Leiomyosarcoma of Kidney: A Rare Case Report

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ABSTRACT

Published on 30th June 2016

Primary sarcomas are very rare aggressive tumours of the kidney. We report a case of a 57 year old lady presenting with right loin pain and anorexia for 3 months. On examination she had a nontender mass which on radiological evaluation proved to be a large heterogeneous renal mass with central necrosis. Right radical nephrectomy was done. Histological examination revealed a neoplasm with cells arranged in fascicles and sheets and a herring bone pattern. Immunohistochemistry confirmed a diagnosis of primary leiomyosarcoma kidney. The patient is being followed up; the case is being reported for its rarity.

Keywords: Loin mass, Primary leiomyosarcoma kidney

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INTRODUCTION

Primary sarcomas constitutes from 0.8- 2.7% of renal tumors in adults.[1,2] Of these leiomyosarcomas constitute 50-60% of all renal sarcomas.[3,4] Though rare, but they are aggressive tumour arising from renal capsule, renal vein, pelvic musculature, or renal parenchyma. Mean age at diagnosis is in sixth decade. Usual clinical presentation mimics renal cell carcinoma with classical triad i.e. flank pain, hematuria & abdominal mass.[5] Currently available imaging systems are unable to differentiate between leiomyosarcoma & RCC, so diagnosis is usually made postoperatively with histology and immunohistochemistry. We hereby represent a case of 57 years old female diagnosed to have leiomyosarcoma. To the best of our knowledge it is one of the few reported cases from Indian subcontinent.

CASE REPORT

A 57 year old woman presented with pain in right loin region and anorexia for 3 months. There was no history of hematuria, bladder and bowel disturbances. On clinical examination she was pale looking with a non-tender ballotable mass in right loin region. She was known hypertensive and diabetic for 8 years and was on regular treatment for the same.

On routine investigation her hemoglobin was found to be 7.7gm%. Urine analysis revealed no abnormality, renal function tests were in normal range. CT scan showed a large heterogeneous mass of 20x 20x 17 cm arising from interpolar & lower polar region of right kidney extending exophytically with curvilinear broken calcifications. Internal density ranged from 30-35 HU with moderate post contrast peripheral enhancement (60 HU) with poorly enhancing central necrotic areas. There was no invasion or thrombosis of renal vein or IVC **(Figure 1).**

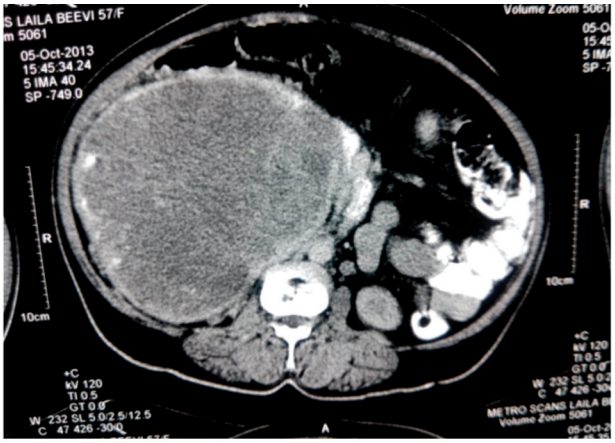

Figure 1. **CECT Abdomen- Tumor Originating from RT Kidney**

Right radical nephrectomy was performed and mass was delivered in Toto. On gross examination the mass was 22X15X10 cm in size weighing 2500gms. Cut section showed ill-defined circumscribed mass of variegated appearance with extensive areas of hemorrhage and necrosis with few viable areas. Sinus fat showed involvement by the tumour **(Figure 2).**

Cite this article as: Vasudevan S, Kumar V, Krishna B. Leiomyosarcoma of Kidney: A Rare Case Report. Kerala Medical Journal. 2016 Jun 30;9(2):73–5.

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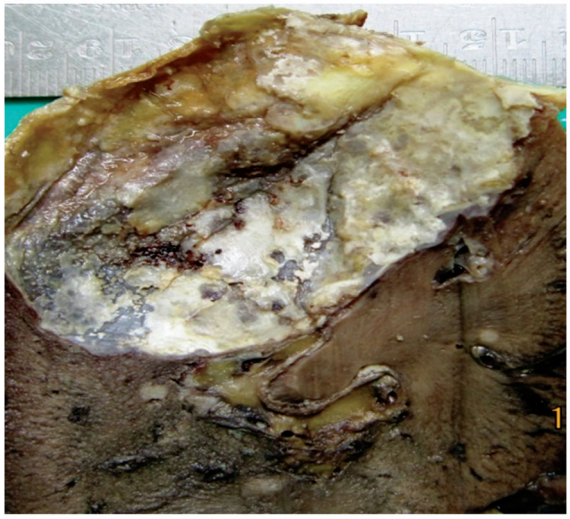

Figure 2. **Gross image of Tumour Mass**

Histopathological examination revealed a neoplasm composed of cells arranged in fascicles and sheets and herring bone pattern. Individual cells were spindly with moderate amount of eosinophilic cytoplasm, pleomorphic elongated vesicular nuclei with coarse chromatin. Many bizarre cells with abundant cytoplasm and irregular nuclei were seen. Atypical mitotic figures were noted. Mitotic rate was <10/10HPF. Area of hyalinization, myxoid changes and necrosis were noted. Neoplasm was seen to involve intrarenal sinus fat and perinephric fat.

Sections were further stained for immunohistochemical markers to confirm the diagnosis. It was negative for CK and positive for vimentin and SMA **(Figure 3).**

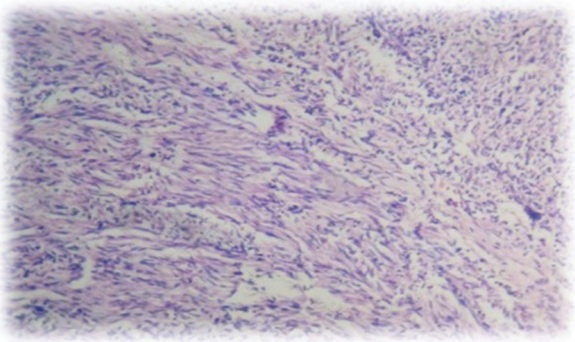

Figure 3. **IHC-Showing Negativity for Cytokeratin**

DISCUSSION

Renal leiomyosarcomas are a rare entity constituting 0.8-2.7% of malignant renal tumors in adulthood. They arise from smooth muscle cells. Definite diagnosis can be made only on microscopy with IHC as mentioned in the above case. Leiomyosarcomas commonly arise from uterus, stomach, intestine, retroperitoneum and also blood vessels. Of nonperitoneal soft tissue sites it usually arises from lower extremities but can also arise from head and neck regions. Leiomyosarcomas usually arise from capsule, smooth muscles, blood vessels and hilum of the kidney.[4,8] Mean age of presentation is fifth or sixth decade of life.[9] with female predominance. But cases have been reported in neonatal period. Renal leiomyosarcomas secondary to metastasis is rare which commonly appear as intra parenchymal lesion or diagnosed on microscopy. Renal leiomyosarcomas are bulky tumors that replace and invade the renal tissue as seen in this case. Local recurrence is common mainly when tumor is not well-circumscribed and has extended beyond the kidney. Common site of metastasis are lung, liver and regional lymph nodes if the tumor is not confined to kidney. Spontaneous rupture of sarcoma is rare which is common in angiomyolipomas and occasionally in renal cell carcinomas and Wilms tumor.[7] Common findings in CT scan and angiography in case of renal sarcomas include: 1. Tumors arise from renal capsule or renal sinus, 2. Mass usually confined to the kidney 3.Vascular pattern of the tumor. Renal leiomyosarcomas show tortuous tumor vessels without pooling of contrast material or arteriovenous shunting.

Grossly the tumors appear like leiomyoma with well-defined margins and whorled cut surface. The malignant variety however appears fleshy and has areas of hemorrhage, necrosis and cystic degeneration.[10]

Microscopically, they represent features of smooth muscle tumor with alternating fascicles of spindle shaped cells. The cells have eosinophilic cytoplasm and blunt ended nuclei. Necrosis, nuclear pleomorphism and mitotic figures are suggestive of malignancy.

Sarcomatoid variant of RCC, epitheloid variant of angiomyolipoma, renal synovial sarcoma are differential diagnosis of renal leiomyosarcomas. For evaluation of such tumors immunohistochemistry is mandatory.[8] Sarcomatoid variant of RCC exhibits typical foci of renal cell carcinoma and positivity for cytokeratin but negativity for smooth muscle actin. Epitheloid variant of angiomyolipoma is negative for cytokeratin but positive for smooth muscle and melanocytic markers. Renal synovial sarcomas exhibit positivity for Bcl2 and smooth muscle markers.

Radical nephrectomy is the treatment of choice for renal leiomyosarcomas.[1,2] Renal leiomyosarcomas have shown aggressive nature due to rapid growth rate, frequent metastasis and local as well as systemic recurrences. Common sites of metastasis are lung, liver and colon which indicates unfavorable prognosis. Adjuvant

Chemotherapy and radiotherapy are recommended due to aggressive nature of tumor. However, the role is still questionable due to lack of data on this rare renal malignant tumor. Mean survival rate varies from 6 months to 2 years after diagnosis.

CONCLUSION

Though rare, possibility of renal leiomyosarcomas should always be kept while evaluating a well- defined mass with whorled cut surface of smooth muscle tumor in the kidney especially in older females due to their aggressive nature and distant metastasis. Final diagnosis can only be made by microscopic examination with immunohistochemistry in case of renal leiomyosarcomas.

END NOTE

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Conflict of Interest: All the authors state that there is no conflicts of interest pertaining to the study.

Editor's Remarks: The case reported is of rare occurrence. It is of value to understand the pathological varieties of renal tumours seen.

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