

Primary leiomyosarcoma of kidney in a young adult

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ABSTRACT

Primary renal leiomyosarcoma (LMS) is an extremely rare tumor constituting to <2% of all malignant renal tumors. Here, we present the case of a 28-year-old female with a right lower polar renal tumor, who underwent partial nephrectomy which on microscopy suggested LMS. Due to the lack of a follow-up protocol and low survival rates, regular imaging and a multidisciplinary approach were considered in the patient.

Key words: Kidney, Leiomyosarcoma, Partial nephrectomy, Young adult

Primary sarcomas of the kidney represent 1–2% of all malignant tumors of kidney [1] and leiomyosarcoma (LMS) accounts for 60–70% of all sarcomas of the kidney. Females are more commonly affected usually in the 4th–6th decades [2]. Flank pain, palpable mass, and hematuria constitute the major symptoms with the right side being more frequently affected [3]. A radical nephrectomy with wide margins is the treatment of choice for LMS of the kidney. However, the literature also supports treatment with partial nephrectomy in selected cases [4]. Here, we present the case of a 28-year-old female with primary LMS of the right kidney who underwent partial nephrectomy with a relevant review of the literature.

CASE REPORT

A 28-year-old female with no comorbidities reported to our department with chief complaints of intermittent, right-sided flank pain of 3 months duration. There was no history of any hematuria, gravelluria, or any altered bowel habits. On general physical examination, she was an average built, moderately nourished and was vitally stable. There was no palpable mass on per abdomen examination.

Her routine blood investigations were within normal limit. Contrast-enhanced computed tomography scan of the whole abdomen revealed a 3.6 cm × 3.2 cm × 2.8 cm mass in the lower pole of the right kidney with post-contrast enhancement and an R.E.N.A.L nephrometry score of 5a (Fig. 1).

She underwent partial nephrectomy. Postoperatively, the gross examination of the specimen revealed a globular mass 4 cm × 2 cm with infiltration of renal parenchyma and without the infiltration of the renal pelvis. All margins were free from tumor. The cut

section revealed a fleshy tan mass with areas of necrosis and hemorrhage. On microscopic examination, the sections showed a well-encapsulated tumor composed of pleomorphic spindle-shaped cells arranged in intersecting fascicles and showing whorled pattern at places with a mitotic count of 18–20/hpf (Fig. 2).

Immunohistochemistry was positive for desmin, vimentin, and smooth muscle actin (SMA) and negative for S100, cytokeratin (CK), CD99, CD56, and calretinin suggestive of leiomyosarcoma. The patient has undergone a whole body positron emission tomography scan at 3 months interval and is free of any residual or recurrent tumor until 1 year of follow-up. The patient is under close follow-up in oncology and urology department.

DISCUSSION

LMS is a rare tumor that arises most commonly from smooth muscle of soft tissues and uterine tissue. In a majority of the cases, renal LMS arises from smooth muscle of renal vessels but can also arise from the renal pelvis and renal capsule [5]. The incidence of the primary LMS of the kidney increases with age, more commonly affecting females and the right kidney [5].

Renal LMS is less common but more lethal than LMS of other genitourinary sites such as prostate, bladder, and paratesticular region [6]. The symptoms increase with the advancing disease stage. The pressure effect results in flank pain and hematuria and it sometimes presents with a palpable mass. Nausea, vomiting, and abdominal pain may occur as a part of systemic symptoms [5]. It is difficult to differentiate LMS of the kidney from renal cell carcinoma (RCC) as they exhibit similar clinical, radiological, and pathological features.

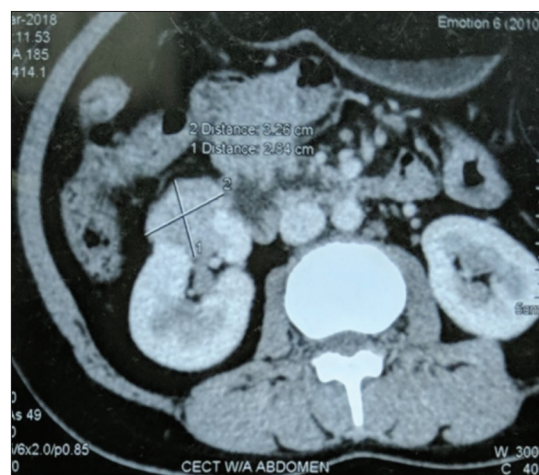


Figure 1: A well-defined, round iso to hyperdense (HU - 45) SOL in lower pole of the right kidney 3.26 cm × 2.84 cm

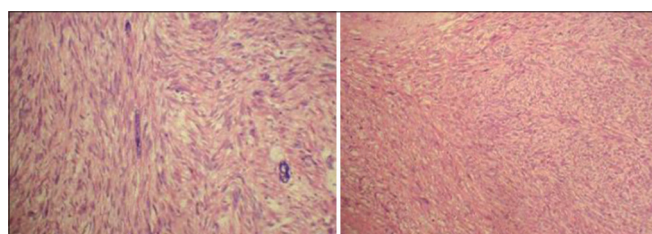


Figure 2: Microscopic image of the kidney tumor composed predominantly of spindle cells with bizarre scattered cells, arranged in whorls, interlacing fascicles, and bundles

The differentiating features being size, site of origin, and lymph nodal involvement. Renal LMS tends to displace rather than invade the renal parenchyma and has a rapid growth rate. They also tend to have frequent metastasis and high local and systemic recurrence rates [7].

Sarcomatoid RCC forms the closest differential diagnosis of renal LMS [8]. In case of fast-growing masses, sarcomas should always be considered. The sarcomas possess a pseudocapsule which is often found to be infiltrated by the tumor. On histopathology, thorough sampling for an absence of any epithelial component should be done to rule out sarcomatoid RCC. On immunohistochemistry, LMS tumor cells are positive for desmin, SMA calponin, H-caldesmon, and negative for CK, S-100, and HMB-45. The angiomyolipoma shows positivity for HMB-45 while CK positivity is seen in a sarcomatoid variant of RCC [9].

In a study done by Lewis *et al.*, unresectable disease and positive surgical margins were reported as the most significant factors predictive of disease-specific mortality [10]. The initial resection tends to be the key and gives the best chance for a long-term cure [11]. Radical nephrectomy with wide margins is recommended in patients with renal LMS. However, partial nephrectomy has also been given as the form of treatment in the literature [4,12].

According to the statistical records of the National Cancer Institute and American Joint Committee on Cancer, only 50% of the patients live longer than 5 years. With increasing grade, this

survival rate drops further. In general, it is seen that there is no survival advantage of neoadjuvant or adjuvant chemoradiation in LMS [3,6]. However, in a study, adjuvant chemotherapy has been seen to be beneficial in partially resected tumors [13]. Kamba *et al.* showed that neoadjuvant chemotherapy with (cyclophosphamide, vincristine, adriamycin, and dacarbazine) can cause downstaging of the tumor and increases the possibility of complete resection [14].

Deyrup *et al.* demonstrated that increasing histological grade was a poor prognostic factor [7]. Primary LMS of the kidney is associated with significantly poor prognosis as compared to other renal malignancies. A detailed morphological and immunohistochemical analysis is required for its diagnosis and management [15]. To accomplish better oncologic control, neoadjuvant and adjuvant treatment modalities should be considered. As no follow-up protocol has been devised due to the paucity of cases, stringent imaging should be considered for follow-up in such cases, and the patients with renal LMS should be referred for multidisciplinary management.

CONCLUSION

Primary LMS of the kidney is rare, but an important tumor that may present similarly to other renal malignancies. A high index of suspicion should be maintained due to the non-specific symptoms. Although radical nephrectomy is the gold standard treatment for renal LMS, our patient being treated with partial nephrectomy is also supported in some reports.

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