Case Report

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

Rawan Hudairy,[1] Omar Buksh,[2] Rabea Akram,[2] Adel Alammari,[2] Jaudah Al-Maghrabi[1] and Zuhoor Almansouri[1]

Department of [1]Pathology and [2]Urology, King Faisal Specialist Hospital and Research Center, Jeddah, Saudi Arabia

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

Correspondence: Rawan Hudairy M.D., Department of Pathology, King Faisal Specialist Hospital and Research Center, Jeddah, Saudi Arabia.
Email: R.hudairy@hotmail.com

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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 resolvement 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

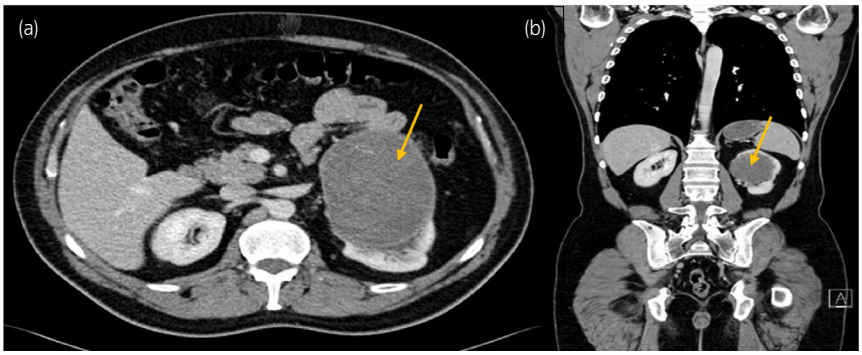

Fig. 1 (a, b) Computed tomography scan revealed a large mass in the upper pole of the left kidney.

was no recurrence in the surgical bed, no abdominopelvic lymphadenopathy, or destructive bone lesions.

On gross examination, there is a 12 × 11.5 × 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.

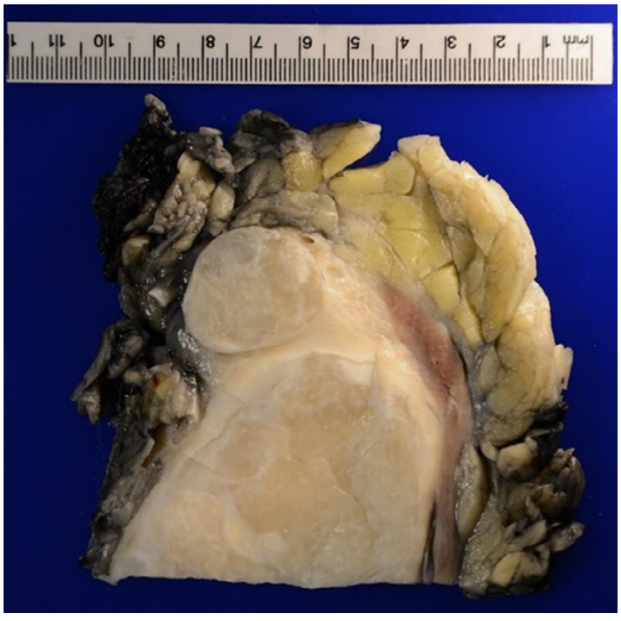

Fig. 2 The kidney revealing a welsl-circumscribed mass sparing a rim of the kidney at the periphery. It has a lobulated soft predominantly gelatinous/myxoid cut surface.

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,[16] 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 leiomyoma, myxoid schwannoma, myxoid neurofibroma, and mucinous

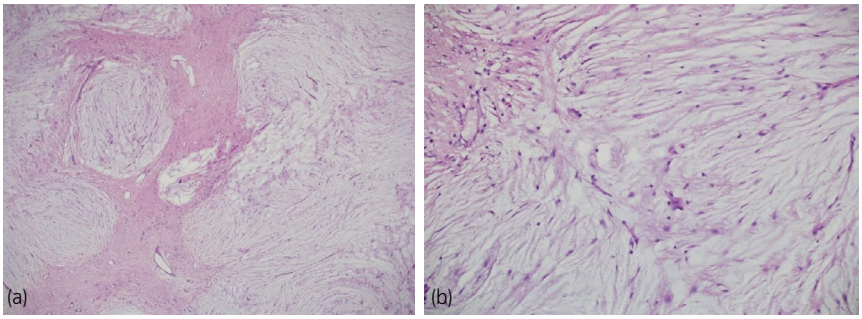

Fig. 3 (a) The low power (4×) highlights the lobulated nature of the myxoid-rich tumor. (b) The high power (20×) reveals the hypocellular bland spindle cells proliferation.

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/lower pole	15	Nephrectomy	4
Melamed *et al*.	52/female	Right renal colic	Left/lower pole	7	Nephrectomy	15
Melamed *et al*.	68/female	Asymptomatic	Right/upper pole	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/lower 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/lower pole	9	Nephrectomy	6
Hakverdi *et al*.	59/male	Lower urinary tract infection	Right/upper pole	6	Nephrectomy	15
Tenkorag *et al*.	50/female	Right flank pain	Right/middle pole	4	Radical nephrectomy	3
Salehipour *et al*.	56/male	Right flank pain and hematuria	Right/lower 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 obstruction	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	

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) *FUS-DDIT3* or uncommonly t(12;22)(q13;q12) *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.

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 wight 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, Dastgheib 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, Omana 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 Bhattar 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 literaturereview. *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 variety of benign genitourinary tumour. *J. Clin. Diagn. Res.* 2015; **9**: 11–12.
12 Yildirim U, Erdem H, Kayikci 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.