Case Report

Renal myxoma – a case report of a rare kidney tumor, its differential diagnosis and literature review

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Abbreviations & Acronyms
CT = computed tomography
CBC = complete blood count
SMA = smooth muscle actin
FISH = fluorescence in situ hybridization
LGFMS = Low-Grade Fibromyxoid Sarcoma
RMS = Rhabdomyosarcoma

Introduction: Myxoma is a rare benign kidney tumor. Only 18 cases have been described in the literature. We report a new case of renal myxoma that arises from the renal parenchyma.

Case presentation: A 56-year-old man, medically free, presented complaining of intermittent vague left-side abdominal pain for 1 year, otherwise no history of gross hematuria or lower urinary tract symptoms. Clinical examination revealed a soft, non-tender abdomen. All laboratory investigations were within normal. Abdominal and pelvic computed tomography scan with contrast revealed a large mass located in the upper pole of left kidney with the possibility of renal cell carcinoma. A laparoscopic-assisted left robotic radical nephrectomy was performed. Microscopic examination revealed renal myxoma.

Conclusion: Renal myxoma is a very rare benign kidney tumor with excellent prognosis. Considering its rarity, it is important to recognize this entity to avoid diagnostic errors with other neoplasms with prominent myxoid features.

Key words: Rare. Renal tumor. Myxoma, Renal tumor, Myxoma.

Keynote message
We report a new case of renal myxoma. Considering its rarity, it is important to recognize this entity and its differential diagnosis in the kidney to avoid diagnostic errors with other neoplasms with prominent myxoid features.

Introduction
Myxoma is a benign tumor that is proposed to have a primitive mesenchymal origin. It has a strong predilection for the deep muscles of the extremities. It also has been found in various sites of the body.1 It has an overall good prognosis. Renal myxoma was first described by Hulk in 1887.2 Up to date, only 18 reported cases in the literature. Most of the described cases were placed in the renal parenchyma.2

Case presentation
A 56-year-old man, medically free and non-smoker, presented complaining of intermittent vague left-side abdominal pain for 1 year. No history of urinary symptoms, hematuria, or fever. Clinical examination revealed a soft, non-tender abdomen. All laboratory tests including CBC, renal and liver function tests, chemistry, and urinalysis were within normal. The patient was evaluated by CT scan imaging with contrast of abdomen and pelvis, in which a large exophytic mass was found in the upper pole of the left kidney, measuring 12 × 11 × 8.6 cm. The radiological findings were suspicious for renal cell carcinoma (Fig. 1a,b). A laparoscopic-assisted left robotic radical nephrectomy was performed. The patient had complete resolution of his flank pain, did well after surgery, and was discharged in stable condition. After 6 months, a follow-up CT abdomen with contrast was done, there
was no recurrence in the surgical bed, no abdominopelvic lymphadenopathy, or destructive bone lesions.

On gross examination, there is a $12 \times 11.5 \times 8.0$ cm well-circumscribed, encapsulated, large mass, occupying most of the kidney but does not penetrate the renal capsule or invade into the perirenal fat. It has a predominantly lobulated, soft gelatinous/myxoid cut surface and scattered, firm, white-yellowish areas (Fig. 2).

Microscopic examination revealed a hypocellular myxoid tumor, arranged in lobules that are separated by delicate fibrous strands. The lobules consist of bland spindle cells and scattered delicate blood vessels in a myxoid-rich background. No cellular atypia, lipoblasts, or mitosis (Fig. 3a,b).

A panel of immunohistochemistry stains was performed to confirm the diagnosis of renal myxoma and exclude other differential diagnoses of other neoplasms with prominent myxoid features. Tumor cells showing diffuse and strong positivity for vimentin and patchy for CD34. They are negative for Pan-cytokeratin, desmin, SMA, S100, and MUC4. Ki-67 showed a very low proliferative index (<1%). Testing for 12q13 (CHOP) and 16q11.2 (FUS) rearrangement by FISH was negative, which rules out the remote possibility of myxoid liposarcoma. The overall gross, histological, and ancillary tests findings confirm the diagnosis of renal myxoma.

**Discussion**

Myxoma is a benign myxoid lesion, commonly found deep within skeletal muscle.\(^1\)

No specific clinical or radiological findings for renal myxoma. It is mostly reported in adults (age range 36–68 years) with no gender predilection. However, flank pain was the most common clinical feature (Table 1). By radiological imaging, it usually found as a large lobulated, well-demarcated, heterogeneous mass, hyperechoic in ultrasonography and hypo-dense in computed tomography. It pushes into the renal structures without invasion, and overall raises the suspicion of a malignant mass. Pathological examination is crucial for definitive diagnosis.\(^2\) Although myxoma occurring at sites other than the kidney may on rare occasions transform into a malignant entity,\(^1\) this is not similar to renal myxoma, with all reported cases described in the literature showing no invasion, recurrence or metastasis after surgical removal of the tumor,\(^1,15\) further signifying the importance of surgical resection as it offers the best prognosis in cases of renal myxoma.\(^13\)

Macroscopically, the tumor has a mucoid/gelatinous appearance due to glycosaminoglycan production. It is often well-circumscribed and may have focally infiltrative borders. It ranges in size from 4 to 28 cm. Microscopically, it is hypocellular, hypovascular, formed of bland looking spindle-shaped cells with oval nuclei, dispersed in an abundant myxoid stroma and often surrounded by a capsule at the periphery. No mitoses, necrosis, cytological atypia, lipoblast, or giant cells have been reported in any of the cases. It has been found to stain positively for vimentin and negative for EMA, Pan-cytokeratin, desmin, SMA, and S-100. Its morphology and prognosis are indistinguishable from Myxomas in other body sites.\(^3-6\)

The differential diagnosis –although all are uncommon – as well includes neoplasms that exhibit prominent myxoid features including myxoid liposarcoma, low-grade fibromyxoid sarcoma, low-grade myxofibrosarcoma, rhabdomyosarcoma, leiomyosarcoma-myxoid variant, myxoid leiomysoma, myxoid schwannoma, myxoid neurofibroma, and mucinous
tubular and spindle cell carcinoma. A panel of immunohistochemical stains (including S-100, EMA, Pan-cytokeratin, desmin, SMA, CD34, MUC4, and Ki-67) is helpful in addition to the histological morphology to establish the diagnosis and exclude other differential diagnoses.

Myxoid Liposarcoma has a prominent plexiform vasculature with chicken-wire pattern and lipoblasts. Its incidence peaks in adults, in their fourth to the fifth decade with no gender predilection. It is usually located in the thigh but rarely involves the retroperitoneum. So far, intrarenal myxoid liposarcoma has not been reported in the literature. This neoplasm is positive for vimentin and S-100. It is associated with molecular abnormalities with either t(12;16) (q13;p11.2) \textbf{FUS-DDIT3} or uncommonly t(12;22)(q13;q12) \textbf{EWSR1-DDIT3} rearrangements.

LGFMS has a prominent collagenized stroma with an abrupt transition to a myxoid area with scattered large collagen rosettes. MUC4 is highly sensitive and specific. So far, only five cases of renal LGFMS have been described in the literature.⁷

Low-Grade Myxofibrosarcoma has curvilinear vessels with marked pleomorphic scattered cells in between. Renal origin of myxofibrosarcoma is uncommon and seen in less than 3% of all primary renal neoplasms.⁸

RMS often contains hypocellular and hypercellular areas with variable degrees of rhabdomyoblastic differentiation. It is usually positive for desmin, myogenin, and MyoD1. It is rare in the kidney, with only a few cases reported worldwide.⁹

Myxoid Leiomyoma is positive for desmin and SMA. The myxoid variant of leiomyosarcoma has been reported in the kidney.¹⁰ It has marked cytological atypia, ≥1 mitosis per 10 high power fields, and/or coagulative necrosis. It is usually positive for desmin and SMA as well.

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**Table 1** Clinicopathologic data of 19 cases of renal myxoma

| Authors                  | Age (year)/gender | Presenting symptoms            | Side/site                          | Tumor size (cm) | Treatment            | Reference No: |
|--------------------------|-------------------|---------------------------------|------------------------------------|-----------------|----------------------|---------------|
| Bolat et al.             | 27/female         | Asymptomatic                    | Left/upper pole                    | 15              | Nephrectomy          | 4             |
| Melamed et al.           | 52/female         | Right renal colic               | Left/upper pole                    | 7               | Nephrectomy          | 15            |
| Melamed et al.           | 68/female         | Asymptomatic                    | Right/paraclavicular               | 10              | Nephrectomy          | 15            |
| Appel and Schoenberg     | NS                | Hematuria                       | Right/parapelvic                   | 8               | Enucleation of mass  | 15            |
| Kundu et al.             | 36/male           | Hypochondrium mass              | Left/most of renal parenchyma      | 28              | Nephrectomy          | 15            |
| Shenasky and Gillenwater | 62/male           | Hematuria                       | Right/upper pole                   | 4               | Nephrectomy          | 15            |
| Owari et al.             | 62/male           | Asymptomatic                    | Right/middle pole                  | 8               | Nephrectomy          | 15            |
| Val-Bernal et al.        | 37/male           | Asymptomatic                    | Right/capsule                      | 6               | Nephrectomy          | 15            |
| Nishimoto et al.         | 36/male           | Asymptomatic                    | Left/upper pole                    | 9               | Nephrectomy          | 6             |
| Hakverdi et al.          | 59/male           | Lower urinary tract infection    | Right/upper pole                   | 6               | Nephrectomy          | 15            |
| Tenkorag et al.          | 50/male           | Right flank pain                | Right/middle pole                  | 4               | Radical nephrectomy  | 3             |
| Salehipour et al.        | 56/male           | Right flank pain and hematuria  | Right/upper pole                   | 8.5             | Partial nephrectomy  | 2             |
| Souza et al.             | 73/female         | Right flank pain                | Left/middle pole                   | 12              | Partial nephrectomy  | 1             |
| Thakker et al.           | 55/female         | Abdominal pain                  | Right/upper pole                   | 1.7             | Partial nephrectomy  | 5             |
| Yildirim et al.          | 82/male           | Dysuria/Flank pain/Urinary obstr | Left renal pelvis                  | 9               | Radical nephrectomy  | 12            |
| Bernardino et al.        | 85/male           | Hematuria                       | Left/inferior calyx                | 2               | Nephroureterectomy   | 13            |
| Shah et al.              | 43/female         | Asymptomatic                    | Left/mid-upper pole                | 5               | Radical nephrectomy  | 14            |
| Suthar et al.            | 48/female         | Abdominal pain                  | Right/mid-lower pole               | 13              | Radical nephrectomy  | 11            |
| Present case             | 56/male           | Abdominal pain                  | Left/most of renal parenchyma      | 12              | Radical nephrectomy  |               |

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Myxoid Schwannoma and Myxoid Neurofibroma are usually positive for S-100.

Mucinous tubular and spindle cell carcinoma is an indolent renal epithelial neoplasm, formed of tubular architectures admixed with bland spindle cells in a background of myxoid stroma. It is positive for Pan-cytokeratin, low molecular weight keratins, PAX8 and AMCAR.

In conclusion, Renal myxoma is a very rare benign kidney tumor with an excellent prognosis. The mainstay of treatment is radical nephrectomy with no evidence of recurrence or metastases in any case. Considering its rarity, it is important to recognize this entity and its differential diagnosis in the kidney to reach the correct diagnosis.

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Author Contributions
Rawan Eid Hudairy: Resources; writing – original draft; writing – review and editing. Omar Buksh: Resources; writing – review and editing. Rabea Akram: Supervision. Adel Alammari: Supervision. Jaudah Al-Maghrabi: Supervision. Zuhoor Almansouri: Resources; supervision; writing – review and editing.

Conflict of interest
No conflicts of interests.

Approval of the research protocol by an Institutional Reviewer Board
Number is 2021CR11.

Informed consent
Not applicable.

Registry and the registration no. of the study/trial
Not applicable.

References
1. Souza C, Carneiro K, Leite K, A. Junior A, Costa F. Renal myxoma: a case report. J. Bras. Patol. Med. Lab. 2015; 51: 113–6.
2. Salehipour M, Geramizadeh B, Dastghheib N, Makarem A, Asadollah Poor A, Taheri N. Renal myxoma, a case report and review of the literature. Urol. Case Rep. 2019; 23: 21–2.
3. Tenkorang S, Kharbach Y, Omama J et al. Myxoma of the kidney – an unusual benign renal tumor: a case report. J Med Case Rep. 2017; 11: 41.
4. Bolat F, Turunk T, Kayaselcuk F, Ulusan F, Bal N. Primary renal myxoma: a case report. Turk. J. Pathol. 2007; 23: 160–3.
5. Thakker P, Ramsey T, Navarro F. Renal Myxoma, an Incidental Finding. Urol. Case Rep. 2017; 13: 131–2.
6. Nishimoto K, Sumitomo M, Kakoi N, Asano T, Hayakawa M. Case of renal myxoma. Int. J. Urol. 2007; 14: 242–4.
7. Bhattachar R, Aggarwal S, Yadav S, Tomar V. Primary low-grade Fibromyxoid sarcoma of kidney—an extremely rare entity. Indian J. Surg. 2017; 80: 281–3.
8. Seo H, Park S, Lee K et al. Primary renal myxofibrosarcoma in a woman: A case report and literature review. J. Cytol. Histol. 2017; 8: 4.
9. Samkari A, Al-Maghrabi H. Rhabdomyosarcoma of the kidney. J. Pediatr. Surg. Case Rep. 2018; 32: 62–7.
10. Yokose T, Fukuda H, Ogiwara A, Sakai K, Saitoh K. Myxoid leiomyosarcoma of the kidney accompanying ipsilateral ureteral transitional cell carcinoma: A case report with cytological, immunohistochemical and ultrastructural study. Pathol. Int. 1991; 41: 694–700.
11. Suthar D. Renal myxoma - a rare variant of benign genitourinary tumour. J. Clin. Diagn. Res. 2015; 9: 11–12.
12. Yildirim U, Erdem H, Kayici A, Uzunlar A, Tekin A, Kuzey M. Myxoma of the renal sinus: case report and literature review. Turk. J. Pathol. 2012; 28: 76.
13. Bernardino R, Severo L, Lemos L, Pinheiro L. Renal myxoma: an unforeseen diagnosis. Arch. Ital. Urol. Androl. 2020; 92: 273–4.
14. Shah A, Sun W, Cao D. Myxoma of the kidney associated with hemorrhage. Indian J. Surg. 2013; 75: 480–3.
15. Hakverdi S, Gorur S, Yaldiz M, Kiper A. Renal myxoma: a case report and review of the literature. Turk. J. Urol. 2010; 36: 318–21.
16. Kusumi T, Minakawa M, Fukui K et al. Cardiac tumor comprising two components including typical myxoma and atypical hypercellularity suggesting a malignant change. Cardiovasc. Pathol. 2009; 18: 369–74.