

Primary renal leiomyosarcoma: A rare case report

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ABSTRACT

Leiomyosarcoma of the kidney is a rare type of adult renal sarcoma. Here, we presented a case of a 68-year-old female who had a past history of bilateral renal calculi 3 years back with hydronephrotic changes, now presented to the OPD with complaints of abdominal pain for the past few months. On further evaluation, a heterogeneously enhancing necrotic mass was identified in the right kidney measuring 7.5 cm in the greatest dimension. A right radical nephrectomy with left Double J stenting was done. From histopathological and immunohistochemical staining, it was diagnosed as leiomyosarcoma.

Key words: Renal calculi, Renal, Sarcoma

Renal leiomyosarcomas are the most common type of sarcomas, accounting for 50–60% of all the sarcomas arising in the kidney [1]. It accounts for <1% of all renal malignancies accounting for only 0.12% of renal malignancies [2,3]. There are no known predisposing factors. It is also known to preferentially affect females in their 60's [4].

Here, we report a rare case of renal leiomyosarcoma in a 68-year-old female and also reviewed the relevant literature below.

CASE REPORT

A 68-year-old lady presented to the OPD with complaints of mild to moderate abdominal pain which was fluctuating in nature for the past 6 months. The patient also complained of anorexia and significant weight loss for the past 6 months.

Her physical examination showed an easily ballotable, non-tender mass, lobular in shape, and firm in consistency. 3 years earlier, she presented with bilateral renal calculi with hydronephrotic changes which passed spontaneously in urine without any intervention, with medication.

Her routine hematological examination at present showed anemia with hemoglobin of 9 g%. The biochemical investigations were all within normal limits. Urinalysis showed mild hematuria with 4–6 RBCs/hpf. Urine culture turned out to be negative but urine cytology revealed a few atypical cells suspicious for malignancy. No other hematological or biochemical abnormality was detected


except anemia. Occult blood in stool and urine were negative. On further evaluation of the patient, a heterogeneously enhancing necrotic mass was identified in the right kidney measuring 7.5 cm in greatest dimension on computed tomography scan (Fig. 1).

Subsequently, the right nephrectomy was performed with left Double J stenting. Gross examination revealed a well-defined tumor mass measuring 8 cm × 8 cm × 7 cm, with cut surface showing areas whitish whorling with focal areas of necrosis (Fig. 2).

Microscopic examination of the tumor masses revealed a malignant neoplasm composed of spindle cells arranged predominantly in fascicles and somewhere in plexiform pattern, with marked nuclear pleomorphism and prominent nucleoli with acidophilic cytoplasm (Fig. 3). Atypical mitotic activity was present. Sections from the adjacent renal parenchyma did not reveal any neoplastic component apart from changes of chronic tubulointerstitial nephritis. Major vessels showed no sign of tumor dissemination. An extensive regrossing was performed to look for any epithelial components in the tumor mass but revealed none by microscopy. Immunohistochemically, the neoplastic cells showed membrane positivity for desmin (Fig. 4). Hence, from the histomorphology and immunohistochemistry, it was diagnosed as a case of primary renal leiomyosarcoma.

DISCUSSION

Leiomyosarcoma is a malignant tumor of smooth muscle component of soft tissue. It is essentially a tumor of adults and the elderly population, but some cases have also been reported in children [5]. Apart from the uterus, soft tissue leiomyosarcoma

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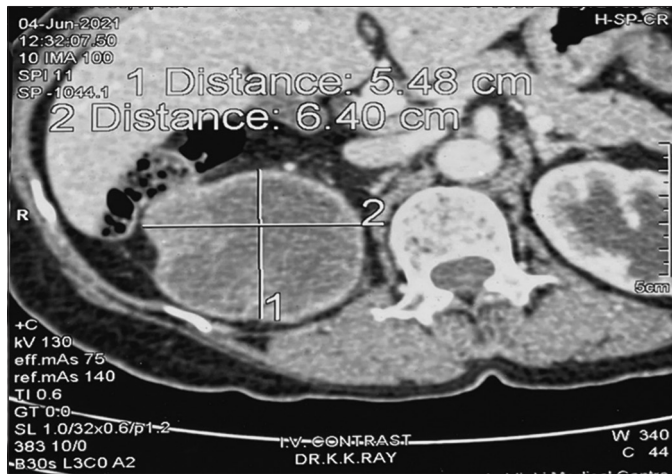


Figure 1: Computed tomography scan image showing heterogeneously enhancing necrotic mass involving whole of the right kidney

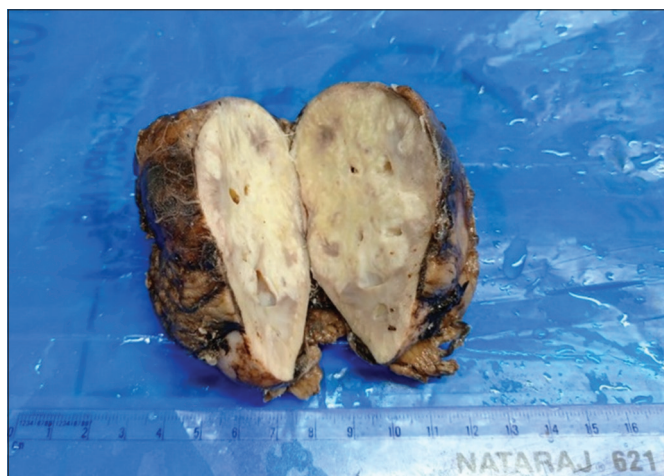


Figure 2: Gross picture of renal leiomyosarcoma

commonly occurs in the retroperitoneum and blood vessels. Leiomyosarcomas of non-peritoneal soft tissue sites usually involves the lower extremity and can also occur in the head and neck region [6]. They are most commonly found in the uterus, stomach, small intestine, and retroperitoneum [7]. Renal leiomyosarcomas are believed to originate from the renal capsule or the smooth muscle fibers in the renal pelvis or from the renal vessels [4].

The incidence of the primary renal leiomyosarcoma increases with the age of the patient. This tumor was found to be more common in females than in males and more common in the right kidney than in the left kidney according to a study conducted by Beardo *et al.* [8] Farrow *et al.* found a preponderance of renal leiomyosarcomas in females and more frequent after the fifth decade of life [9]. Furthermore, in a study performed by Dhamne *et al.*, it was found that the renal leiomyosarcomas have a female preponderance, with females being twice more commonly affected by renal leiomyosarcomas than males with most of the patients presenting in their fourth-sixth decades [10]. Renal leiomyosarcomas are found to be typically occurring between the fourth and eighth decades of life, more commonly found among women and arising from the right kidney as found

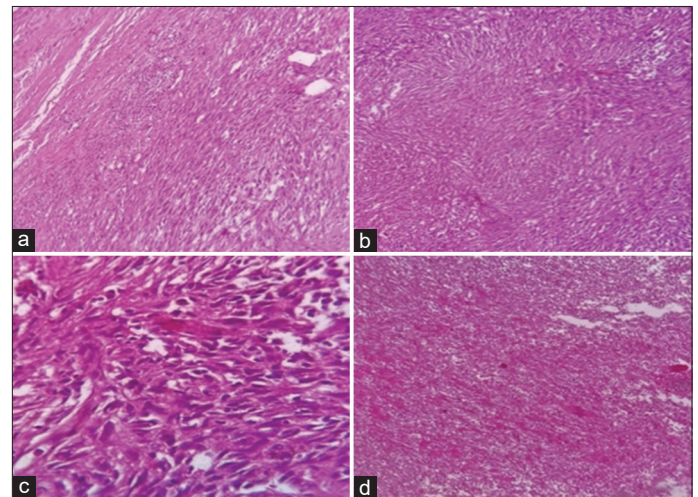


Figure 3: (a) Picture showing tumor portion with adjacent renal tubules $\times 100$ (H and E); (b) Picture showing tumor $\times 100$ (H and E); (c) Tumor composed of spindle cells with eosinophilic cytoplasm. Many atypical mitotic figures can be identified. $\times 400$ (H and E); (d) Areas of extensive necrosis adjacent to tumor cells $\times 100$ (H and E)

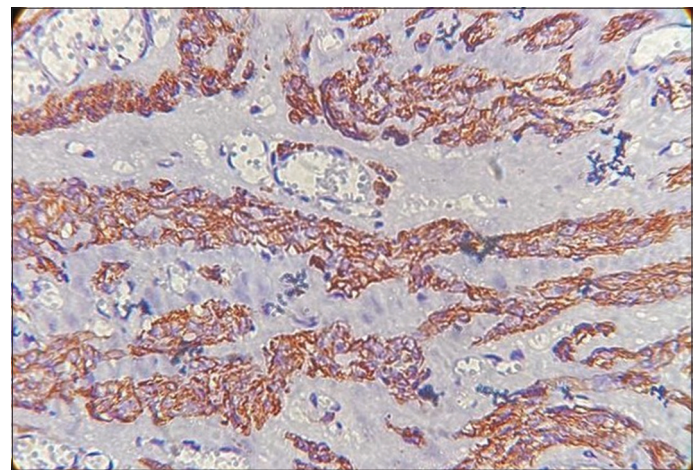


Figure 4: Membrane expression of Desmin $\times 400$

by Miller *et al.* [11] Polianko *et al.* concluded in their study that renal leiomyosarcomas can occur on both the left and right sides equally and can also occurs bilaterally; however, the etiology was not known [12]. The reason behind female preponderance is unknown, but a study performed by Brown *et al.* suggested that some malignancies are associated with genes located on the X chromosome which escape X chromosome inactivation [13].

Clinical features of renal leiomyosarcomas are very similar to more common renal malignancies which consist of hematuria, flank pain, progressive weight loss, and an abdominal mass lesion. As the tumor increases in size, it produces symptoms such as lumbar pain and hematuria and a palpable mass [8].

Grossly, the tumors look like leiomyomas with a well-circumscribed margin and whorled appearance on the cut section. However, the malignant counterpart appears fleshy and has areas of hemorrhage, necrosis, and degenerative changes [9]. Gross examination of the tumor shows evident trabeculations and whorling pattern; the combined light microscopy with immunohistochemistry gives the definitive diagnosis of

leiomyosarcoma. The sarcomatoid variant of renal cell carcinoma is the chief differential diagnosis of renal leiomyosarcoma. Other differential diagnoses which can be confused with leiomyosarcoma are the epithelioid variant of renal angiomyolipoma and renal synovial sarcoma. Hence, immunohistochemistry is an important investigation for the evaluation of such tumors to avoid the wrong diagnosis [11].

On microscopy, renal leiomyosarcomas show smooth muscle cells arranged in a fascicular pattern, with individual cells showing marked nuclear atypia and prominent mitotic figures. They show a positive expression of smooth muscle markers such as smooth muscle actin, calponin, desmin, and h-caldesmon and are negative for cytokeratin, S-100 protein, and HMB45 on immunohistochemistry. The sarcomatoid variant of renal cell carcinoma reveals foci of typical renal cell carcinoma with cytokeratin positivity while the smooth muscle markers are negative. Epithelioid angiomyolipoma shows positivity for both smooth muscle markers and melanocytic markers, while renal synovial sarcomas show positivity for smooth muscle markers and Bcl 2 [11,12].

Renal leiomyosarcomas show aggressive behavior due to rapid growth rate, frequent distant metastases, and high recurrence rates. Distant metastases have been reported to the lungs, liver, and gastrointestinal tract. Radical nephrectomy is the treatment of choice for primary renal leiomyosarcoma [10,11]. Because of the limited data, the role of adjuvant chemoradiotherapy is still in doubt [11]. The mean survival rate of renal leiomyosarcomas ranges from 6 months to 2 years after the diagnosis is made [2,14]. Kwon *et al.* reported a case of renal leiomyosarcoma in a 46-year-old Caucasian male [15]. Renal leiomyosarcomas usually show aggressive behavior, and the prognosis appears to be poor with a median survival between 17.9 and 25 months [16]. Total surgical resection is the major prognostic factor [17]. Radical nephrectomies, size of the tumor <5 cm, low histologic grade of the tumor, and negative lymph nodal status are all associated with a better prognosis in patients with leiomyosarcomas [15].

CONCLUSION

We presented a rare case of renal leiomyosarcoma in a 68-year-old female. A nodular mass with a whorled cut surface resembling a leiomyoma raises a suspicion of a smooth muscle tumor in the kidney. This tumor was sampled extensively. Histopathological

examination alone is insufficient to give a definite diagnosis. Immunohistochemistry is essential to prove the smooth muscle cell origin of the tumor and also for differentiating it from the sarcomatoid variant of renal cell carcinoma.

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