

# Mucinous tubular and spindle cell carcinoma: A difficult diagnosis

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## Abstract

Mucinous Tubular and Spindle Cell Carcinoma (MTSCC) is infrequently seen renal malignancy with favorable outcome, when diagnosed in the early stage. Once out of kidney it is have lethal course. Radiologically MTSCC is heterogeneously hypo-enhancing renal mass in delayed phase, may mimic to papillary renal cell carcinoma and in histopathology showed these tumoral cells shows complex immunophenotype expression. As this does not show enhancement pattern of common renal cell carcinomas, radiological diagnosis may be challenging. Histopathological reporting is also tedious indeed necessary to reach definitive diagnosis, which help in tailoring follow up and prognosis. Metastatic disease has dismal outcome and responds poorly with adjuvant therapy and patient succumbs within short span of time.

**Keywords:** Carcinoma, mucinous, papillary, renal, sarcomatoid

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## INTRODUCTION

Renal cell carcinoma (RCC) is the most lethal of common urological cancers. Clear cell variant is the most common type of RCC described. Mucinous tubular and spindle cell carcinoma (MTSCC) is a rare variant of RCC, described in fewer than 100 cases in the literature. Due to its rarity, immunohistochemistry is used as an adjunct to diagnosis and to rule out close mimickers, notably sarcomatoid papillary Type 1 RCC and low-grade collecting duct carcinoma. In organ-confined MTSCC, complete removal usually confers a good prognosis. We describe a patient with renal mass whose postoperative histology showed MTSCC of the kidney. Its management and the literature on the subject are subsequently discussed.

## CASE REPORT

A 65-year-old female presented with occasional mild pain in the right upper quadrant of the abdomen for the past 6 months. Pain was nonradiating and not associated with any specific aggravating or relieving factors. She used to have two to three such episodes every month. There was no history of flank pain, hematuria, lower urinary tract symptoms, fever, weight loss, bone pains, or loss of appetite. General physical examination was unremarkable. Abdominal examination revealed a soft, ill-defined smooth, firm lump in the right hypochondriac region extending to the right lumbar region inferiorly and epigastric region medially. Mass was not bimanually palpable but was ballotable. It was not possible to get above the swelling. Ultrasound of

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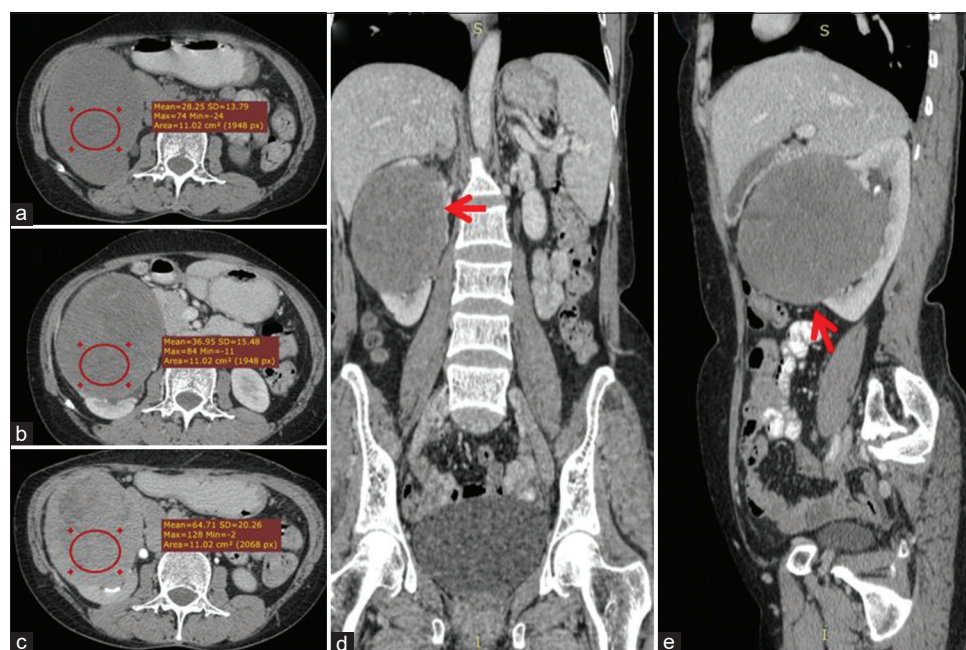
the abdomen revealed a large heterogeneous hyperechoic space-occupying lesion arising from the upper and midpole of the right kidney. A contrast-enhanced computed tomography scan of the abdomen was done, which revealed a large heterogeneously enhancing mass measuring 12 cm × 10 cm arising from the upper and midpole of the right kidney, with the involvement of the renal sinus. There was no tumor thrombus in renal vein [Figure 1]. Nearby structures were not invaded. A metastatic workup (liver function tests, serum alkaline phosphatase, and chest X-ray) was done, which was within normal limits. The patient was counseled and taken up for open right radical nephrectomy. Intraoperatively, the mass was closely abutting the liver, from which it was carefully dissected off. The renal artery was doubly ligated and divided, followed by the renal vein in the same fashion. The radical nephrectomy specimen was delivered out and sent for histopathology. The postoperative course was uneventful, and the patient was discharged 4 days after the operation.

On gross inspection, the radical nephrectomy specimen with an intact capsule measured 14 cm × 11 cm × 6 cm. The cut surface of the lesion showed variegated rounded growth with hemorrhagic pale yellow and necrotic areas. On microscopy, a thickened fibrocollagenous capsule was seen, which was uninvolved by tumor cells. Cuboidal cells were seen arranged in long cords and tubules and making abrupt transitions to spindle cell morphology at focal places. These epithelial structures were arrayed against the background of lightly basophilic

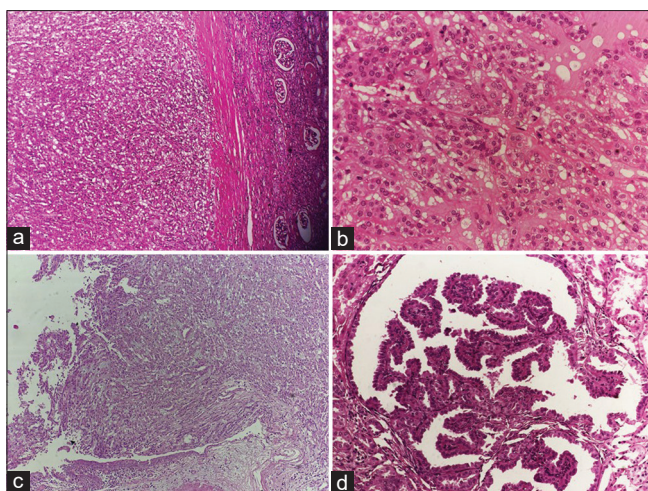
mucinous or myxoid material. The nuclei were spherical or oval with fine chromatin and small nucleoli. Mitotic figures were uncommon. The mucinous background contained mast cells, clusters of foamy histiocytes, and cholesterol clefts. On immunohistochemistry, the tumor cells were positive for CK7, negative for p63, and Alpha-methylacyl-CoA racemase (AMACR) was noncontributory [Figures 2 and 3]. Features were consistent with a diagnosis of MTSCC arising from the upper pole of the right kidney.

## DISCUSSION

MTSCC is an unusual variant of RCC, which shows a biphasic pattern of tubular and spindle epithelial cells in a background of mucinous stroma. It was first recognized as a distinct RCC entity in the 2004 classification of renal tumors.<sup>[1]</sup> It occurs more commonly in females and demonstrates multiple chromosomal losses involving 1, 4q, 6, 8p, 11q, 13, 14, and 15 and gains involving 11q, 16q, 17, and 20q. Grossly, MTSCC is partially encapsulated and well circumscribed. The tumor is characterized by a mixture of tubular and spindle cell components separated by variable amounts of mucin. Alcian blue stain can highlight the scant mucin in the tumor. These tumor cells express complex immunophenotype epithelial markers, CK19, CK7, AE1/AE3, and distal renal tubule markers such as epithelial membrane antigen, CK19, and E-cadherin.<sup>[2,3]</sup> Literature suggests that MTSCC may represent a variant of papillary RCC or a heterogeneous tumor, but genetic studies have proved that it is a distinctive entity.<sup>[4,5]</sup>



**Figure 1:** Computed tomography attenuation of right renal mass on axial (a) noncontrast (28 HU), (b) corticomedullary phase (36 HU), (c) delayed (10 min) phases (64 HU) shows slow but progressive enhancement of the mass. (d) Coronal, (e) sagittal reformats (arrows) show a large well-encapsulated mass in the right kidney arising from the anterior cortex and involving the renal sinus



**Figure 2:** (a) Encapsulated, hypercellular tumor with normal kidney (H and E,  $\times 10$ ). (b) Tumor cells arranged in tubules and cord along with the abrupt transition to bland spindle cells (H and E,  $\times 20$ ). (c) Tumor cells filling the renal pelvis (H and E,  $\times 20$ ). (d) Papillary adenoma. (H and E,  $\times 20$ )

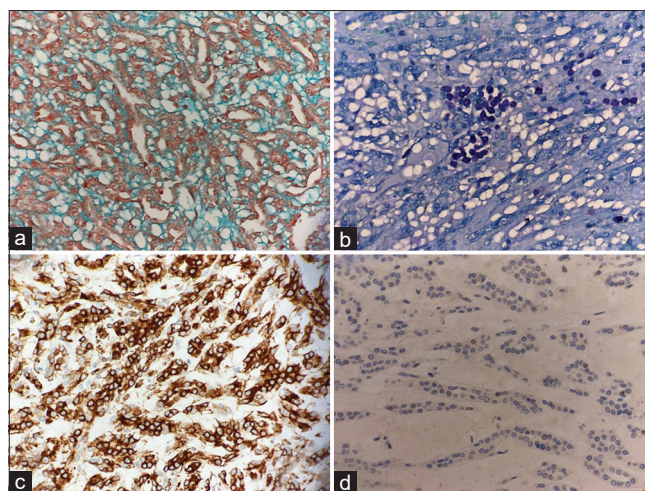
Comparative genomic hybridization and fluorescence *in situ* hybridization have emerged to differentiate it from papillary RCC, which does not show extracellular mucin but has specific cytogenetic alterations. Papillary RCC with sarcomatoid differentiation shows atypical, large, and polygonal cells, whereas MTSCC shows low-grade nuclei. Sarcomatoid RCCs may show uniform tumoral spindle cells but always with areas of considerable nuclear pleomorphism and mitotic activity. Collecting duct carcinomas (papillary variant) show large, eosinophilic tumor cells, with important cytonuclear atypia in the tubular component.<sup>[6-8]</sup> In distinction to the more aggressive sarcomatoid papillary RCC, MSTCC if confined to the kidney and removed entirely has a good prognosis.<sup>[2,6]</sup> In a cohort of 28 patients, the 3-year overall survival was 84.8%. However, for metastatic MTSCC, time to treatment failure was  $<6$  months with the exception of one patient who achieved long-lived response with sunitinib.<sup>[2]</sup>

## CONCLUSION

MTSCC is a rare variant of RCC. Histomorphological features are the gold standard for making a diagnosis and differentiating from papillary RCC with sarcomatoid features, with which it shares histological similarities. Immunohistochemistry may be used as an adjunct to diagnosis. If confined to the kidney, complete removal confers a good prognosis. In metastatic disease, treatment with tyrosine kinase inhibitors has been tried, but the overall efficacy is unproven.

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient (s) has/have given his/her/their consent for his/her/their images and



**Figure 3:** (a) Alcian blue highlighting the mucinous stroma ( $\times 40$ ). (b) Toluidine stain highlighting the mast cells ( $\times 40$ ). (c) Cytokeratin 7 immunoreactivity in tumor cells (IHC  $\times 40$ ). (d) P63 is negative in the tumor cells (IHC  $\times 40$ )

other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

## Conflicts of interest

There are no conflicts of interest.

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