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Incidentally Discovered Primary Renal Leiomyosarcoma in a Middle Aged Female Presenting with Symptoms of Incomplete Abortion: A Rare Case Report

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Abstract

Background Primary renal leiomyosarcoma (LMS) is an exceptionally rare malignant mesenchymal tumor, constituting <1.5% of all primary renal malignancies. Diagnosis is difficult because clinical and radiologic features overlap with other renal neoplasms.

Case Presentation We describe a 37-year-old woman, notably younger than the typical age reported for renal LMS, who presented with abnormal uterine bleeding and incomplete abortion. During gynaecologic imaging, an incidental exophytic right-renal mass was detected. CECT and CT angiography revealed a heterogeneously enhancing lesion with necrosis abutting adjacent structures. The patient underwent laparoscopic radical nephrectomy. Histopathological examination showed fascicles of spindle cells with pleomorphism, atypical mitoses, and necrosis involving <50% of tumor area. Immunohistochemistry confirmed smooth muscle differentiation with positivity for vimentin, desmin, and SMA, while negative for cytokeratin, S100, and HMB-45. A final diagnosis of well-differentiated primary renal leiomyosarcoma (FNCLCC grade 1, stage pT3aNxMx) was rendered. The postoperative course was uneventful, and the patient remains well at 3-month follow-up.

Conclusion This case is novel for its incidental detection during gynaecologic evaluation for incomplete abortion and for occurring in a young female patient (37 years), whereas most reported renal LMS present later in life and at higher grade. These features underscore the importance of considering LMS in atypical contexts and highlight the need for awareness among both urologists and gynaecologists.

Key words renal leiomyosarcoma, primary renal sarcoma, kidney neoplasm, smooth muscle tumor, radical nephrectomy, case report

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Introduction

Renal tumors can be renal cell in origin or can be metanephric, mixed, embryonal and mesenchymal [1]. Most common renal tumors are renal cell tumors, accounting for 2% of all cancers globally [2]. Mesenchymal tumors constitute only about 4.5% of all adult primary renal tumors [3]. LMS are malignant tumors of smooth muscle origin. Primary renal LMS are extremely rare, constituting only <1.5% of all primary renal malignancies [4-6]. Owing to its rarity, there is scarcity of knowledge regarding treatment, metastasis and long-term survival.

To add to the current understating of renal LMS, we present a case of incidentally discovered primary renal LMS in a middle-aged female.

Case presentation

Clinical presentation

A 37-year-old lady presented to gynaecology OPD with chief complaints of abnormal bleeding per vaginum for a period of 5 days. On local examination, external genitalia was unremarkable and external os was visible and unremarkable. On per abdomen examination, no significant abnormality was detected. Preliminary blood investigations were within normal limits.

Imaging

On further workup, a whole abdomen ultrasonography was performed. On USG, the uterus was retroverted, normal in size with presence of heterogenous echotexture with few well defined heterogenous hypoechoic lesions, largest in anterior myometrium measuring 8x6 mm, suggestive of multiple leiomyomas. Also seen was ill defined heterogenous hyperechoic lesion measuring 9.1x7.9 mm, within the endometrial cavity showing internal vascularity, suggestive of retained products of conception (likely the cause of abnormal uterine bleeding in the patient) for which a hysteroscopic D&C was done with Histopathological diagnosis of retained products of gestation. Further in the same USG, urinary bladder and left kidney were unremarkable on USG. However, right kidney was slightly enlarged in size and showed an exophytic heterogenous echoic lesion arising from the mid pole, extending and abutting the gall bladder and liver with mild internal vascularity and measuring 74x51 mm.

Further radiological investigations were done to find the nature and extent of the lesion in the kidney. Contrast enhanced CT scan showed the exophytic mass to be arising from the anterior mid pole of the right kidney, significantly extending anteriorly with loss of fat planes with gall bladder, second part of duodenum and adjacent segment V of liver. The findings were suggestive of neoplastic etiology of right kidney (**Figure 1**).

CT renal angiography was performed which revealed similar radiological findings. Further, areas of necrosis were noted. The mass was seen bulging into the perinephric fat and extending superiorly. However, no extension was seen beyond gerota's fascia. The mass was indenting the right renal vessels with contact angle of 180 degrees. Right renal vein and IVC were normal. Ipsilateral adrenal gland was also unremarkable (**Figure 2**).

Surgical management

The patient was planned for laparoscopic radical nephrectomy of right kidney under general anesthesia. Intra operative findings revealed right kidney to be grossly enlarged involving the upper and mid pole and adherent to 2nd part of duodenum. One renal artery and 2 renal veins were clipped and divided. No liver, visceral and peritoneal metastasis were seen. The specimen was sent for histopathological examination.

Histopathological examination

On gross examination, the right kidney measured 11x10x5 cm and weighed approximately 500gm. A mass was identified involving the upper and middle pole of the kidney and measuring 7x6x5cm. Capsular breach was identified, and the mass was seen extending to perinephric fat. The mass macroscopically seemed

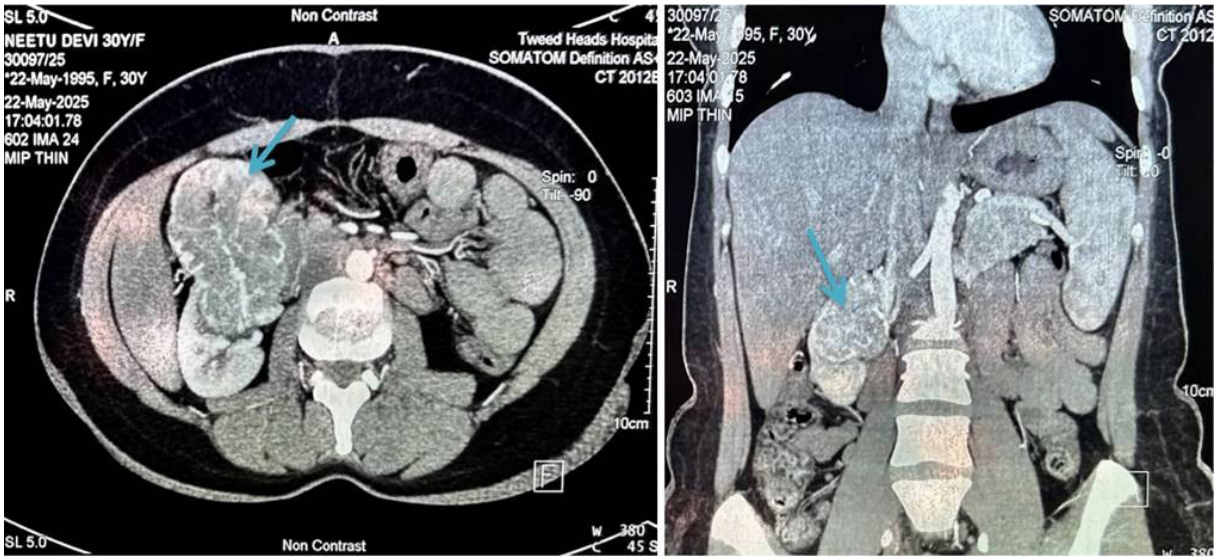

Figure 1. CECT Abdomen – An approximate 75 x 54 mm size exophytic enhancing soft tissue mass lesion is seen arising from the anterior mid pole of right kidney, significantly extending anteriorly and having lost fat plane with gall bladder, 2nd part of duodenum and adjacent segment V of liver. Findings were suggestive of neoplastic aetiology of right kidney. Another 16 x 12mm size soft tissue lesion is seen at mid pole of right kidney just above the large lesion likely neoplastic aetiology.

to arise from renal pelvis. Hilar structures could not be identified. On serial sectioning, corticomedullary junction was identified. Tumor on serial sectioning appeared solid white in appearance with hemorrhagic areas. On microscopic examination, the tumor was composed of plump spindle cells arranged in bundles and long intersecting fascicles, with individual cells having oval to elongated, blunt-ended nuclei and moderate to abundant pale to eosinophilic cytoplasm, showing moderate to marked nuclear pleomorphism. Mitotic figures including atypical ones were numerous (**Figure 3**). Tumor necrosis was seen involving <50% of the total tumor area. Lymphatic or vascular invasion were not identified. Margins of resection were free of tumor. On immunohistochemistry, the tumor cells were positive for vimentin, desmin and smooth muscle actin and negative for CK7, PanCK, MyoD1, S100, p53, CD10, HMB-45 and EMA (**Figure 4**). Histopathological reporting was done according to CAP cancer templates [7]. Based on the morphological and immunohistochemical profile, a diagnosis of Well-differentiated leiomyosarcoma was made. The tumor was involving the upper and middle pole of right total nephrectomy specimen and was involving the perinephric fat and reaching upto the gerota's fascia. The pelvic calyceal system was seen involved by the tumor, however renal sinus was free of tumor. Final diagnosis was given as Leiomyosarcoma, FNCLCC grade 1, of right kidney with pTNM staging as pT3aNxMx. No regional lymph nodes were submitted.

Follow up

Post op period was uneventful. Urinary catheter was removed on post op day 1 and drain was removed on post op day 2. At present, the patient has been followed up for 3 months, during which she has remained asymptomatic with no clinical or radiological evidence of recurrence or metastasis. The patient has been counselled regarding the importance of regular clinical review and imaging, and she has been enrolled in our institutional sarcoma follow-up program. The planned protocol includes physical examination and abdominal imaging (ultrasound or CT) every 6 months for the first 2–3 years, in addition to annual chest imaging to monitor for pulmonary metastasis. After this period, surveillance will be continued on an annual basis or sooner if new symptoms develop.

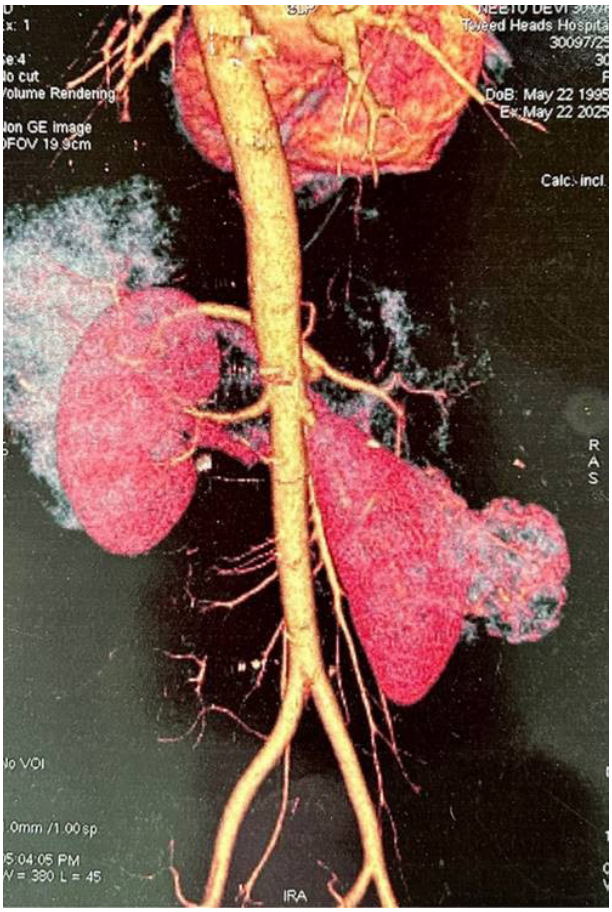

Figure 2. CT renal angiography – Large well defined smoothly marginated heterogenously enhancing exophytic mass is seen arising from mid pole of right kidney measuring "7.4 x 5.1 cm" with necrotic areas(white arrow). No calcification is seen. No fat density is seen. It is bulging into perinephric fat and extending superiorly. No extension seen beyond Gerota's fascia. Right renal vein and IVC are normal. Ipsilateral left adrenal gland is normal. It is abutting inferior surface of right lobe. It is indenting right renal vessels with contact angle of 180°.

Discussion

Renal cancers represent about 1.3% of all cancers in India in accordance with the GLOBOCAN 2020 data [8, 9]. Renal cell carcinoma is the most common renal malignancy and accounts for 85% of all kidney cancers [10]. Mesenchymal tumors constitute only about 4.5% of all adult primary renal tumors [3]. Within adult mesenchymal tumors, 5 entities have been included in the current WHO classification: PEComa of kidney, Epithelioid angiomyolipoma, Renal hemangioblastoma, juxta glomerular cell tumor and Renomedullary interstitial cell tumorl. Leiomyosarcomas of renal origin are rare and account for just <1.5% of all primary renal malignancies [4-6]. A systemic review and meta-analysis done by Periasamy et all1, involving Pubmed and Embase databases from inception to March 2023, revealed only 85 publications of primary renal LMS.

LMS are malignant tumors of smooth muscle origin. Within kidney, LMS usually arise from smooth muscle of inner layer of renal capsule, or smooth muscle of intrarenal renal blood vessels or the renal pelvis [11, 12]. Predisposing or etiological factors of renal LMS are largely unknown.

Most cases of renal LMS present with late, non-specific symptoms in middle-aged or older adults [12-15]. In contrast, our patient was incidentally diagnosed during evaluation for incomplete abortion, with no renal or abdominal complaints, and was only 37 years old. These two aspects, atypical clinical context and relatively young age - make this case clinically significant. They broaden the spectrum of how renal LMS may first come to attention and emphasize that vigilance beyond typical urologic presentations is warranted.

Renal LMS, like LMS in general, shows a strong female predilection [12-14] and typically presents between the 4Th and 6Th decade of life with a mean age of 58.5 [12, 15]. In the largest systematic review and individual patient data analysis to date by Periasamy et all1, only 85 published cases could be identified. 52.7% patients were female and median age was 55.5 years. In the case reported by Sood et all [6], the patient was 51-year-old female. Our patient, by contrast, was 37 years old, placing her more than a decade younger than the median. This difference in age highlights an important clinical outlier and broadens the spectrum of expected demographics for renal LMS.

There are no specific presenting symptoms to differentiate renal LMS from other renal malignancies. The presenting symptoms usually occur late in the course of disease, like RCC and can be

varied, such as hematuria, abdominal pain, abdominal distention and an abdominal mass [14]. In the study by Periasamy et al. [11], the most common presenting complaint was pain, followed by palpable mass and hematuria, while in the case reported by Sood et al [16], the patient presented with abdominal pain and swelling in left flank. In the present case there was no renal or abdominal symptoms, and the renal mass was incidentally discovered on imaging studies for patient presenting with history of incomplete abortion which was confirmed on D&C.

Unlike RCC and other renal neoplasms which have characteristic radiological findings, renal LMS does not have any known specific imaging characteristics [17, 18]. Radiological investigations such as CECT, CT renal angiography and USG are important in evaluating the location and extent of renal LMS, as in other malignancies. RCC is one of the most important differential diagnosis of renal LMS, but cannot be reliably differentiated by any imaging technique. Thus, CT guided core needle biopsy is recommended for obtaining a histopathological diagnosis and planning an appropriate treatment [13, 19].

Histomorphological features of renal LMS are similar to LMS elsewhere in the body and show intersecting bundles of spindle shaped cells with blunt ended nuclei and moderately abundant eosinophilic cytoplasm. The cells typically show cellular pleomorphism; atypical mitosis and tumor necrosis can be seen. Pre-operative imaging could not reliably distinguish LMS from mimics such as RCC (including sarcomatoid change) or angiomyolipoma. Histology demonstrated a high-grade-appearing spindle neoplasm; therefore, a broad IHC panel was used. Diffuse SMA/desmin/vimentin positivity confirmed smooth-muscle differentiation. Lack of epithelial markers (CK7, Pan-CK, EMA) argued against RCC/sarcomatoid carcinoma; absence of HMB-45 excluded PEComa/angiomyolipoma; S100 negativity made malignant peripheral nerve sheath tumor unlikely; MyoD1 negativity excluded skeletal-muscle lineage. Together with morphology, these findings established primary renal LMS. Grading of LMS is done according to the French federation of cancer center sarcoma group (FNCLCC) [8] with the grading based on three parameters: tumor differentiation, mitotic count and tumor necrosis. While our case showed classic leiomyosarcoma morphology with spindle cells, fascicular arrangement, pleomorphism, and atypical mitoses, the mitotic index did not reach the threshold for grade 2, and necrosis was <50%. Thus, despite perinephric fat invasion (pT3a stage), the histological parameters resulted in a grade 1 tumor. This discordance low histologic grade despite advanced anatomic stage is rarely reported. It underscores the need to consider both grade and stage when prognosticating, as stage reflects local extent, while grade predicts biological aggressiveness. In our case, the tumor was grade 1, however majority of primary renal LMS have been reported to be high grade ie grade 2 or 3 [12, 19].

In Periasamy et al.'s [11] series, most tumors were classified as FNCLCC grade 2 or 3. Similarly, in other reported series including those of Miller et al [12] and Deyrup et al [20], high-grade morphology was predominant, correlating with the aggressive clinical course and poor prognosis typically described for renal LMS. In contrast, our patient's tumor was discovered incidentally

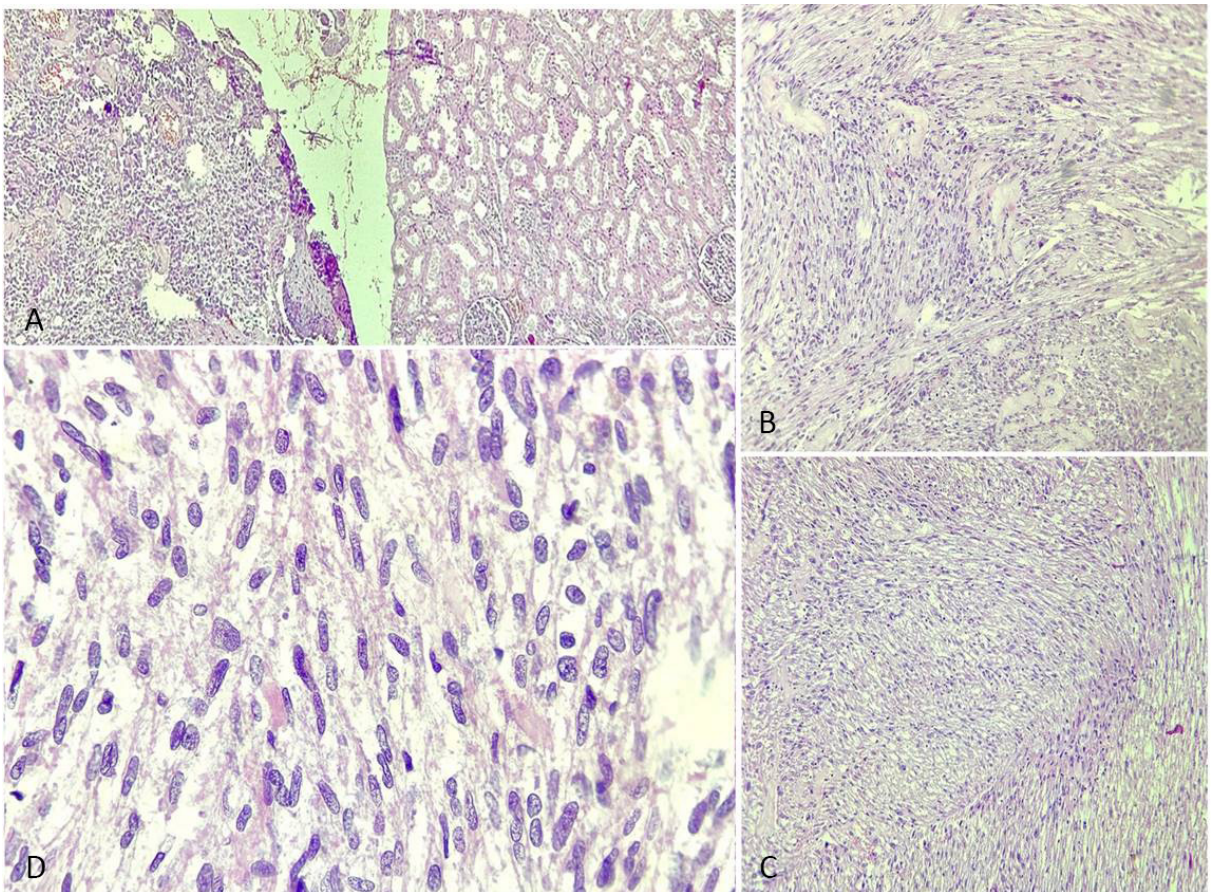

Figure 3. Renal leiomyosarcoma (H&E stained sections). (a) Tumor cells (black arrow) in fascicles adjacent to normal renal tissue (blue arrow), H&E, 4×; (b-c) Fascicles of spindle cells (black arrow) with oval to elongated, blunt-ended nuclei and moderate eosinophilic cytoplasm, H&E, 10×; (d) High power showing marked nuclear pleomorphism and atypical mitoses (black arrow), H&E, 40×.

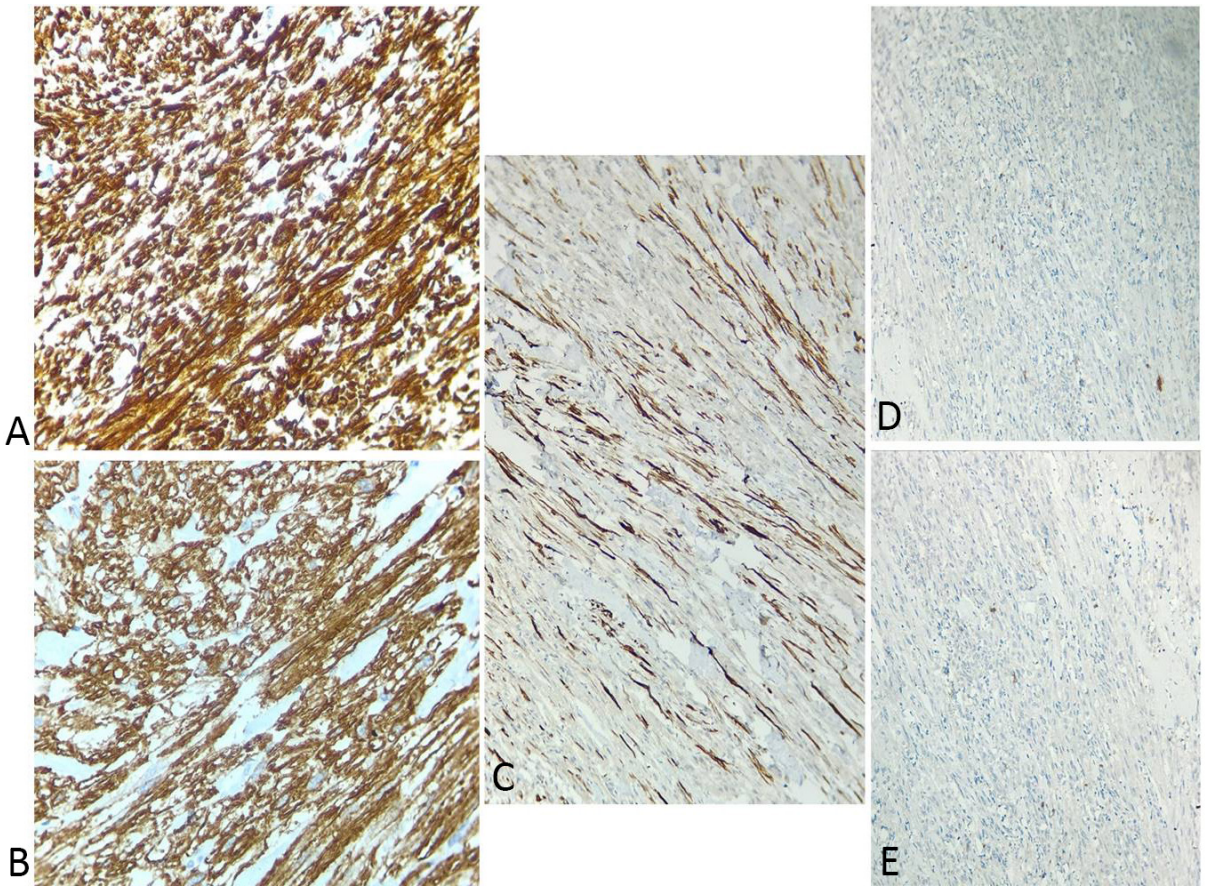

Figure 4. Immunohistochemistry of renal leiomyosarcoma. (a) Strong cytoplasmic positivity for Vimentin (clone V9, Dako), 40×; (b) Diffuse cytoplasmic positivity for SMA (clone 1A4, Dako), 40×; (c) Cytoplasmic positivity for Desmin (clone D33, Dako), 40×; (d) Tumor cells negative for MyoD1 (clone 5.8A, Dako), 10×; (e) Tumor cells negative for Pan-CK (clone AE1/AE3, Dako), 10×.

during evaluation for incomplete abortion, without any renal or abdominal complaints, and histologically graded as FNCLCC grade 1. This unusual combination of low grade and incidental discovery in a young female differentiates our case from the majority of previously published reports.

Due to scarcity of data on primary renal LMS, there are no specific treatment protocols dictated by randomized control trials. The currently accepted treatment protocol is complete surgical excision of the tumor for resectable tumors with negative margins. Radical nephrectomy is a preferred treatment option [21]. Neoadjuvant and postoperative chemotherapy and radiotherapy are acceptable and can be given in cases with intermediate or high-grade tumors, large tumors or when there is a high risk of distant metastases [18].

Renal LMS is associated with a poor prognosis with a median overall survival of 25 months [18]. Prognostic factors include stage, histological subtype, grade, age of the patient and gender. According to Deyrup et al. increasing grade of renal LMS and survival is inter-related [20].

Distant metastases to sites such as liver, lungs, bones and soft tissue were reported in 30% of the patients in the case series by Novak M et al [13]. Other complication which can be observed is vascular extension such as tumor thrombus to renal vein with presence with extension to intrahepatic portion of inferior vena cava as seen in the case by Srinivasan et al [22]. Patients can present to the emergency as well with spontaneous rupture [23].

Differentials of primary renal LMS that should be considered while making the final diagnosis include: Retroperitoneal LMS with extension to kidney, Sarcomatoid carcinoma of kidney, Angiomyolipoma and Renal leiomyomas. Retroperitoneal LMS can be ruled out using radiological techniques. Sarcomatoid carcinoma of kidney on histopathology will show a malignant epithelial component which be absent in LMS. Further, in sarcomatoid carcinoma, on IHC, spindle cell component will be positive for CK and negative for SMA while tumor cells of LMS will be positive for SMA and negative for CK. Angiomylipoma, on microscopy, shows mature adipose tissue with thick hyalinized vessels on histology with cells showing positivity for HMB-45, which will not be seen in LMS. Renal leiomyomas unlike LMS, will not show atypia, mitosis or tumor necrosis on microscopy.

Given the aggressive nature of renal leiomyosarcoma and its known tendency for local recurrence and distant spread to sites such as the lungs, liver, bone, and soft tissue, short-term follow-up alone is insufficient. Long-term surveillance is therefore essential. The structured follow-up as described in the case presentation section aims to detect recurrence at an early stage, when curative or palliative interventions may still be feasible, and reflects the broader consensus on monitoring patients with renal and other visceral sarcomas.

Conclusion

Primary renal LMS is rare. There is often delay in diagnosis due to non-specificity of symptoms and radiological findings.

Recommended treatment option is radical nephrectomy with negative margins. Renal LMS carries poor prognosis with a shortened period of survival, making it crucial to be considered as a differential while dealing with spindle cell tumors of the kidney.

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No applicable.

Ethical policy

The investigation was conducted in accordance with the Declaration of Helsinki of 1975. Written informed consent was taken from the patient.

Availability of data and materials

Data sharing is not applicable to this article as no new data were created or analysed in this study.

Author contributions

Rashi Maheshwari: Data Curation, Visualization, Writing - Original Draft; Shikhar Chohan: Data Curation, Visualization, Writing - Original Draft; Ayushi Aggarwal: Data Curation, Visualization; Sufian Zaheer: Conceptualization, Data Curation, Validation, Formal analysis, Writing - Review & Editing, Visualization.

Competing interests

The authors declare that they have no competing interests.

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