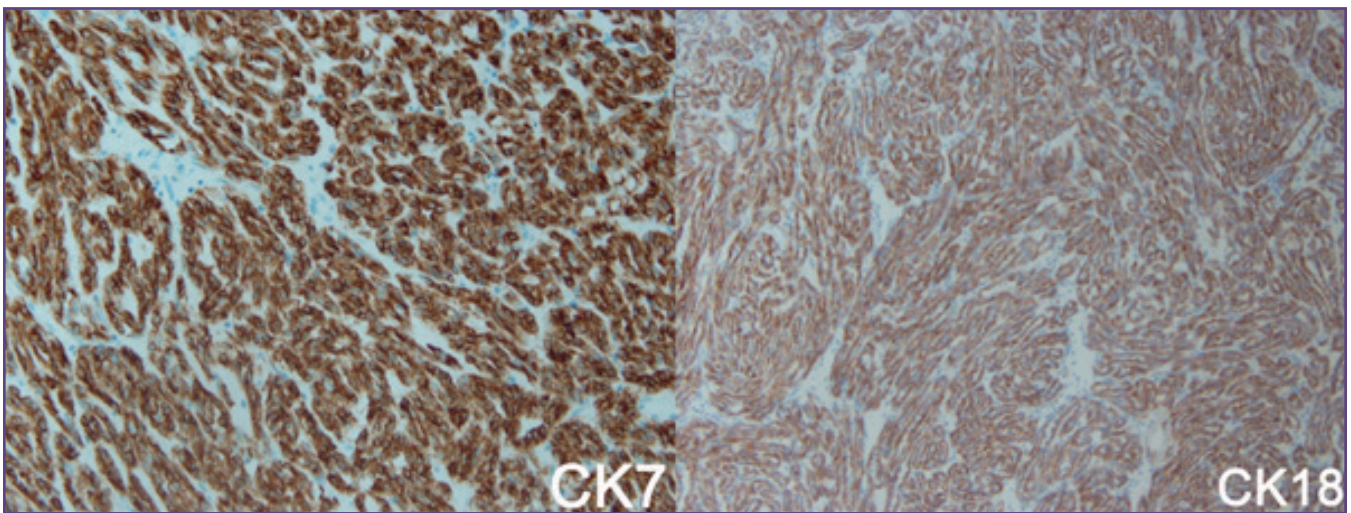


Fig. 4: IHC CK 7 and CK 18 positive epithelial and spindle cells.

incidentally picked up on routine physical examination or during imaging studies. CT imaging shows MTSCC and RCC with slightly different ranges of attenuation values on unenhanced scan. MRI shows similar signal intensity of both the tumour and renal parenchyma on T1-weighted imaging and heterogenous on T2-weighted imaging. FNAB (fine needle aspiration biopsy) may be diagnostic of MTSCC. In such cases the preoperative diagnosis aids in the case management with limited nephrectomy. Our case showed a heterogeneously enhancing lesion with internal hypodense areas and involvement of the renal pelvis (in contrast to the post nephrectomy where renal pelvis was free of tumour). The huge size of the tumour and radiologically involvement of the renal pelvis

probably prompted for a total nephrectomy. The tumour is well circumscribed ranging from 1.0 cm to 18.0 cm in diameter and is usually confined to the kidney.^[1,5] Our case measured 16.5 cm in its greatest dimension. Cut surface shows a grey to pale yellow areas often involving the renal medulla.^[5] Histology shows a low grade tumour composed of elongated tubules or cordlike growth pattern with branching and freely anastomosing, lined by cuboidal cells with eosinophilic focally vacuolated cytoplasm separated by pale mucinous stroma and spindle cell areas.^[2,5] Fine et al classified them as classic with abundant extracellular mucin stroma and mucin poor with little or no extracellular mucin.^[3] Tumours may show focal papillary growth pattern, necrosis, ectopic bone and neuroendocrine differentiation

including carcinoid tumour and small cell neuroendocrine carcinoma.^[2,5] Differential diagnosis includes collecting duct carcinoma, papillary RCC solid variant, sarcomatoid RCC, metanephric adenoma and carcinoid. All the above conditions lack a mucinous stroma. Both RCCs show classical areas of RCC whereas collecting duct carcinoma shows highly atypical tubules with desmoplastic stroma and metanephric adenomas are CK 7 negative unlike MTSCC which are CK 7 positive.^[4] IHC shows CK 7, CK 19, CK 18, EMA, vimentin and P504S(AMACR) positivity.^[5] Histogenesis of the tumour remains debatable and is uncertain. ^[6] AMACR, CK 7, EMA and vimentin positivity favour a distal tubule origin but AMACR is seen in proximal convoluted tubules as well.^[6] Low cuboidal cells, elongated tubular glands and myxoid stroma are reminiscent of loop of Henle.^[7] Molecular studies show multiple chromosome losses(1,4,6,8,9,13,14,15 and 22)^[2,3] Two cases with neuroendocrine differentiation have been reported.^[7] Described as a low grade tumour, histologically is consistent with low grade unless sarcomatoid pattern is noted.^[4] Staging is same as RCC. The tumour is always benign and extremely rare to present as metastasis to lymph nodes and other organs. As a subtype of RCC careful follow-up is recommended.

Conclusion

MTSCC is a low grade tumour, a new entity recently included in the WHO classification with good prognosis. One needs to be aware of its classical histological picture and limited nephrectomy can be considered if preoperative diagnosis is available. A massive renal mass excised with a diagnosis of RCC/TCC surprisingly showed a lesion with a new histomorphology and good prognosis favourable to the patient.

Declarations

Funds:

None

Competing interests

None declared

Referances

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