A rare case of mucinous tubular and spindle cell renal cell carcinoma with sarcomatoid change

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ABSTRACT

Mucinous tubular and spindle cell renal cell carcinoma is a rare and recently described subtype of RCC. We present the case of a 45-year-old woman with incidentally detected left renal mass. Imaging revealed a large mass and left radical nephrectomy was performed, histopathology revealed it to be a mucinous tubular and spindle cell carcinoma with sarcomatoid change. She is under close follow-up for the last 6 months.

Key words: *Mucinous, Renal cell carcinoma, Sarcomatoid change, Spindle cell*

Mucinous tubular and spindle renal cell carcinoma (MTSRCC) is a subtype of RCC, which has a polymorphous histology and spindled epithelial cell as an inherent carcinomatous component. The tumor has been recognized as a distinct entity by the World Health Organization tumor classification of 2004 [1]. To date, <100 cases of these tumors have been reported in the literature [2]. Sarcomatoid change in RCC has an overall incidence of 8% and is well documented to occur in the more common subtype of RCC. Although MTSRCC by itself does not have aggressive behavior, there have been reporting cases of MTSRCC with sarcomatoid change having aggressive behavior in the literature search [3].

We present the case of a 45-year-old woman with incidentally detected left renal mass, and later on, histopathology revealed it to be a mucinous tubular and spindle cell carcinoma with sarcomatoid change.

CASE REPORT

A 45-year-old woman presented to our department with a chief complaint of incidentally detected left renal mass. She was a housewife by occupation, and there were no other complaints of hematuria, flank pain, or weight loss. There was no history of fever, hypertension, tuberculosis, and diabetes. On physical examination, it revealed the presence of a mass occupying the left lumbar region and moving with respiration. It was bimanually palpable and ballotable.

All the hematological investigations including renal function tests were normal. Chest x-ray was noncontributory. On ultrasonography (USG), there was a fairly defined mixed echogenic space occupying lesion (SOL), 64×43 mm in the lower aspect of the left kidney. There was another well-defined hyperechoic SOL, about 25×23 mm in diameter in the upper pole of the left kidney, probably an angiomyolipoma (AML). On computerized tomography (CT) scan, there was a large heterogeneously enhancing soft tissue mass of size 45 mm×69 mm×46 mm (enhancement from 39 to 50 H.U.) seen involving lower part of the left kidney. The left renal vein was free from any thrombus. Fascial planes were intact. There was also a mixed density SOL in the upper pole of the left kidney having fat and soft tissue component probably an AML (Fig. 1). Left radical nephrectomy was performed with an uneventful recovery.

On gross pathologic examination, there was tumor of 5.5 cm×3.5 cm×2.5 cm, involving lower pole of the left kidney with variegated cut surface (Fig. 2). The macroscopic extension was limited to the renal parenchyma. Histological features of tumor composed of tubules and cords formed by cuboidal cells with hyperchromatic nuclei, separated by bubbly myxoid stroma, along with an abrupt change into spindle cell morphology (Fig. 3). The spindle cell component was composed of fascicles of highly pleomorphic malignant cells with hyperchromatic nucleus and prominent nucleoli (Fig. 4), with brisk mitotic activity. At some areas, a collection of foamy histiocytes and inflammatory cells including plasma cells were noted. Histologic type was confirmed to be as mucinous tubular and spindle cell carcinoma of the kidney with sarcomatoid change. All the margins were uninvolved including Gerota's fascia, and lymphovascular invasion was absent. Immunohistochemistry (IHC) was done which showed that vimentin was focally positive in tumor cells. The epithelial membrane antigen (EMA), alpha-methylacyl-coenzyme A racemase (AMACR), and cytokeratin 7 (CK7) were also positive (Figs. 5 and 6) in tumor cells. At present, she is in regular follow-up and doing well for 6 months after surgery.

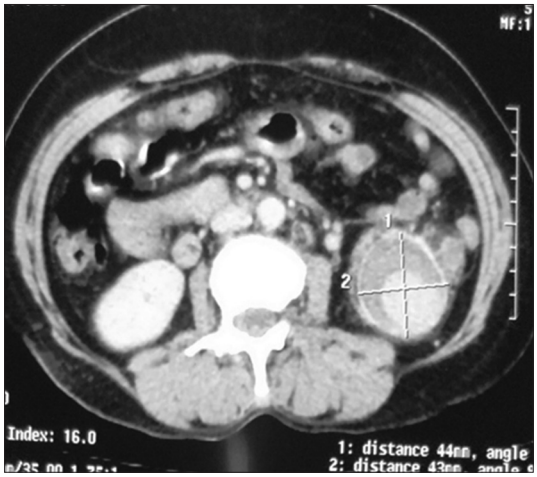

Figure 1: Contrast-enhanced computed tomography showing angiomyolipoma and heterogeneously enhancing mass in the left kidney

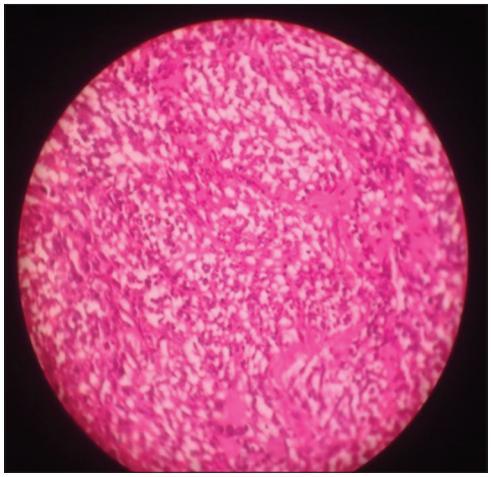

Figure 4: Tumor having spindle cells with hyperchromatic nucleus and prominent nucleoli (40× magnification)

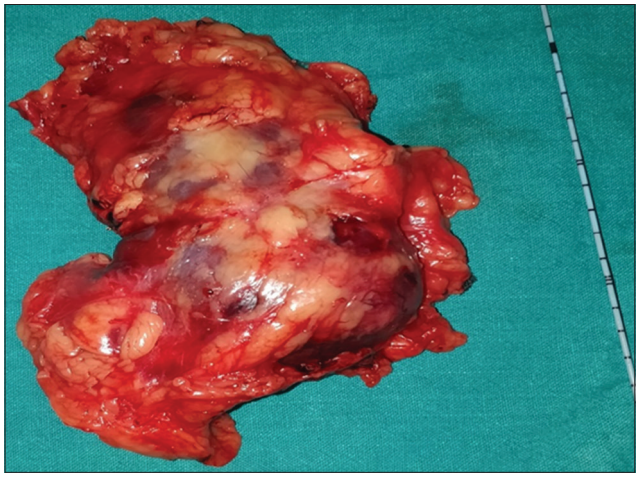

Figure 2: A picture of gross specimen

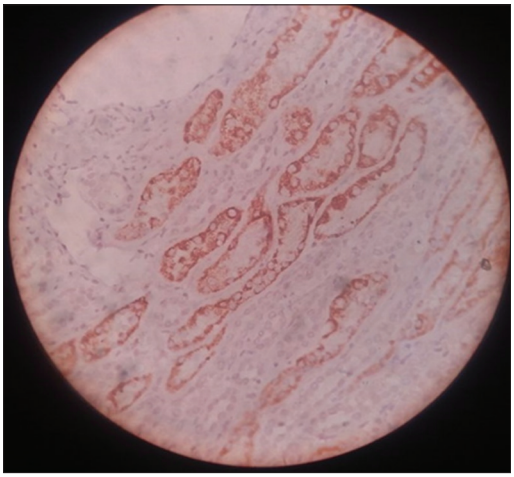

Figure 5: Alpha-methylacyl-coenzyme a racemase positive in tumor cells (40× magnification)

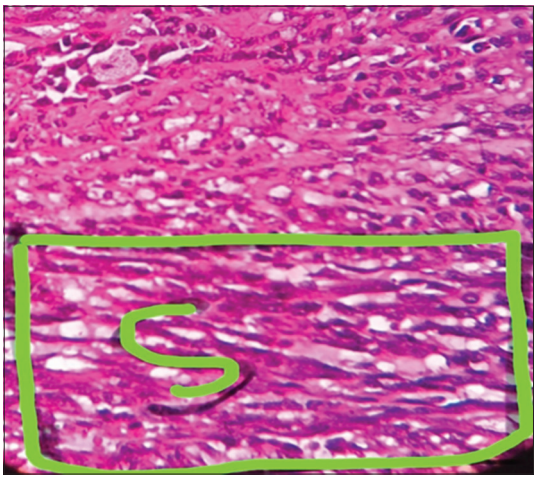

Figure 3: Tumor showing sarcomatoid changes 40× magnification

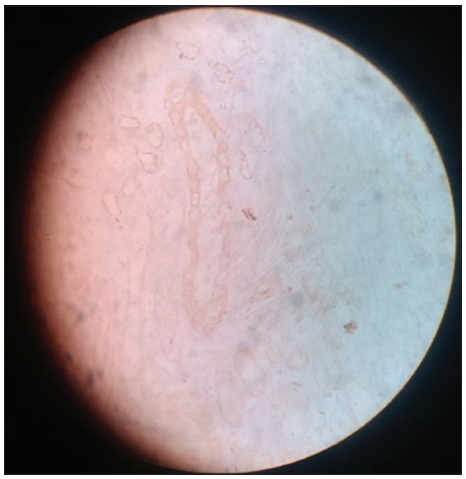

Figure 6: Cytokeratin 7 positive in tumor cells (10× magnification)

DISCUSSION

MTSRCC of the kidney is a rare entity. It was first described in 1998 and previously classified in the category "RCC, unclassified" by He *et al*. [4]. The issue of the origin of MTSRCC is still unsettled. Expression of epithelial markers, namely AMACR, EMA, CK7, and vimentin, has been reported in 80–100% of the cases, thus supporting a possible origin from distal convoluted epithelial cells as in the present case [5].

The tumor is more prevalent among adults with a mean age of 53 years (range: 17–82), with a marked female preponderance (1:4), not seen to be associated with nephrolithiasis. Clinically,

they are generally asymptomatic or present with flank pain or hematuria and are detected on USG. The age, sex, and clinical presentation of the present case were similar to that described in the literature [6].

The CT features of MTSRCC are attenuation values ranging from 31 to 40 HU on an unenhanced scan and attenuation values ranging from 38 to 50 HU on corticomedullary phase which was very much similar in our case [3].

There is no consensus regarding the treatment of mucinous tubular and spindle cell carcinoma of the kidney with sarcomatoid change. It is recommended that although an innocent outcome is likely, a close follow-up is warranted. We followed up this patient with 3 monthly visits with physical examination, laboratory tests, abdominal USG, and a chest X-ray. With regard to the therapy of MTSRCC, patients with localized disease are usually treated with resection, either partial or radical nephrectomy, and in our case, partial nephrectomy was not done as the tumor was more than 4 cm in size. For metastatic diseases, there are no reports of systemic treatment guideline published to date.

With regard to MTSRCC, where spindle cells dominate, the most critical differential diagnosis is sarcomatoid RCC, which can develop in any form of RCC, and usually confers an aggressive behavior. Basal-appearing spindle cells are an inherent defining component of mucinous tubular and spindle cell carcinoma. The presence of these benign-appearing spindle cells does not give a bad prognosis to this carcinoma. Sarcomatoid change in RCC has a worse prognosis and is defined histologically by the presence of spindle cells with nuclear pleomorphism and prominent nucleoli [7]. These cytologic features were observed in our case, but the tumor was not aggressive.

Simon *et al*. [8] found that, in case of sarcomatoid change, the tumor is negative for CK7 and EMA. Furthermore, Dhillon *et al*. [9] found high MIB1 labeling index and negativity for CK7 and AMACR in cases with sarcomatoid change, showing aggressive behavior. In our case, CK7, AMACR, and EMA were positive. Kuroda *et al*. performed comparative genomic hybridization analysis of a case of the high-grade MTSCC that showed a gain of chromosomes 1q, 7, 16, 19q, and Y and loss of chromosomes 1p, 6p, 8p, 11q (del (11) (q23)), and 13. Subsequent G-band karyotyping of the same case showed a gain of chromosomes 2, 5, 7, 12, 16, and 20 and loss of chromosome 15 [10]. Cossu-Rocca *et al*. reported that MTSCC lacks the gain of chromosomes 7 and 17 and loss of chromosome Y as seen in papillary RCC [11].

CONCLUSION

MTSRCC is a rare entity and its association with sarcomatoid change is a rare possibility. Sarcomatoid change is generally associated with aggressive behavior but is not always necessary. Diagnosis requires histopathology, IHC, and cytogenetics. Close follow-up of the entity is essential until a better understanding of the biology of this rare tumor is obtained.

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