



Unraveling Rare Renal Malignancy At Our Institute: A Comprehensive Clinicopathological Retrospective Case Series

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Abstract

Background:

Renal malignancies are predominantly clear cell renal cell carcinomas; however, rare subtypes such as collecting duct carcinoma, renal sarcomas, medullary carcinoma, and primary renal lymphoma pose diagnostic and therapeutic challenges due to their aggressive nature and lack of standardized treatment protocols.

Objective:

To analyse clinical presentations, histopathology, treatment modalities, and outcomes in a series of patients with rare renal malignancies diagnosed at Bhaktivedanta hospital and research institute.

Methods:

Retrospective review of 3 patients diagnosed with rare renal malignancies excluding clear cell RCC. Data on demographics, presenting symptoms, radiological features, histopathological diagnosis, treatment, and follow-up outcomes were collected and analysed.

Results:

The series included 3 patients with diagnoses of renal sarcoma. Patients presented primarily with haematuria and flank pain. Imaging showed heterogeneous renal masses with variable enhancement patterns. Histopathology and immunohistochemistry confirmed diagnoses. Radical nephrectomy was performed in all operable cases, with adjuvant chemotherapy or radiotherapy based on tumor type. Median follow-up was 18 months; overall survival was short due to aggressive tumor biology.

Conclusion:

Rare renal malignancies exhibit aggressive clinical behaviour and poor prognosis. Early diagnosis with histological confirmation and multimodal treatment strategies may improve outcomes. Further research is required to establish effective therapeutic protocols.

Keywords: Renal Sarcoma , Rare Renal Malignancy , Aggressive Renal Tumor

Introduction

Primary renal sarcomas are rare malignant tumors arising from mesenchymal structures of the kidney, comprising less than 1% of all primary renal neoplasms (1). These tumors originate from smooth muscle, vascular endothelium, nerve sheath cells, or primitive mesenchymal tissues, giving rise to a heterogeneous group that includes leiomyosarcoma, synovial sarcoma, Ewing sarcoma/PNET, angiosarcoma, malignant peripheral nerve sheath tumor, liposarcoma, and undifferentiated pleomorphic sarcoma (1,2).

The clinical presentation of renal sarcomas is often nonspecific, with symptoms such as flank pain, abdominal mass, or haematuria, frequently mimicking renal cell carcinoma. Due to the silent growth pattern and large retroperitoneal space, tumors often present at advanced stages, sometimes exceeding 10–15 cm at diagnosis (1,3). Systemic symptoms such as fever and weight loss are more common in Ewing sarcoma/PNET (4).

Radiologically, renal sarcomas typically appear as large heterogeneous masses with necrotic or haemorrhagic areas. Imaging alone cannot reliably distinguish sarcoma from carcinoma, making histopathological examination essential. Immunohistochemistry (IHC) and molecular diagnostics play critical roles, especially in translocation-associated sarcomas like synovial sarcoma (SYT-SSX fusion) and Ewing sarcoma (EWSR1-FLI1 fusion) (3,4).

Management is challenging due to the rarity of these tumors and absence of standardized guidelines. Radical nephrectomy remains the cornerstone of treatment, while adjuvant therapies depend on histological subtype. Prognosis varies, with angiosarcoma and Ewing sarcoma having particularly poor outcomes (1,5).

This study presents a detailed case series of three rare renal sarcomas, aiming to highlight diagnostic clues, subtype-specific characteristics, and clinical outcomes to contribute to existing literature.

Methods

This retrospective case series included 3 patients diagnosed with primary renal sarcoma between 2024-2025 at our institute.

Inclusion Criteria

1. Histologically confirmed primary renal sarcoma
2. Complete clinical, radiological, and pathological records
3. Underwent surgical excision

Data Collection

Data were extracted on:

1. Demographics and clinical presentation
2. Radiologic findings (CT/MRI)
3. Gross and microscopic pathology
4. Immunohistochemistry results
5. Molecular diagnostics (RT-PCR, FISH) where indicated
6. Treatment modality (surgery, chemotherapy, radiotherapy)
7. Follow-up and outcomes

Histopathological evaluation was performed with haematoxylin and eosin staining. IHC markers were used depending on morphology. Molecular testing was performed for suspected synovial sarcoma and Ewing sarcoma.

Case Series

Case 1: UNDIFFERENTIATED PLEOMORPHIC SARCOMA

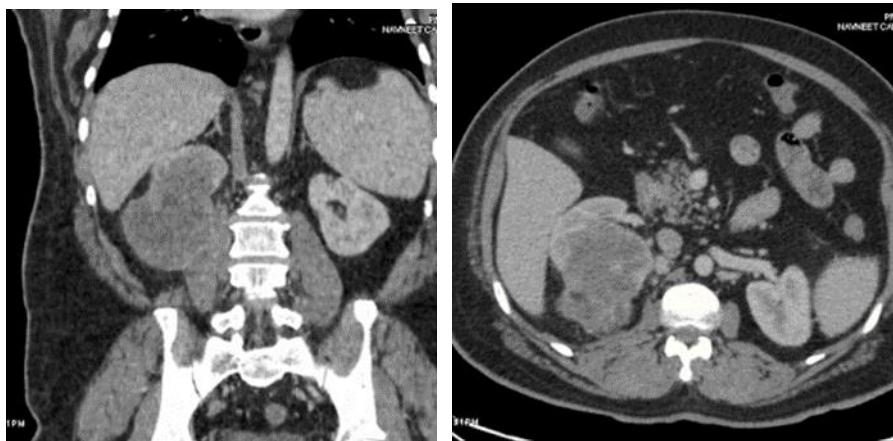
Patient: 54-year-old male

Presentation: Right flank pain with intermittent on and off haematuria

Imaging:

Contrast-enhanced CT revealed a 7.3 x 7.8 x 6.4 cm heterogeneous mass localized to the cortical region of the right kidney with extending into right pelvicalyceal system, inferomedially infiltrating the left psoas muscle. (figure 1).

Figure 1- CT revealed a 7.3 x 7.8 x 6.4 cm heterogeneous mass localized to the cortical region of the right kidney

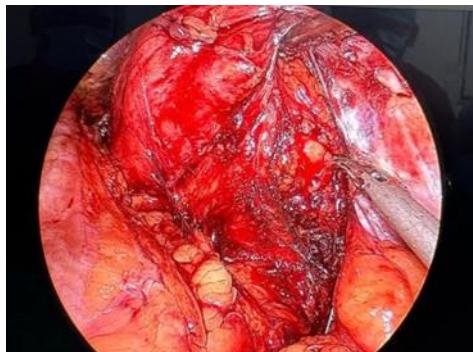


PET CT was done suggestive of: - An ill-defined metabolically active (SUVmax: 50.9) heterogeneously enhancing lobulated exophytic mass lesion interspersed with areas of necrosis is epicentered in the cortex of the right kidney, extending into the right pelvicalyceal system, infero-medially infiltrating the left psoas muscle, measuring approximately 9.3 (AP) x 8.3(W) x 9.9(SI) cm in size. No involvement of right renal vessels. Few small mildly metabolically active (SUVmax: 3.1) perilesional and retrocaval nodes are seen, the largest node measuring approximately 1.7 x 1.4 cm in size.

Management: Right laparoscopic radical nephrectomy.

Intraoperative findings: a large, variegated mass approx. 7x8x8cm occupying the entire kidney, sparing a part of upper pole, 6 firmly adherent to the abdominal wall and right Psoas muscle with perihepatic adhesion. (figure 2)

Figure 2: Intraoperative imaging of right renal mass



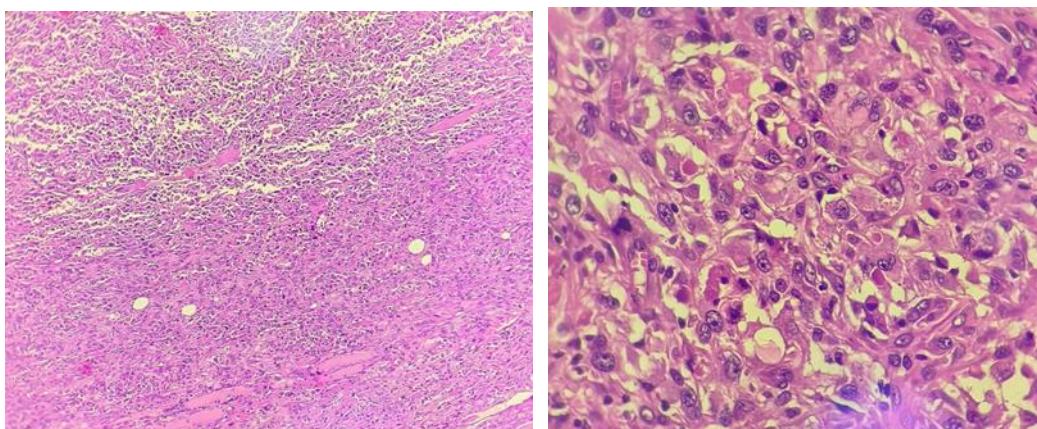
Histopathological findings (figure 3):

1. The tumor was composed of diffusely infiltrating spindled to epithelioid, markedly pleomorphic cells.
2. Individual tumor cells showed variable amounts of cytoplasm and had enlarged vesicular to hyperchromatic nuclei with prominent nucleoli at places.
3. The mitotic rate was greater than 20 per 10 high-power fields. Atypical mitoses were noted.
4. Bizarre pleomorphic tumor cells and multinucleated tumor cells were noted.
5. Areas of necrosis were noted.
6. The stroma was hyalinized in areas with collagen bundles and showed focal calcification.

Impression:

Undifferentiated pleomorphic sarcoma involving the right kidney and adherent skeletal muscle.

Figure 3- Histopathology showing spindled to epithelioid cells with prominent nucleoli



Outcome And Follow Up:

1. Patient was tolerated the surgery well and then after 6-month patient had complaints of right flank pain again.
2. CT was done which is Suggestive of: - FDG avid heterogeneously enhancing, large, lobulated solid cystic mass in the right renal fossa, infiltrating the right psoas muscle and posterior abdominal wall and showing FDG avidity in the solid component, mass measures 16.7x13.5x21.5 cm (APxTrxCC), SUVmax 32.3. The mass is superiorly infiltrating seg VI of liver. It is abutting the right 12 rib, lumbar vertebrae, sacrum and right iliac bone th without erosion. Inferiorly it is reaching up to the pelvis. Low grade FDG uptake in another smaller necrotic lesion abutting the IVC in the post operative site, measures 3.2x2.8, SUVmax 5.2. Few other discrete nodular masses with similar morphology noted abutting and infiltrating the right anterolateral abdominal wall in the right iliac fossa, largest measures 7.8x8.1x7.2 cm, SUVmax 27.3. overall findings were suggestive of disease recurrence.
3. patient was started on intravenous Nivolumab as immunotherapy.

Case 2: RENAL LEIOMYOSARCOMA

Patient: 47-year-old female

Presentation: Weight loss, and anorexia over 4 months and palpable mass in left lumbar and umbilical region (figure 4)



•Imaging:

CT showed Well defined solid heterogeneously enhancing lesion (6.8 x 7 x 6.2 cm) with necrotic areas arising from the lower pole of left kidney. Mass effect in the form of left moderate to gross hydronephrosis with parenchymal thinning. Delayed uptake and excretion of contrast from left kidney - findings suggestive of neoplastic aetiology. Well defined heterogeneously enhancing retroperitoneal masses with necrotic areas within? conglomerated necrotic retroperitoneal lymphadenopathy? retroperitoneal neoplastic lesion. Enhancing lesion in the left psoas muscle. Non-enhancing tiny lesion in the subcutaneous left anterior abdominal wall as described above. (figure 5)

Figure 5 - Well defined solid heterogeneously enhancing lesion (6.8 x 7 x 6.2 cm) with necrotic areas arising from the lower pole of left kidney



PET CT- Metabolically active heterogeneously enhancing partially exophytic lobulated mass lesion epicentered in the lower pole of left kidney, with its size and extent, as described, indicating primary disease involvement. Further biopsy correlation is recommended.

Multiple metabolically active discrete and conglomerate retro pancreatic, peripancreatic, retroperitoneal and bilateral common iliac adenopathy – likely nodal metastases.

Mildly metabolically active heterogeneously enhancing intermuscular deposit in the left psoas muscle – likely metastasis.

Management: Left Open Radical Nephrectomy with post operative chemotherapy (docetaxel + gemcitabine)

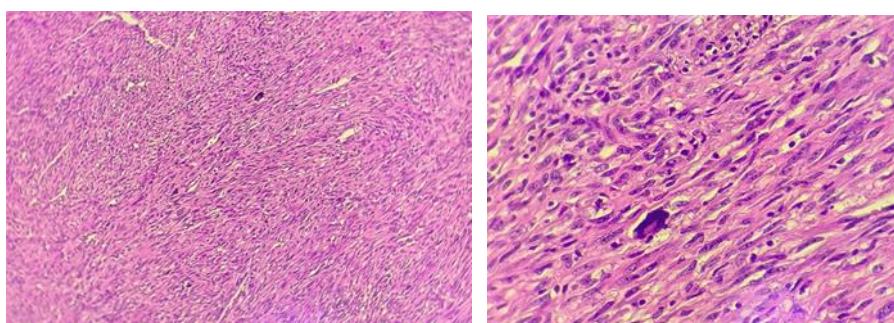
Intraoperative findings:

left sided renal mass greyish red, variegated appearance, hard very vascular, necrotic areas seen, irregular shaped approx. 7x5.5x6cm occupying the inferior and mid pole, hydronephrotic upper pole. 2)a very hard conglomerated greyish white lymph nodal mass in para-aortic, para-caval region, adherent to the underlying major vessels, very vascular approx. 5.5x6x5cm. (figure 6)

Figure 6 – left sided renal mass with enlarged conglomerated lymph node

Histopathology: Multiple sections studied show high grade spindle cell sarcoma composed of intersecting fascicles and whorls of spindly tumor cells with eosinophilic cytoplasm and oval to spindly nuclei with blunt edges (figure 7)

1. Tumor cells show marked nuclear pleomorphism.
2. Focal areas of epithelioid morphology are noted.
3. Bizarre pleomorphic tumor cells and multinucleated tumor giant cells are noted.
4. Mitotic rate is 8-10/10 hpf. Atypical mitoses are seen.
5. Necrosis (<50% of tumor) is seen.
6. Hemangiofibromatous vasculature is noted.
7. Tumor involves lower pole of left kidney.
8. Tumor measures 8.4 x 7.5 x 6.5 cm.
9. Tumor infiltrates into perinephric fat and sinus fat.
10. Tumor cells show patchy positivity for SMA and vimentin, focal positivity for desmin.
11. They are negative for pan-ck, pax8, cd99, h-caldesmon, cd34, s100 protein, myogenin, stat6, hmb45 and cd117.
12. Multifocal high grade leiomyosarcoma of left kidney and ureter with metastatic conglomerated para-aortic, para-caval and retroperitoneal lymph nodes.

Figure 7- spindly tumor cells with eosinophilic cytoplasm and oval to spindly nuclei with blunt edges.

Outcome: Local recurrence at 5 months (figure 8); palliative care was initiated and then patient succumbed because of severe disease progression at 11 month post diagnosis.

Figure 8 – CT showing local recurrence after 5 months

Case 3: UNDIFFERENTIATED PLEOMORPHIC SARCOMA

Patient: 73-year-old male patient

Presentation: Haematuria and right abdominal discomfort.

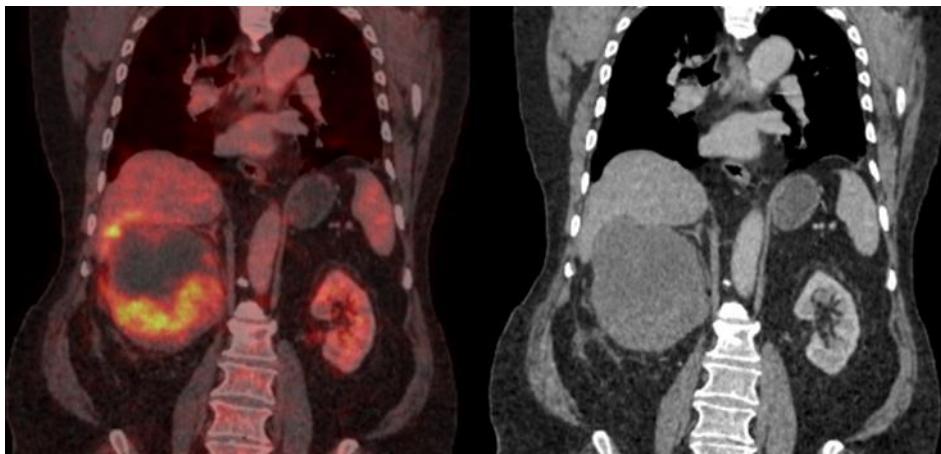
Imaging: CT revealed A well-defined lobulated thick walled hypodense cystic lesion showing solid component within and more than 3 irregular enhancing septations are more than 3 mm in size with extensions as follows - indenting the inferior surface of segment VI of right lobe of liver with maintained fat planes, extension in right perinephric space, compressing the upper and middle minor and major calyx with normal contrast uptake and excretion, abutting the medial border of right psoas muscle. Capsular hypodense heterogeneously enhancing lesion is noted in the inferior surface of segment VI of right lobe of liver? local metastasis. Nodular heterogeneously enhancing lymph node anterosuperior to the right kidney within the perirenal fascia limited by the Gerota's fascia

suggestive of regional metastatic lymphadenopathy. ---The above findings are likely suggestive of neoplastic aetiology (Cystic RCC unless otherwise proven - Bosniak type IV cyst). According to TNM staging, T4 N1 M0.

PET CT- A well-defined metabolically active (SUVmax: 11.5) heterogeneously enhancing solid-cystic mass lesion is seen involving the upper pole of right kidney, infiltrating the right perirenal fascia, without involvement of right renal artery or vein, indenting into the segment VI of liver, measuring approximately 9.8(AP) x 10.6(W) x 12.7(SI) cm in size. Few metabolically active (SUVmax: 15.1) enhancing nodular lesions are seen surrounding the afore-mentioned mass lesion, the largest lesion measuring approximately 2.1(AP) x 1.6 (W) cm in size.

A focal subcapsular hypodensity is seen in the segment VI of liver, without any metabolic activity, measuring approximately 1.7 x 1.0 cm in size (figure 9)

Figure 9- CT and PET CT suggestive of right renal mass encasing segment VI of liver



Treatment: Right open radical nephrectomy

Intraoperative findings: - approx. 15 x 10 cm² right kidney with mass variegated in appearance with involvement of perinephric fat with right kidney adhered to gerota's fascia (figure 10)

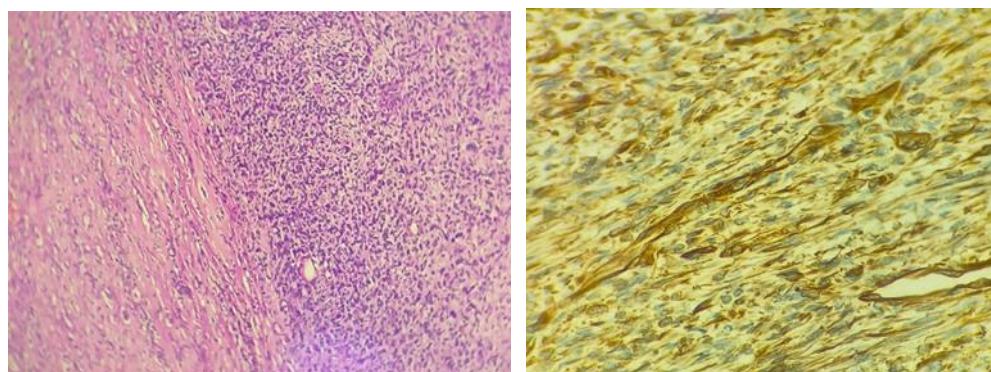
Figure 10- showing right kidney with mass with perinephric fat



Histopathology: Tumor is composed of fibro sarcomatous to storiform areas of spindly cells. Individual tumor cells show round to ovoid to spindly, markedly pleomorphic, vesicular nuclei. Bizarre pleomorphic and multinucleated tumor giant cells were seen. Mitosis +(18-20/10 hpf). Tumor infiltrates into perinephric fat as well as gerota's fascia. On IHC tumor cells are positive for Vimeticin and negative for CK, PAX8, GATA3, CD34, S100, desmin, HMB45 and p40 (figure 11).

Suggestive of: Undifferentiated pleomorphic sarcoma of right kidney infiltrating perinephric fat and gerota's fascia.

Figure 11 – fibro sarcomatous to storiform areas of spindly cells, tumor cells are positive for Vimetin and negative for CK, PAX8, GATA3, CD34, S100, desmin, HMB45 and p40.



Outcome: Disease progressed with local recurrence at 5 months; patient succumbed at 12 months post-diagnosis.

SUMMARY:

Case number	Presentation	Management	Histopathology	Adjuvant therapy	Recurrence	Outcome
1	Flank pain with hematuria	Right laparoscopic radical nephrectomy	Undifferentiated pleomorphic sarcoma	Nivolumab and Chemotherapy	Yes	Alive with disease at 12 months
2	Lumbar pain, weight loss and anorexia	Left open radical nephrectomy	Leiomyosarcoma	Docetaxel +gemcitabine (Chemotherapy)	Yes	Death at 11 months
3	Hematuria	Right open radical nephrectomy	Undifferentiated pleomorphic sarcoma	No	Yes	Death at 12 months

Discussion

Primary renal sarcomas are exceptionally rare, accounting for <1% of primary renal tumors, a figure consistently reported across multiple retrospective series (1,2). Their rarity contributes to a lack of standardized diagnostic or therapeutic protocols. Our 3-patient case series—including leiomyosarcoma (n=1) and undifferentiated pleomorphic sarcoma (UPS) (n=2)—demonstrates several clinical and pathological patterns consistent with previously published studies, while highlighting the aggressive nature of these malignancies.

In this case series of three patients with primary renal sarcomas, the most common presenting symptom was

hematuria, observed in **66.7% (2/3)** of cases, while **flank or lumbar pain** was reported in **66.7% (2/3)**. Systemic symptoms such as weight loss and anorexia were present in **33.3% (1/3)** of patients, reflecting advanced disease at presentation.

All patients (**100%**) underwent **radical nephrectomy**, which remains the mainstay of treatment for renal sarcomas. A minimally invasive laparoscopic approach was used in **33.3% (1/3)** of cases, while **66.7% (2/3)** required an open surgical approach, likely due to tumor size or local extension.

Histopathological examination revealed **undifferentiated pleomorphic sarcoma** in **66.7% (2/3)** of patients and **leiomyosarcoma** in **33.3% (1/3)**,

consistent with existing literature identifying leiomyosarcoma and undifferentiated sarcoma as the most common renal sarcoma subtypes.

Adjuvant therapy was administered in **66.7% (2/3)** of cases. One patient (**33.3%**) received immunotherapy with nivolumab, while another (**33.3%**) was treated with combination chemotherapy (docetaxel and gemcitabine). Despite adjuvant treatment, **tumor recurrence occurred in 100%** of patients, underscoring the aggressive nature of renal sarcomas and their high propensity for recurrence.

Overall survival outcomes were poor, with **mortality observed in 66.7% (2/3)** of patients in 1 year follow up. Notably, the only surviving patient (**33.3%**) had a favourable outcome despite recurrence and had received immunotherapy and chemotherapy, suggesting a potential role of multidisciplinary approach in selected cases, although conclusions are limited by the small sample size.

Clinical Presentation

In our study, the most common presenting symptoms were flank pain, haematuria, and weight loss, consistent with the nonspecific presentations described by Cheville and Menon and Parham et al. (1,5). Tumor sizes ranged from 6–12 cm, reflecting delayed detection due to retroperitoneal location. Two patients presented with regional lymphadenopathy and evidence of local invasion, mirroring observations in previous renal sarcoma series (3,4).

Radiologic Features

Imaging revealed large, heterogeneous masses with necrotic areas, sometimes compressing adjacent structures. These features are nonspecific and often mimic RCC, underscoring the importance of histopathological diagnosis (1,3,4,7). PET CT was useful for assessing metabolic activity and detecting metastasis or recurrence.

Histopathology and Immunohistochemistry

Histology and IHC were essential for definitive diagnosis. The leiomyosarcoma case demonstrated classic spindle cell morphology with diffuse SMA and desmin positivity, consistent with Cheville and Menon (1). UPS cases showed highly pleomorphic spindle-to-epithelioid cells, vimentin positivity, and lack of lineage-specific markers, in agreement with Miettinen and Fetsch (6). Molecular testing is required for

translocation-associated sarcomas, although our series did not include synovial or Ewing sarcomas; in literature, these require SYT-SSX1/2 or EWSR1-FLI1 fusion testing for diagnosis (3,5).

Treatment

All patients underwent radical nephrectomy, consistent with literature recommending surgery as the cornerstone of management (1,2,12). Margin status strongly influenced outcomes, as previously reported (13). Adjuvant chemotherapy was administered to the leiomyosarcoma patient (docetaxel + gemcitabine) but was not curative, consistent with previous reports of variable chemo-responsiveness in renal sarcomas (7,15,16). Immunotherapy was offered for recurrent UPS in one patient, reflecting evolving systemic management approaches.

Outcomes and Prognosis

Median follow-up was 18 months. Two UPS patients developed early local recurrence and metastasis, while the leiomyosarcoma patient succumbed to systemic disease within 5 months. These findings align with global data showing poor survival in renal sarcomas, particularly high-grade subtypes and those with local invasion or metastasis (1,4,6,18). Our results highlight that UPS tends to recur rapidly, while leiomyosarcoma has slightly slower progression but remains aggressive.

Comparison With Literature

Key observations from our series that align with previous studies include:

1. Subtype prevalence: Leiomyosarcoma and UPS are among the most reported renal sarcomas (1,2,5).
2. Diagnostic limitations: Imaging cannot reliably differentiate sarcoma from RCC; IHC and molecular studies are crucial (3,4,7).
3. Therapeutic approach: Radical nephrectomy remains primary treatment; adjuvant therapies are subtype-specific and often of limited efficacy (12–16).
4. Prognosis: High-grade tumors with local invasion, vascular involvement, or metastasis have poor outcomes, with 5-year survival <35% (6,13,18).

This series differs slightly from other reports in the higher proportion of UPS and the rapidity of recurrence, likely reflecting referral bias and tumor aggressiveness.

Limitations

The main limitations include the small sample size, retrospective design, and short follow-up. Heterogeneity of sarcoma subtypes further limits generalizability.

Conclusion

Rare renal sarcomas are aggressive and diagnostically challenging tumors, representing less than 1% of all primary renal neoplasms. Our case series highlights that these tumors frequently present at advanced stages due to nonspecific symptoms such as flank pain, hematuria, and constitutional complaints, often resulting in large tumor size and local invasion at diagnosis. Imaging alone is insufficient for accurate differentiation from more common renal malignancies, underscoring the critical role of histopathology, immunohistochemistry, and molecular testing in establishing a definitive diagnosis.

Surgical resection, specifically radical nephrectomy, remains the cornerstone of treatment for localized disease, but adjuvant therapies—including chemotherapy, radiotherapy, or immunotherapy—must be tailored to the histological subtype. Despite aggressive multimodal management, prognosis remains poor, particularly for high-grade subtypes such as undifferentiated pleomorphic sarcoma and Ewing sarcoma/PNET, which demonstrate rapid recurrence and early metastasis. Leiomyosarcomas may show slightly slower progression but still carry significant mortality risk.

This series reinforces the need for early recognition and prompt intervention, as well as close postoperative surveillance to detect recurrence. The rarity and heterogeneity of renal sarcomas pose significant challenges for standardized treatment protocols. Therefore, multicentre collaboration and larger prospective studies are essential to better understand tumor biology, optimize therapeutic strategies, and improve overall survival. Moreover, integration of emerging systemic therapies, including targeted agents and immunotherapy, warrants further exploration in this patient population.

This series highlights the **aggressive behaviour, high recurrence rate, and poor prognosis** associated with primary renal sarcomas, even following radical surgery and adjuvant therapy. Larger studies are needed to better define the role of systemic therapies and improve long-term outcomes in this rare malignancy.

In conclusion, rare renal sarcomas demand a multidisciplinary approach involving urologists, oncologists, pathologists, and radiologists, with individualized treatment plans guided by tumor subtype, stage, and patient factors. Raising awareness of these aggressive tumors and compiling comprehensive case series will be crucial in advancing knowledge and improving clinical outcomes for this uncommon but deadly group of renal malignancies.

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