CASE REPORT & LITERATURE REVIEW: MUCINOUS TUBULAR & SPINDLE CELL CARCINOMA

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INTRODUCTION:

Mucinous tubular and spindle cell carcinoma (MTSCC) is histologically distinctive low grade malignant neoplasm first described in 2002 while incorporated in WHO classification of renal neoplasms in 2004. Grossly it ranges in size from 2 to 10cm, is well circumscribed and usually off-whit to yellow. It is composed of variable components of tubular architecture, extracellular mucinous material and spindle cell areas. It was previously thought to be a variant of papillary renal cell carcinoma. Few cases have been reported so far with female predominance. Loss of chromosomes 1, 4, 6, 8, 9, 13, 14, 15 and 22 while gains of chromosomes 3, 7, 16 and 17 are characteristic of this tumor. Rare cases show metastatic disease. However, follow up data of all cases is not available.

CASE PRESENTATION:

A 50 years old female patient presented with flank pain and hematuria. Clinical workup and CT scan abdomen was performed, which revealed a solid mass at upper pole of left kidney. Left radical nephrectomy was performed.

Figure 1: Grossly tumor had multinodular, grey-white cut surface

Grossly 9.5cm well circumscribed, multinodular, encapsulated off-white, soft, friable tumor was identified at upper pole of left kidney involving the cortex. Tumor was 9.5cm away from ureteric resection margin. Histologically, tumor was arranged in nodules separated by fibrous septa. There were elongated interconnected tubules containing slit like lumina, solid compressed cords and papillary structures. The tubules and papillary structures were lined by cuboidal tumor cells, with indistinct cell borders, having round to oval bland nuclei, inconspicuous nucleoli and eosinophilic cytoplasm. Additional sections were taken which revealed; focal clear cells, foamy macrophages, myxoid stroma and few psammoma bodies. Mitotic activity was very low while necrosis was absent. Adrenal gland was free of tumor.

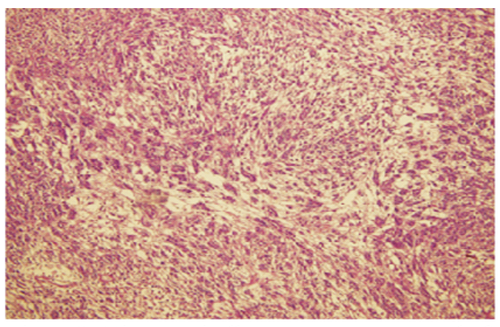
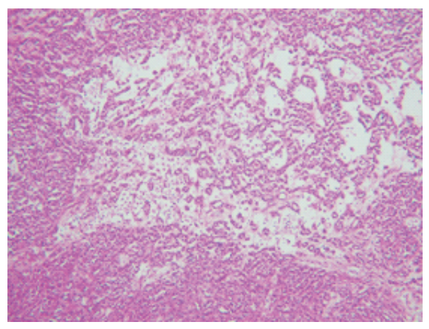
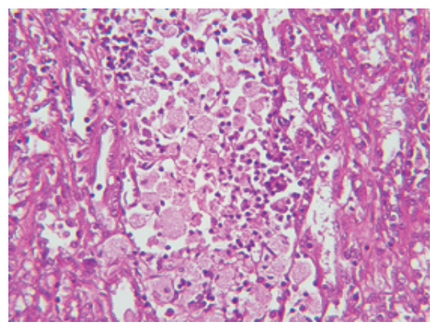


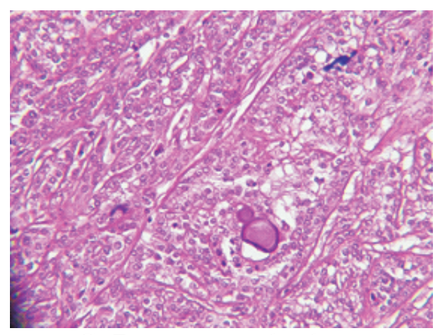
Figure 2: Microscopically, tumor was composed of compressed tubules, surrounded by myxoid stroma



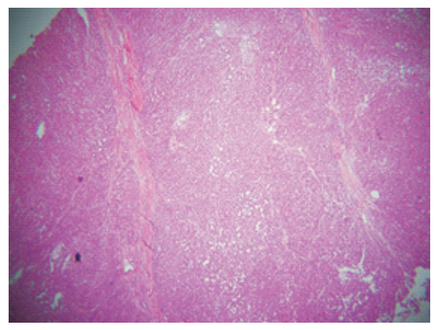
A. Myxoid stroma



B. Foamy macrophages



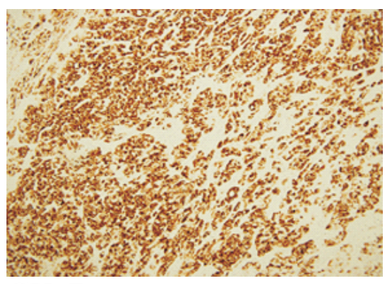
C. Psammoma body



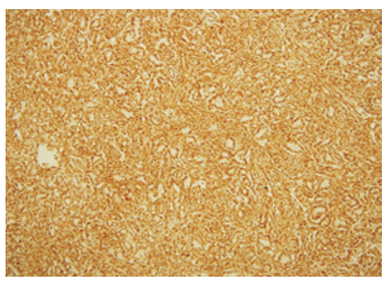
D. Fibrous septa

Figure 3: Different variations in the tumor

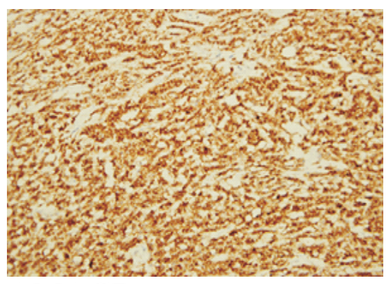
Differential diagnosis included MTSCC, papillary renal cell carcinoma and clear cell papillary renal cell carcinoma.
Immunohistochemically, tumor was positive for vimentin, CK-7, AMACR and CD-15. CD-10 was weak focal positive. Alcian blue highlighted the myxoid stroma.



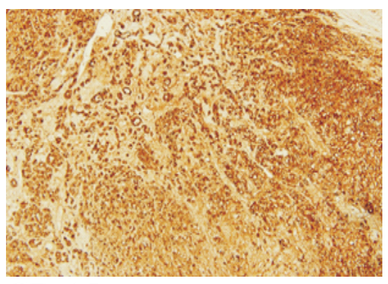
CK-7



Vimentin



AMACR



CD-15

Figure 4: Immunohistochemically tumor was positive for CK-7, Vimentin, AMACR and CD-15.

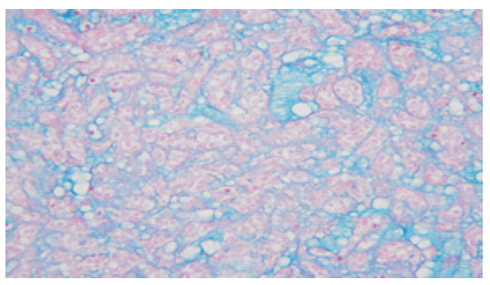


Figure 5: Alcian blue highlighting the myxoid stroma

The immunohistochemical and histochemical stains confirmed the diagnosis of MTSCC. AMACR positivity rules out clear cell papillary renal cell carcinoma while presence of myxoid stroma highlighted by alcian blue ruled out conventional papillary renal cell carcinoma. The tumor was staged as pT2pNxpMx with Furhman's nuclear grade II.

DISCUSSION:

MTSCC is a rare distinct subtype of RCC thought to have proximal tubular differentiation. It is a low grade malignancy incorporated in WHO classification of renal neoplasms in 2004. It occurs more commonly in females with female/male ratio of 3:1. Most of them are discovered incidentally during imaging studies done for other reasons like anemia or occult blood loss in urine. As in our case, they can also present with flank pain and hematuria.
Microscopically, the tumor is usually composed of compressed tubules lined by cuboidal cells with bland nuclear features and eosinophilic often vacuolated cytoplasm. Surrounding stroma is myxoid with pools of extracellular mucin. Variations including papillary change, clear cell change, oncocytic change, sarcomatoid change, ectopic bone formation and neuroendocrine differentiation have also been reported in literature. Some tumors are mucin poor and predominantly show compressed tubules lined by tumor cells. Our case was mucin rich with scattered foamy macrophages and oncocytic cells. No metaplastic change or neuroendocrine change was seen.
Important differential diagnosis includes papillary RCC with sarcomatoid change, mesenchymal tumors, clear cell papillary RCC, metanephric adenoma and Xp11.2 translocation carcinoma.
Immunohistochemically, MTSCC is positive for AMACR, CK-7, EMA, E-cadherin and vimentin. CD-15 positivity can also be seen. Tumor is usually negative for CD-10.Immunostains on our case showed strong staining for vimentin, CK-7, CD-15 and AMACR as validated by literature while alcian blue highlighted myxoid stroma. However, CD-10 also showed focal positivity which is rarely reported. Strong staining for AMACR ruled out clear cell papillary renal cell carcinoma. However, overall staining pattern further solidifies the present belief of MTSCC having proximal tubular differentiation.
At cytogenetic level loss of chromosomes 1, 4, 6, 8, 9, 13, 14, 15 and 22 while gain of chromosomes 3, 7, 16 and 17 is noted. The typical mutation in 3p, VHL mutation found in papillary RCC is not found in MTSCC.
It is important to differentiate this entity from papillary RCC with sarcomatoid features and mesenchymal tumor because MTSCC has favorable prognosis while other two have poor prognosis. Rare cases of MTSCC are known to metastasize. However data is limited to determine long term prognosis.

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