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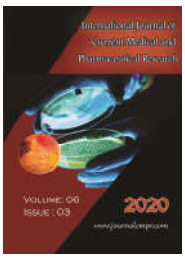

Research Article

MUCINOUS TUBULAR AND SPINDLE CELL CARCINOMA OF KIDNEY :A CASE SERIES

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ABSTRACT

Mucinous tubular and spindle cell carcinomas (MTSRCC) of kidney is rare , low grade polymorphic tumor . Recent studies have described a wide morphological spectrum for this tumor. Prior to this these tumors were diagnosed as variants of solid papillary carcinomas with compressed and elongated papillae, metanephric adenomas, and sarcomatoid carcinomas. Exact cell of origin is not well defined, evidences suggests that they arise from loop of henle or collecting duct epithelium. We report our clinical, histological, and immunohistochemical experiences with 3 such cases.

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INTRODUCTION

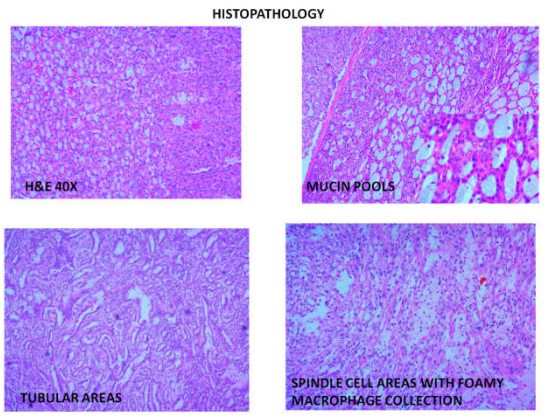
Mucinous tubular and spindle cell carcinoma of kidney is a rare polymorphic low grade renal tumor. It was first described as a distinct entity in the last decade and fewer than 100 cases have been reported in literature ever since [(1).] These showcase a variety of morphological variations as well as the immunohistochemical and molecular aspects of this rare tumor. Here, we report three cases of this enigmatic tumor which we have encountered in our practice with a brief review of literature.

Case 1: 63 year old male was incidentally detected to have a left renal mass when being evaluated for right lower abdominal pain possibly due to appendicitis. Contrast CT scan of abdomen showed 9x 6cm enhancing lower polar mass in left kidney for which left laparoscopic radical nephrectomy was done. Gross examination revealed a well circumscribed and capsulated 9cmx8cm lesion with grey white to yellowish cut surface.

Case 2: 71 year old male presented with a mass involving inferior pole of right kidney which was heterogenous, well defined and exophytic measuring 4.2x3.4cms. He had a history of left radical nephrectomy for renal cell carcinoma, at another centre, 5 years prior to this. At that time, the right renal lesion was 2cms in size for which he was being followed up. He underwent partial nephrectomy and macroscopic examination revealed a 4cmx4cm grey white encapsulated tumor.

Case 3: 58 year old male presented with a left renal mass incidentally detected during routine health checkup. Contrast enhanced CT scan of abdomen showed a 2.3x2.6cm well defined enhancing mass in the lower pole of left kidney for which robotic partial nephrectomy was done. Gross examination revealed a 2.5x2 cm vaguely circumscribed grey white lesion. Microscopy of all three cases revealed tubules, cords and papillae composed of low grade cuboidal cells along with spindle cells lying within mucinous stroma which stained with alcian blue. 2 cases were staged as pT1a while one as pT2a. None of the cases showed lymphovascular invasion or extension into perinephric fat/ renal sinus fat. On immunohistochemical studies, tumor cells revealed positivity for CK7, CK19,AMACR and EMA while they were negative for p63,WT1and CD10.

On follow up, all the three patients have no fresh complaints related to the tumor.



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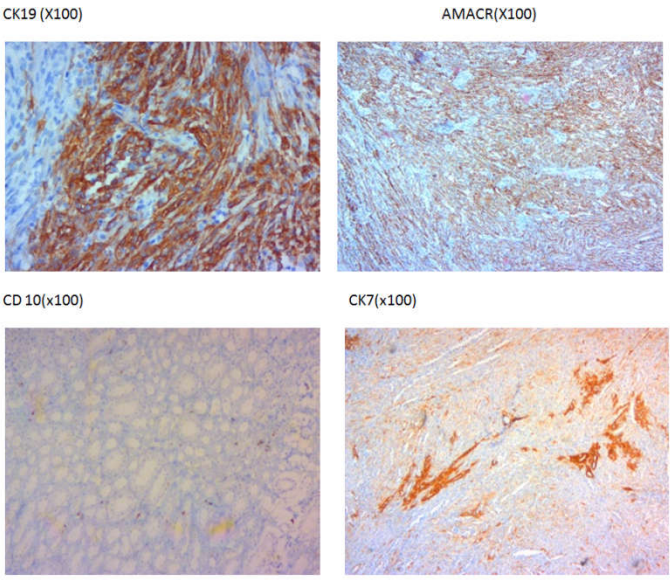
Immunohistochemistry



DISCUSSION

Mucinous tubular and spindle cell carcinoma of the kidney is a rare subtype of renal cell carcinoma (RCC) which was first recognized as a distinct entity in the 2004 World Health Organization (WHO) tumor classification. [(2)] As suggested by its name, classical histomorphology of this tumor comprises mucinous/myxoid stroma, bland spindle cells and tubules. Although tumors with similar morphology were recognised earlier, they were referred to by different names. It was Parwani *et al* who first reported MTSRCC as a distinct low-grade renal tumor with myxoid appearance and distal nephron differentiation in their series of 4 cases. [(3)]

Mucinous tubular and spindle cell carcinomas(MTSRCC) comprise less than 1% of all renal tumors with a mean patient age of 58 years and female preponderance, unlike our exclusively male cohort.[(4)] The classical presentation of RCC with the triad of flank pain, abdominal mass and hematuria is often lacking and majority of these tumors are incidentally detected during abdominal imaging for other reasons as in two of our cases[(5)] They are typically visualised as well-demarcated, exophytic and expansile renal masses with a hypovascular pattern on contrast enhanced studies mimicking papillary RCC .[(6)] Hes *et al* proposed an association of this tumor with renal calculi which we noted in one of our cases.(7)An association with synchronous conventional RCC and renal cysts has also been described by Hes *et al* and Ferlicot *et al*.[(7,8)]

On gross examination, MTSRCCs are usually well circumscribed and partially encapsulated solid masses centred in the cortex with a homogenous cut surface unlike clear cell RCCs. Size can range from less than a cm to more than 18 cm in diameter.(6,9) Classically, this tumor shows varying proportions of tubular and spindle cell components within mucinous stroma. Fine *et al* identified unusual features including papillations, focal clear cells, necrosis, oncocytic tubules, foamy macrophages, heterotopic bone, psammomatous calcification and lymphocyte cuffing thereby mimicking other histologic subtypes of RCC.(10)

On immunohistochemical studies, the tubular and spindlecells express markers of proximal and distal tubular differentiation. They are consistently positive for paired box transcription factor 2 (PAX2) andPAX8, epithelial membrane antigen (EMA), alphamethylacyl-CoA racemase (AMACR), low molecular weight cytokeratins (CK8/18, CK19 and CK7) and Ecadherin and variably express vimentin and high molecular weight cytokeratin (34BE12) . CD10, CD15 and RCC marker which are positive in clear cell carcinomas are often negative as are ulex europaeus agglutinin (UEA-1), P63, CK20, carbonic anhydrase IX and GATA3.(6,9,11) Immunohistochemical evidence of neuroendocrine differentation has also been reported in some of these tumors.

In the presence of areas with unusual architecture, high-grade spindly cells, extensive necrosis,high proliferation index and AMACR negativity, the possibility of sarcomatoid change in MTSRCC must be considered and these predict an aggressive clinical course.(12)

Though often thought to arise from the tubular epithelium, Banyai *et al* proposed that MTSRCC develops from embryonal rest–like precursor lesions with impaired differentiation.(13) Peckova *et al* worked extensively on molecular genetic abnormalities in MTSRCC and identified chromosomal losses including 1, 4, 6, 8, 9, 13, 14, 15, and 22. Though considerable morphological and immunohistochemical overlap with papillary RCCs has been described, FISH based analyses of these tumors have consistently shown lack of gains of chromosomes 7,17 and loss of chromosome Y characteristic of papillary RCC. (14,6)

Classical MTSRCCs have an indolent course and excellent prognosis post excision with rare recurrences and metastases. Metastasis is more frequent in tumors with atypical histopathological features though there are no published consensus guidelines for systemic treatment in such cases. Hence, a close follow up is warranted in all patients.

CONCLUSION

Mucinous tubular and spindle cell renal carcinoma is a distinct and rare low grade malignant renal tumor with a favourable outcome in majority. However pathologists and clinicians must be aware of the morphological variations in these tumors and their implications so as to arrive at the right diagnosis and to guide patient therapy and follow up.

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