

## Mucinous tubular and spindle cell carcinoma kidney with sarcomatoid and rhabdoid differentiation- A rare entity

Fadiya Zainudeen<sup>1</sup>, Umasankar .P<sup>2</sup>, Lakshmi Priya .U<sup>3,\*</sup>, Jayalakshmy P S<sup>4</sup>, Asiq Sideeque<sup>5</sup>

<sup>1,2,3</sup>Assistant Professor, <sup>4</sup>Professor, <sup>5</sup>Professor and HOD, Dept. of Pathology, MES Medical College, Palachode P.O, Perinthalmanna, Kerala, India

**\*Corresponding Author:**

Email: lakshmipriya@gmail.com

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### Abstract

Mucinous tubular and spindle cell neoplasm is a rare renal tumor with favourable outcome. Sarcomatoid change is described in many renal cell carcinomas, indicative of a worse clinical course. We report a case of mucinous tubular and spindle cell neoplasm with areas having sarcomatoid and rhabdoid morphology with large areas of necrosis. Sarcomatoid change is very rare in mucinous tubular and spindle cell tumor and we could find only 4 similar cases in the published literature.

**Keywords:** Mucinous tubular and spindle cell, Sarcomatoid, Rhabdoid.

### Introduction

Mucinous tubular and spindle cell neoplasm is a rare renal tumor included in 2004 WHO classification of renal tumors.<sup>1</sup> It was first described as an entity by Mac Lennan et al in 1997.<sup>2</sup> They described the lesion as “low-grade mucinous tubulocystic renal carcinoma” in a series of 13 cases, morphologically different from all other renal tumors known at that time. They also proposed that the tumor is of collecting duct origin. The tumor is characterized by histological features such as tubules and cords of composed of low grade cuboidal cells, mucinous extracellular stroma and low grade spindle cell areas. The neoplasm is generally considered as low grade with good prognosis. However rare cases of metastatic MTSCC have been reported.<sup>3,4</sup> Cases showing neuroendocrine differentiation are also described.<sup>5,6</sup> There are also reports of mucin poor variant, diagnosis of which causes considerable diagnostic challenge for histopathologist.<sup>7</sup>

Sarcomatoid transformation is well documented and can be seen in about 8% of common histological types of renal cell carcinomas.<sup>8</sup> Sarcomatoid morphology is usually associated with poor prognosis. High grade spindle cell areas in MTSCC are extremely rare.

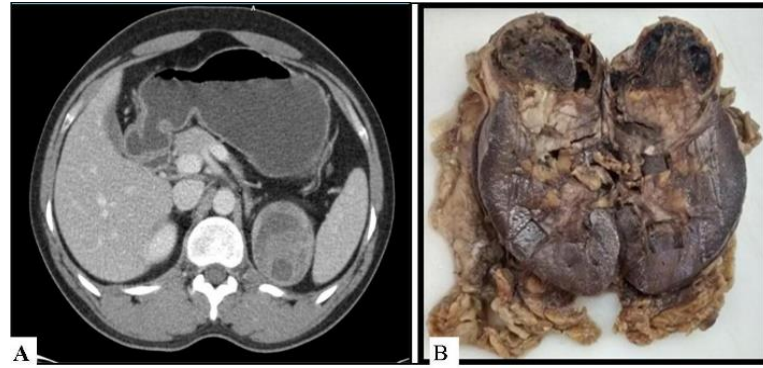
### Case Report

45 year old male presented with history of hematuria for two months. Ultrasound and CT scan revealed an upper pole lesion involving the left kidney (Fig. 1A). Left radical nephrectomy was done. Cut section revealed a well defined neoplasm involving the upper pole (Fig. 1B). Tumor had a variegated appearance with areas of necrosis, hemorrhage and cystic change. Tumor was seen infiltrating into renal

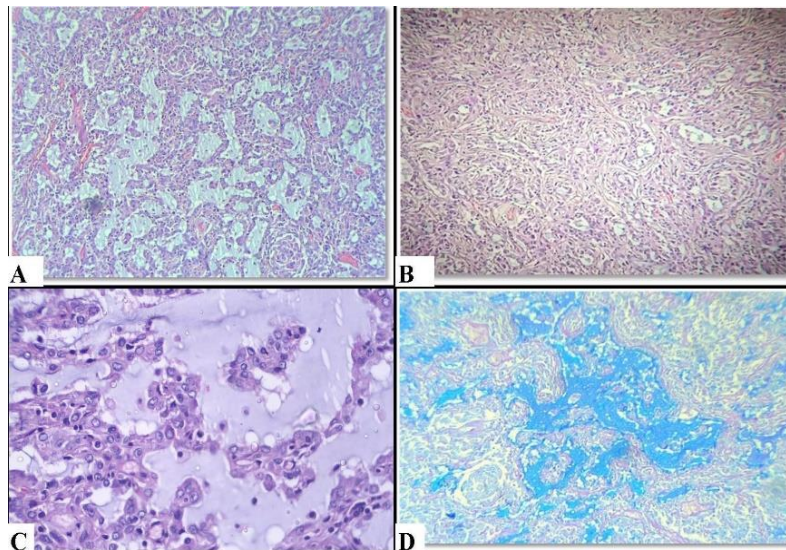
sinuses. Perinephric fat and renal vessels were grossly free of neoplasm. No lymph nodes were identified grossly.

Microscopic examination in H&E sections had the characteristic tubules, interlacing cords, trabeculae and papillae of low grade cuboidal cells with extracellular mucinous material in between (Fig. 2 A, C). Mucin was brightly stained using alcian blue (Fig. 2D). Areas with low grade spindle morphology was seen in between (Fig. 2B). There were also mucin poor areas. Also seen were areas with spindle cell morphology having marked atypia (Fig. 3A), necrosis (Fig. 3B) and frequent mitosis. Rhabdoid morphology was seen in areas (Fig. 3C). Vessels walls had prominent hyalinization (Fig. 3D). In areas cells had plasmacytoid morphology, arranged as whorls, resembling ‘squamous morules’ (Fig. 3E). Geographic necrosis with palisading of tumor cells surrounding the vessels were noticed in areas. Bizarre cells and multinucleate tumor giant cells were present. (Fig. 3A inset). Areas also had collections of foamy macrophages and cholesterol clefts. There were areas with sclerosis and calcification. In areas tubules were seen forming complex architecture resembling a glomerulus, surrounded by an empty space (Fig. 3F). There were areas with prominent lymphoplasmacytic infiltrate, in between the tumor cells and in areas perivascular.

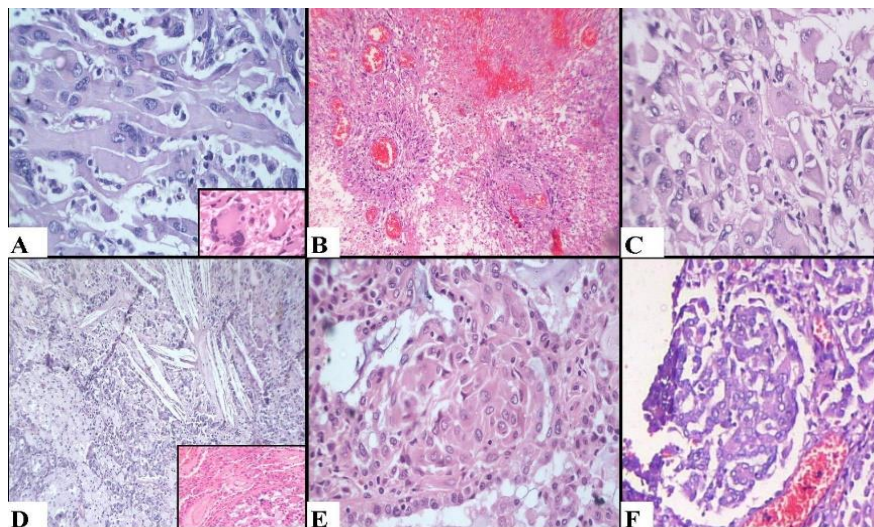
Immunohistochemistry was done (Fig. 4). Tumor cells were positive for cytokeratin 7, cytokeratin 19 and AMACR (alpha methyl racemase coenzyme A). Vimentin showed bright positivity in sarcomatoid areas and was negative in low grade areas. Tumor cells were negative for CD10. Desmin was done on sections with rhabdoid morphology and was negative.



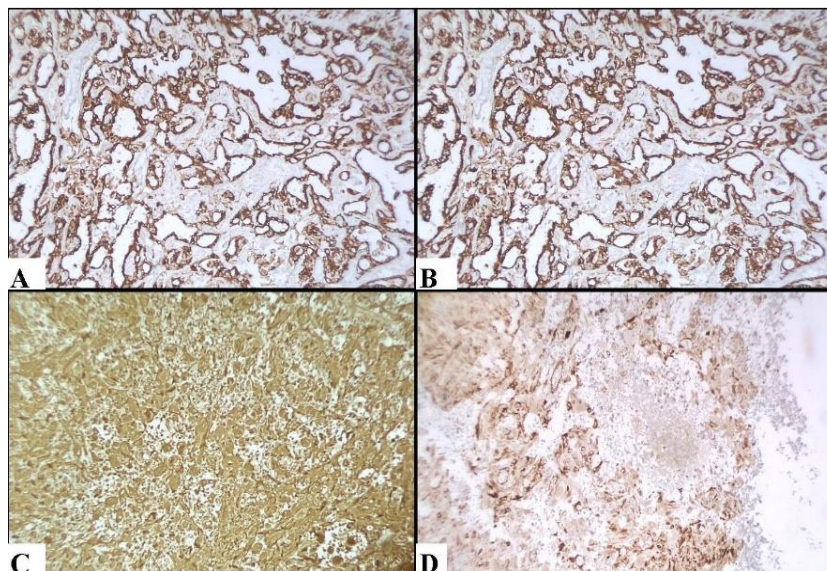
**Fig. 1:** A) CT scan with an upper pole lesion involving the left kidney; B) Nephrectomy showed well defined variegated neoplasm involving the upper pole



**Fig. 2:** A) Tumour cells arranged in cords with extracellular mucin in between(H&E 100x); B) Areas with low grade spindle cell morphology(H&E 100x); C) Tumour cells and mucin in high power(H&E 400x); D) Alcian blue stain highlighted the mucin



**Fig. 3:** A) Tumour cells with high grade spindle cell morphology(H&E 400x); B) Areas of necrosis(H&E 100x); C) Tumour cells with rhabdoid morphology(H&E 400x); D) Areas with collections of foamy macrophages and cholesterol clefts and hyalinised vessels in inset(H&E 100x); E) Squamoid morule formation(H&E 400x); F) Tubules forming glomeruloid tufts(H&E 400x)



**Fig. 4:** A) Tumour cells showed strong positivity for CK 7; B) Tumour cells showed strong positivity for CK 19; C) Sarcomatoid areas positive for vimentin; D) Tumour cells positive for AMACR

### Discussion

Since first described in case series in 1997 less than 100 cases of MTSCC could be identified in the published literature.<sup>2</sup> Sarcomatoid transformation in MTSCC is extremely rare and was first described by Simon R.A et al in 2008.<sup>9</sup> We could trace only four cases of sarcomatoid transformation in the published literature till date. Three out of these four case reports described aggressive courses with distant metastasis and fatal outcomes.<sup>9-12</sup> Hence high grade sarcomatoid morphological features in MTSCC indicates poor prognosis.

MTSCC can show mucin poor areas as seen in this case. The mucin poor variant can cause considerable diagnostic challenge for the reporting pathologist.<sup>7,14</sup> 'Glomeruloid' structures formed by complex tubules occupying an empty space seen in this case was also observed in a previous case report of MTSCC with neuroendocrine differentiation.<sup>15</sup> Presence of foamy macrophages and prominent lymphoplasmacytic infiltrate were also described in published literature.<sup>13</sup> However we could not identify any case reports of MTCC with tumor cells having rhabdoid or plasmacytoid morphology.

Low grade areas were positive for CK 7, CK 19, AMACR and was negative for CD10. Vimentin was expressed in sarcomatoid areas only. These observations were consistent with that of previous observations.<sup>10,13</sup> There is no consensus regarding the ontogeny of MTSCC. Expression of AMACR, CK7 & CK 19 suggests proximal tubular origin, similar to papillary renal cell carcinoma. Considering the overlap in the morphological and immunohistochemical features of both lesions, some researchers have a view that MTSCC is a variant of papillary RCC.<sup>16,17</sup> However genetic analysis done by some researchers do not

support this theory.<sup>18</sup> Peckova K et al had proven that low grade MTSCC can have multiple genetic alterations such as losses affecting chromosomes 1, 4, 8, 9, 14, 15, and 22. The study also proved tumors having morphological overlap with papillary RCC had more variable chromosomal changes including gains of both chromosomes 7 and 17, similar to papillary RCC.<sup>19</sup>

MTSCC is a rare renal neoplasm considered as low grade with favorable outcome. However this popular belief is now shaken by a few reports of metastasis and a few reports of high grade sarcomatoid change. Ursani NA et al and Thway K et al have described distant metastasis in cases with low grade MTSCC morphology.<sup>3,4</sup> Some researchers had successfully attempted tyrosine kinase inhibitors like sunitinib in such cases.<sup>20</sup> There are also reports of MTSCC with neuroendocrine differentiation.<sup>5</sup> Larger worldwide studies are needed to understand more about the biological potential of MTSCC.

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