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Mucinous Tubular and Spindle Cell Carcinoma, a Low Incidence Renal Tumor: Case Report and Literature Review

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Abstract

Introduction: Renal mucinous tubular and spindle cell carcinomas are uncommon, distinctive renal tumors accounts for less than one percent of renal cell carcinoma. Because of such a rare and important presentation, we report a mucinous tubular and spindle cell carcinoma of kidney which occurred in a 60-year-old woman. **Case Presentation:** A 60-year-old woman presented complaining of vague epigastric pain since 2 months. She did not have any urinary problems such as haematuria or loin pain. Contrast enhanced CT scan of the abdomen and pelvis detected a large left renal mass lesion (11 × 9 cm in maximum dimensions) arising from middle zone of the left kidney. The lesion shows heterogenous contrast enactment with radiologic criteria suggestive of aggressive neoplastic lesion (Renal cell carcinoma). Left radical nephrectomy was done through flank incision. The histopathology examination reported mucinous tubular and spindle cell carcinoma. After 4 months, a contrast enhanced CT scan of the abdomen and pelvis was performed. No detectable local or distant recurrence seen. Right kidney was of normal appearance and excretory function. **Conclusion:** Radical nephrectomy is the preferred treatment for this tumor as it has a favorable prognosis. However, a small percentage of cases may experience recurrence or metastasis, necessitating regular follow-up.

Keywords

Mucinous Tubular and Spindle Cell Carcinoma, Renal Cell Carcinoma, Radical Nephrectomy, Case Report

1. Introduction

Renal mucinous tubular and spindle cell carcinomas are uncommon, distinctive renal tumors accounts for less than one percent of renal cell carcinoma (RCC) according to the blue book of World Health Organization 2016 [1]. It is a tumor primarily of adults, with a mean age of 58 years (13 - 81 years). Mucinous tubular and spindle cell carcinoma shows a female predominance, with a female to male ratio of 3:1. The very early cases were reported by Ordonez and Mackay, and MacLennan *et al.* as "RCC with unusual differentiation, originating in loop of Henle" and "low grade collecting duct carcinoma", respectively [2] [3].

Although some tumors are symptomatic, such as lumber region pain, abdominal mass and hematuria, the majority are discovered accidentally during abdominal imaging for other complains. An association with renal stones and end stage renal disease have been reported [4].

Mucinous tubular and spindle cell renal cell carcinoma (MTSRCC) was described as a separate subtype of RCC in the 2004 World Health Organization (WHO) tumor classification [5]. Histologically, it is characterized by the proliferation of small cuboidal and spindle cells, arranged in elongated tubules or sheets, and typically associated with a mucinous background. The most important differential diagnosis of MTSRCC is papillary renal cell carcinoma type 1, with sarcomatoid transformation [6] [7]. Fine *et al.* [8] described the histological types of MTSRCC and divided them into two categories: 1) Classic, abundance of extracellular mucin stroma; and 2) mucin-poor, little to no extracellular mucin. The presence of some features in pathology reports suggests adverse prognosis such as: necrosis, solid growth, sarcomatoid transformation, lymphovascular space invasion, and increased mitoses [8].

In some cases, it can be challenging to differentiate MTSRCC from a solid variant of papillary RCC, sarcomatoid RCC, or myoid-predominant angiomyolipoma by the morphology alone. The differentiation will need immunohistochemical staining [9]. On immunohistochemistry, MTSCC shows positive reactions for alpha methylacyl-CoA racemase (AMACR), cytokeratins CK7, CK19, epithelial membrane antigen (EMA), vimentin, and E-cadherin. The line of differentiation of MTSRCC remains controversial and has been supposed to be either the loop of Henle or the collecting duct system. This is mainly because of variation of immunohistochemical test results and electron microscopic features [9]. In cytogenetic studies, genetic abnormalities found in MTSRCC cells are mainly in the form of monosomies in chromosomes 1, 4, 6, 8 and 13, and total or partial trisomies of chromosomes 7, 11, 16 and 17 [8].

Radiologically, MTSRCC displays a common appearance that is different from clear-cell RCC but similar to papillary RCC. It usually appears as a well-demarcated, exophytic renal mass and shows a spherical or ovoid shape on computed tomography scan. Tumors less than 5 cm usually demonstrate homogenous pattern of enhancement while those larger than 5 cm often show heterogeneous enhancement pattern [10].

The prognosis for MTSRCC with classic morphology is generally favorable and complete surgical excision appears to be adequate treatment [5]. These tumors are generally of low pathological stage at diagnosis and are suitable for partial or radical nephrectomy. Few cases have demonstrated tumor recurrence, regional lymph-node spread, distant site metastases, as well as tumor-related deaths [11].

Although MTSCC was initially defined as a low-grade neoplasm with a good prognosis by the WHO in 2004 [5], its classification was revised in 2016 and the term 'indolent course' was omitted [12]. Most of cases have a benign course; however, there are some instances where the disease was aggressive, particularly when sarcomatoid changes were detected [13]. In such cases, the survival rate may be less than a year, necessitating close follow-up.

For metastatic diseases, there are no reports of systemic treatment guideline published to date. Recently, one case of metastatic MTSRCC showed a response to sunitinib (a tyrosine kinase inhibitor) has been documented [14].

Because of such a rare and important presentation, we report a mucinous tubular and spindle cell carcinoma of kidney which occurred in a 60 year old woman. The work has been reported in line with the SCARE criteria [15].

2. Case Report

On 10/11/2024 a 60 year old Sudanese housewife came to the GIT outpatient clinic of Prince Abdel-Mohsen Hospital, Alula city, Saudi Arabia, complaining of vague epigastric pain since 2 months. She performed an abdominal ultrasound examination, which showed a left renal mass lesion about 11 × 9 cm so she was referred to urology clinic. Her medical, surgical and family history was irrelevant, and did not had any urinary problems such as haematuria or loin pain. Her clinical examination was unremarkable except for a palpable abdominal mass felt below the left subcostal margin and crossing the med-line. Her ESR was 72 mm/hr the first hour haemoglobin was 10.8 g/dl and urine microscopy was negative. All other lab tests and echo cardiogram were within normal range.

On 13/11/2024 a contrast enhanced CT scan of the abdomen and pelvis was performed (**Figure 1**). The study reported a large soft tissue mass lesion measuring about 9.1 × 10.2 cm is seen arising from middle zone of the left kidney. The lesion shows heterogenous contrast enactment and is seen obviously compressing stomach and pushing proximal aorta and its main branches to the right side. Renal artery is seen compressed and pushed by the lesion, yet patent. Renal vein could not be identified, mostly compressed by the lesion. Multiple collateral vessels are seen near renal hilum. No definitive extra-lesional extension or distant metastatic deposits seen. Neither ascites nor lymphadenopathy seen. No detectable extra lesional extension or distant metastasis deposits seen. The Conclusion was a large left renal mass lesion with radiologic criteria suggestive of aggressive neoplastic lesion (Renal cell carcinoma). CT chest was free.

On 20/11/2024 the patient was operated by the urology department surgical

team; two consultants and one specialty doctor. Under general anesthesia, patient in left lateral position, Left radical nephrectomy was done through flank incision. Extraperitoneal approach, peeling of the posterior peritoneum, exposing the gerrotta's fascia which included the tumor and the kidney, selective ligation of the renal vessels, ligation of left ureter until its middle third and ligation of the ovarian vessels were done. Good hemostasis, inserted a drain, closure of the abdomen in layers were performed. The Duration of the operation was around 1.5 hours. Estimated blood loss was around 500 cc and there were no intra-operative complications recorded. The patient recovered smoothly (**Figure 2**).

Post-operative antibiotics, analgesia, antiemetics and venous thromboembolism prophylaxis were prescribed. Patient started ambulation after 12 hours and eating next morning after she was able to pass flatus. The post-operative period was smooth and unremarkable, and the patient was discharged on the seventh day after her operation. Stitches were removed after 10 days post-operative in her follow up outpatient clinic appointment.

The histopathology examination reported a specimen composed of kidney measures 9*5*2 cm with attached intra-parenchymal encapsulated solid tumor measures 14*11*8 cm. Cut section showed a pale yellow homogenous surface with small areas of hemorrhage, ureter measures 8 cm in length. No lymph nodes was positive. The tumor was limited to the kidney with no features of sarcomatoid or rhabdoid features or necrosis, no vascular or lymphatic invasion. All margins were negative for invasive carcinoma. Final diagnosis was mucinous tubular and spindle cell carcinoma (**Figure 3(a)**, **Figure 3(b)**).

On 24/3/2025 a contrast enhanced CT scan of the abdomen and pelvis was performed. No detectable local or distant recurrence seen. Right kidney was of normal appearance and excretory function (**Figure 4**).

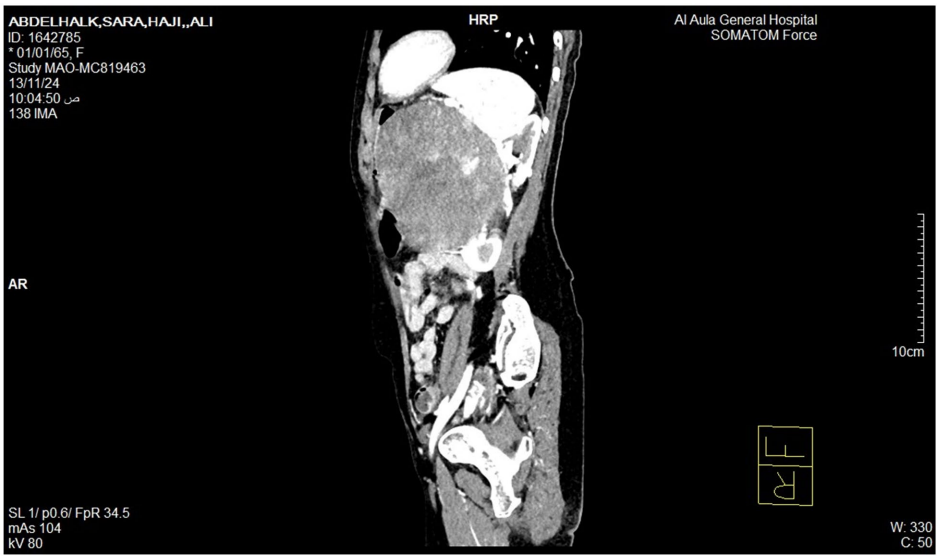


Figure 1. Sagittal image shows a pre-operative contrast-enhanced abdominopelvic CT scan of the patient with a huge left renal mass.

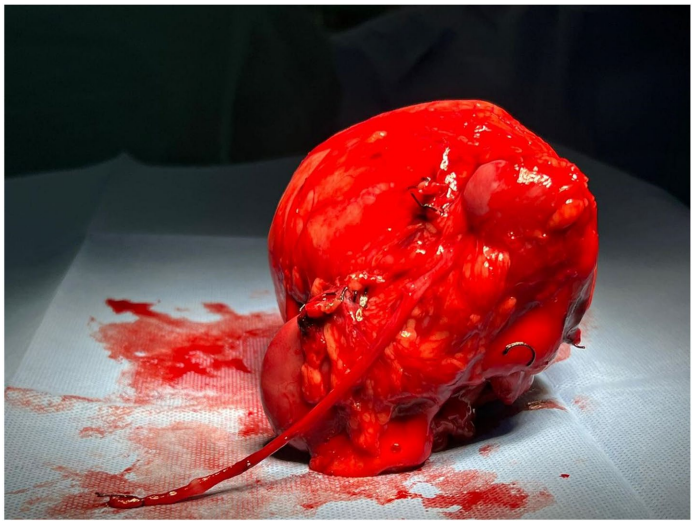
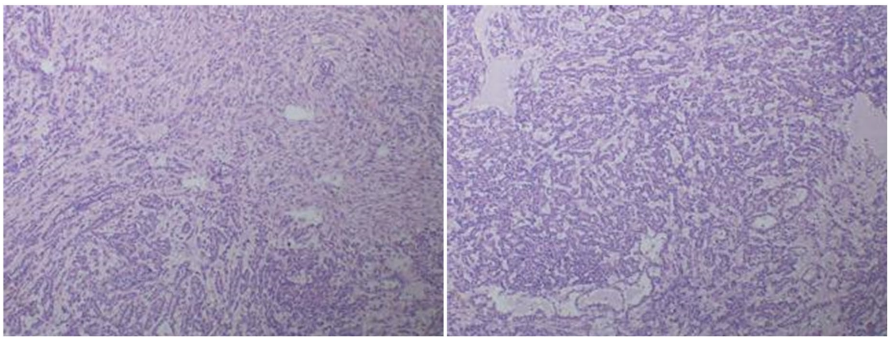


Figure 2. Macroscopic findings of the renal tumor after surgical removal. A 9 × 10 cm, pale yellow, solid tumor is located in the upper pole of the left kidney, upper ureter excised.



(a) (b)

Figure 3. Histopathology Hematoxylin and eosin-stained renal tumor, containing tubulopapillary lesions.

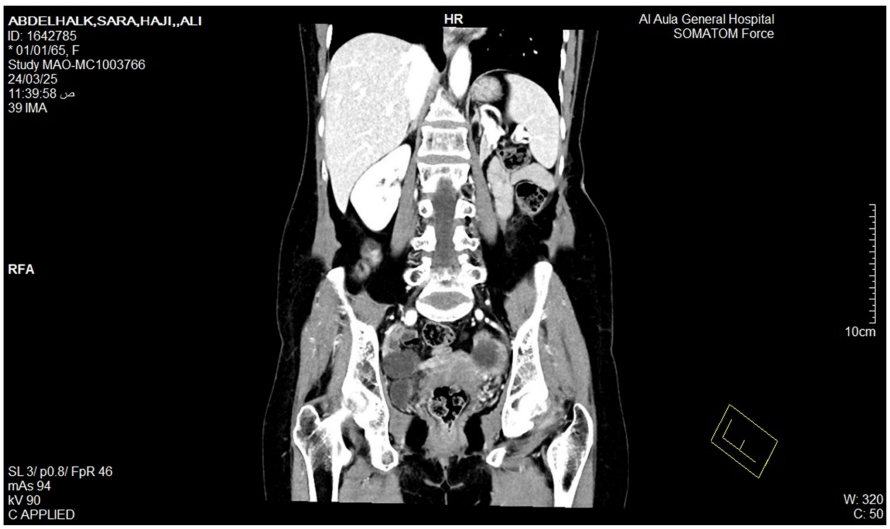


Figure 4. Post operative follow up contrast enhanced CT scan of the abdomen and pelvis showing no recurrence, coronal view.

3. Discussion

In this report, we present a typical case of renal mucinous tubular and spindle cell carcinoma. A female patient with nonspecific abdominal pain, ultrasound detection of a huge renal mass, CT diagnosis of renal cell carcinoma, radical nephrectomy and a final diagnosis was made sure by histopathological examination. No recurrence detected up till the writing of these words.

Most MTSCC cases are incidentally detected on abdominal imaging [16]. MTSRCC show a hypovascular pattern and require differentiation from other hypovascular tumors such as papillary and chromophobe RCC. Distinguishing MTSRCC from papillary and chromophobe RCC is challenging based on the CT features only as MTSCCs have been reported to have no characteristics on CT and magnetic resonance imaging (MRI) [10]. Thus, the preoperative differential diagnosis of MTSCC from papillary RCC is certainly difficult.

Although MTSCC is typically indolent, some cases have been reported that exhibit an aggressive clinical prognosis. Some reports also identified metastasis in 9.5% - 24% of patients [17]. Moreover, these aggressive MTSRCC have been reported to be associated with sarcomatoid changes and high-grade transformation in histopathology [18].

Although no recurrence or metastasis has occurred till the moment, this patient should be followed up carefully, as the tumor may recur. Overall, MTSRCC exhibits a lower malignant degree and a good prognosis compared to other tumors of RCC [19]. Generally, radical nephrectomy is the treatment of choice and no additional treatment is required following surgery in most cases. The tumor is typically of low pathological stage at the time of excision and it is extremely rare for cases to present with lymph nodes spread and other organs metastasis at the time of diagnosis. To date, a very small number of cases have presented with distant metastasis [20].

In the study conducted by Ged *et al.* [17] involving 25 patients, those displayed low-grade histological features typically had localized tumors, and only 1 out of 20 of those individuals developed recurrent metastatic disease. In contrast, among the 5 patients who had underlying high-grade histological features, they either developed or presented with metastatic disease. The overall survival rate at 3 years following diagnosis was found to be 84.8%. Noteworthy, all the deaths were attributed to metastatic disease [17]. For metastatic disease, targeted therapies, such as the use of tyrosine kinase inhibitors (such as sunitinib) have shown promise [14], while immunotherapies, such as ipilimumab and nivolumab have resulted in complete remission in some cases [21] [22].

The main limitation of this work was the short follow up time from the diagnosis till the publication of the report (6 months). In such a rare disease, lake of clear guideline is confusing and more studies are needed to adequately describe the characteristics, optimal treatments of MTSRCC.

4. Conclusion

Mucinous tubular and spindle cell carcinoma is a low grade malignant renal cell

neoplasm with specific histologic, immunohistochemical, and molecular features. Most reported cases had a favorable outcome after surgical removal. A close follow-up of these patients is important to detect recurrences early enough.

Statement of Ethics

Ethical approval was not required for this study in accordance with local or national guidelines. Written informed consent for publication of this case report and any accompanying images was obtained from the patient.

Author Contributions

Abdullah Elrashidy, Gamal R. Abdelhaliem, Mohamed Elbaiomy: urology surgical team and publication. Abdelwahab M. Gabal: radiological examination. Fikry H. Elberawy: histopathological examination. Suzan Elsharkawy: literature review and scientific writing.

Data Availability Statement

All data generated or analyzed in this study are included in this article. Further inquiries can be directed to the corresponding author.

Conflict of Interest Statement

The authors declare no conflict of interest.

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