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Primary Leiomyosarcoma of the Ureter, a case report

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Abstract: *Around 95% of tumors of ureter are primitive epithelial tumors. Most of them are of transitional cell origin. Malignant tumors of smooth muscle of the ureter are extremely rare and about 20 cases of leiomyosarcoma of ureter have been reported till date.*
Key words: *leiomyosarcoma, ureter, hydroureteronephrosis*

I. Introduction

We report a case of a 48yr old woman, with a complaint of continuous dull aching pain and a lump in left upper abdomen from past 3 months. Transabdominal sonography, CECT abdomen and IVU revealed a lump close to lower pole of left kidney with obstruction of left ureter with hydroureteronephrosis. CT guided biopsy of the tumor suggested a malignant tumor. Tumor was excised with safe margin of ureter and ureteroureterostomy was performed after mobilization of left kidney and lower segment of ureter. Histopathology and immunohistochemistry test revealed it to be leiomyosarcoma.

II. Case report

A 48 year old woman reported to our hospital with complaints of a lump in left upper abdomen and a continuous dull aching pain in left flank for past three months. She had no history of gross hematuria or urinary symptoms. No history of kidney stones or urinary tract infection. On clinical examination an oval swelling of 7x6 cm was palpable in the left lumbar area extending to left subcostal region. [Fig. 1] Surface was irregular, lobulated. Tender on deep palpation. It was a retroperitoneal solid swelling. Left renal angle was free. Lump was not ballotable. Renal parameters were normal.

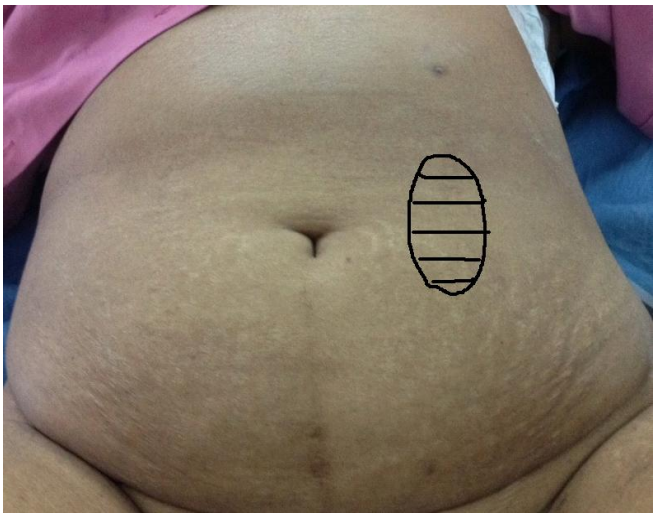

Fig. 1

An ultrasound revealed left sided hydroureteronephrosis with proximal ureterectasis and a multilobulated retroperitoneal mass lesion in the left para aortic area. [Fig. 2a,b]

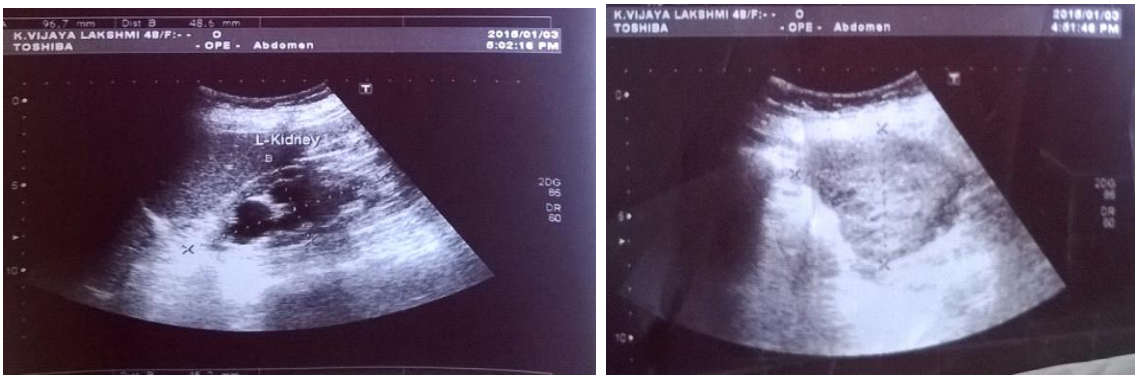

Fig. 2a,b

Plain computed tomography showed a lobulated solid mass (7.5x5.3x7.3cm) in the retroperitoneum on left side infiltrating psoas muscle and exophytic left renal pelvis. Left sided hydronephrosis with parenchymal thinning (8mm) and dilatation of proximal ureter. A contrast study showed heterogeneous enhancement.
CT guided FNAC was nonconclusive, so a CT guided biopsy was done which revealed features of malignancy.

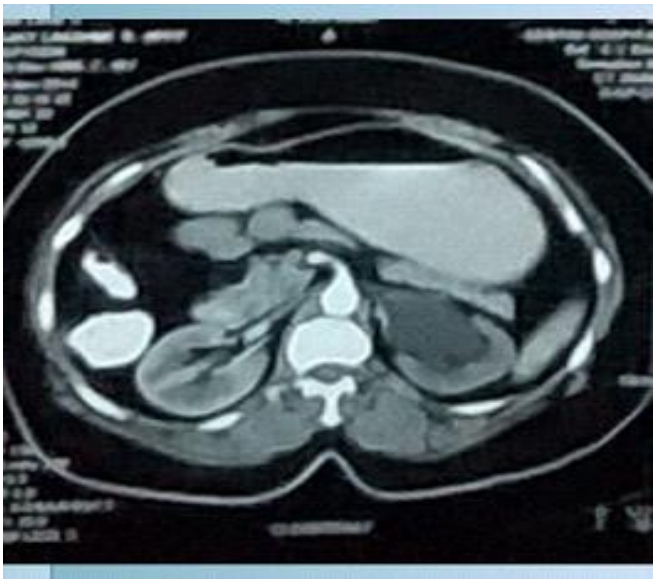

Fig.3

MRI of mid abdomen and spine showed heterogenous well defined mass lesion in the mid lumbar region in the retroperitoneum on left side obstructing the lumen of the left ureter. Medial and posterior wall of the ureter was involved in the affected segment. There was no obvious extension into the spinal canal. Fig.4a, b

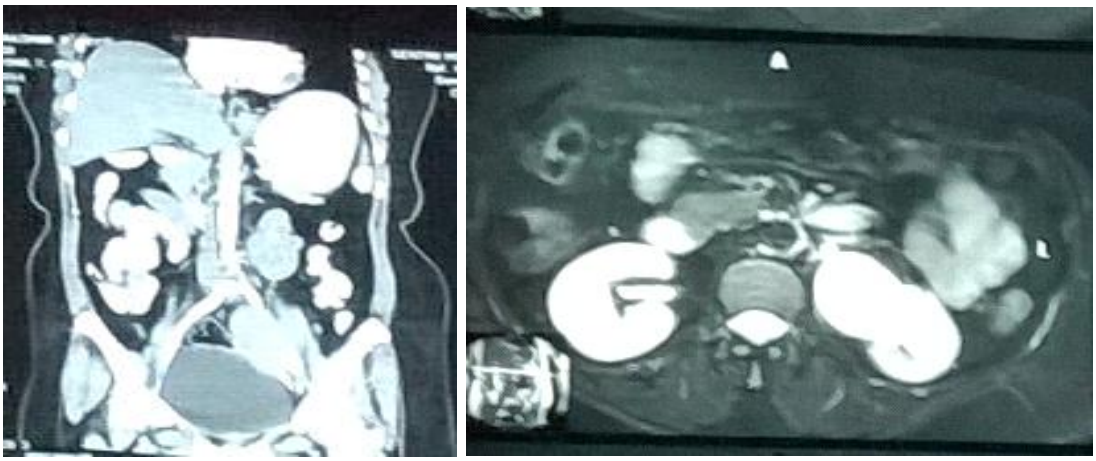

Fig.4a, b

Intravenous urogram

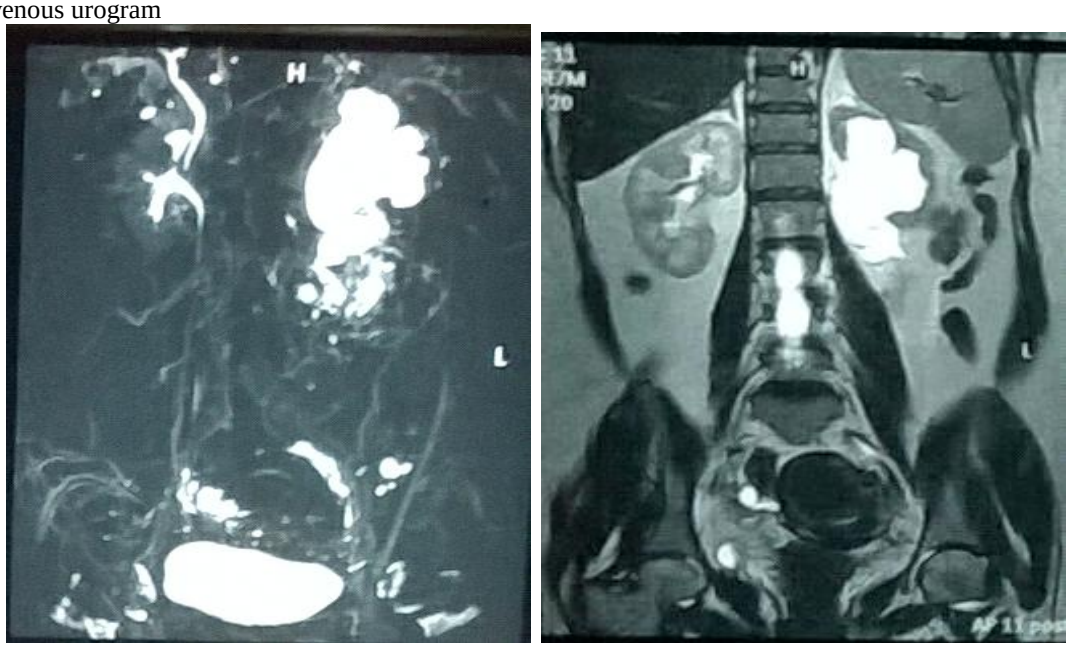

Fig.5a, b

A cystoscopic guided Dj stent placement was done in the left ureter by gentle manipulation before the laparotomy. A midline incision made and dissection carried out by extra peritoneal approach. Left ureter found passing through the center of the mass. A firm tumor mass of 8x6x5 cm was excised after identifying ureter at its upper and lower poles and a ureteroureterostomy was performed over DJ stent after mobilization of left kidney and lower segment of the ureter. Fig.6a, b

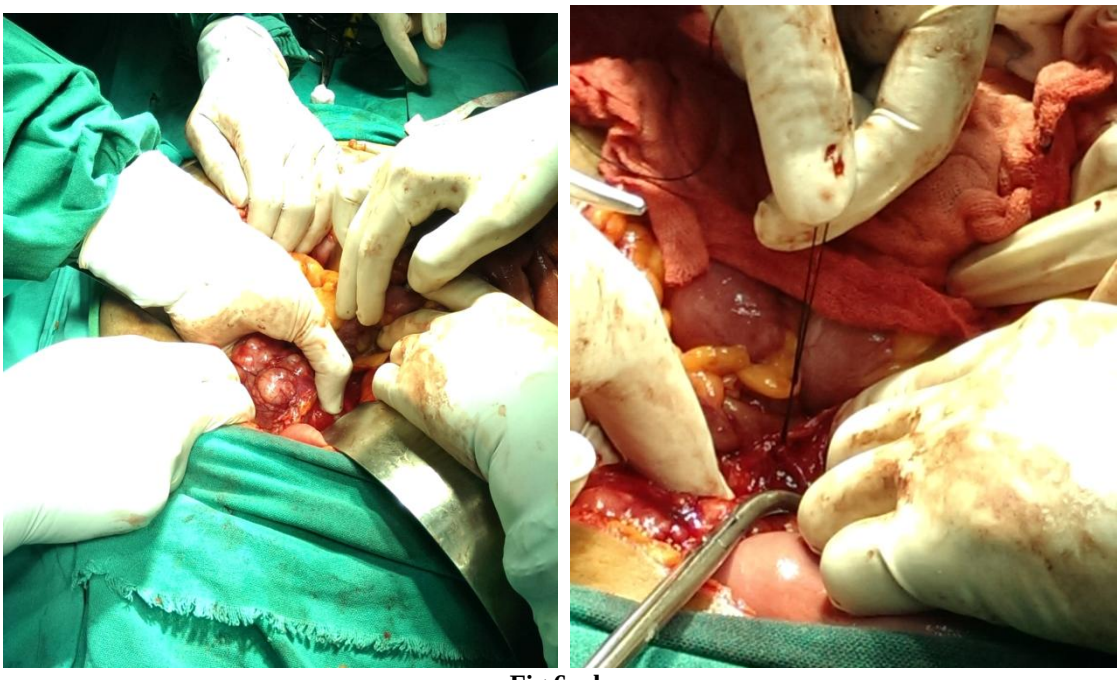

Fig.6a, b

On gross examination specimen looked grey white with external nodular surface. Cut section showed central necrotic and hemorrhagic areas surrounded by greywhite fleshy area. Microscopic picture showed spindle shaped tumor cells arranged as fascicles and had a mitotic picture of pleomorphism, hyperchromatic nuclei.

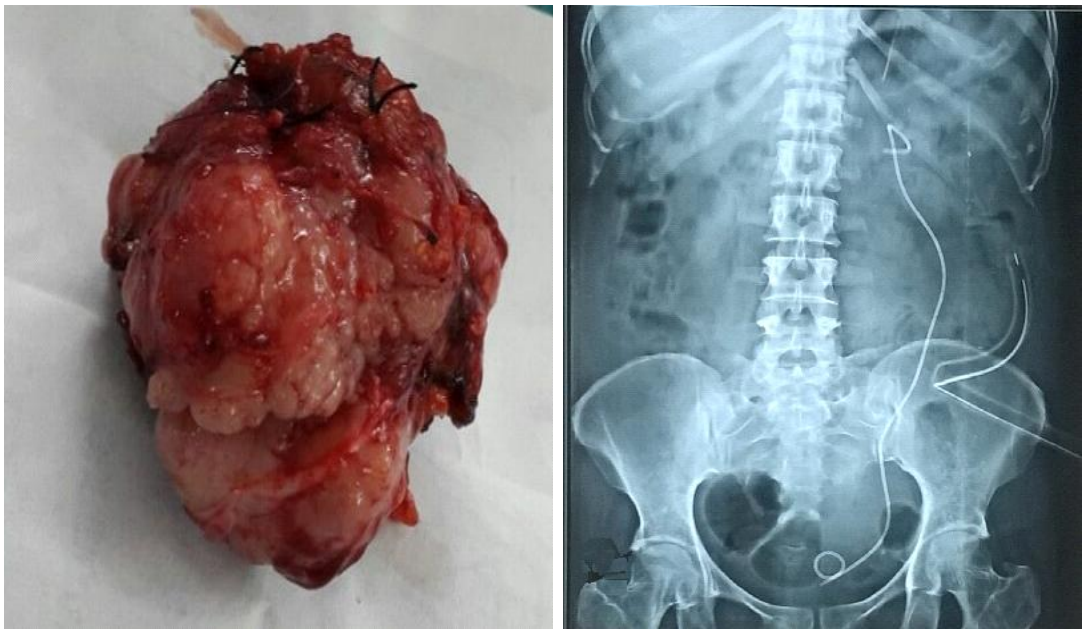

Fig.7a, b

Immunohistochemical staining and microscopic study was positive for vimentin and smooth muscle actin, confirmed the diagnosis of leiomyosarcoma.

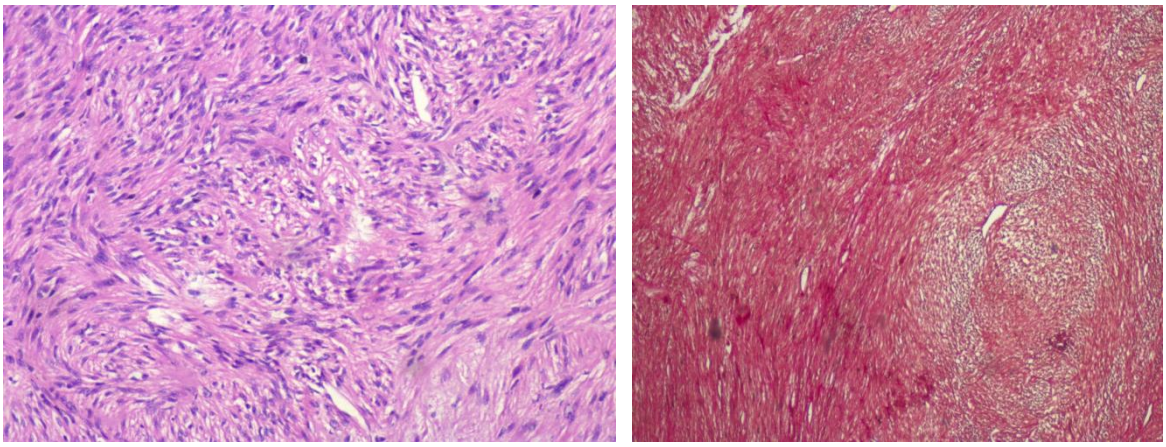

Fig.8a, b

III. Discussion

Transitional cell carcinoma makes up more than 90% of upper urinary tract tumors. In non transitional cell carcinoma most common is squamos cell carcinoma (0.7 to 7%), adeno carcinoma (1%). Multiple types of sarcomas have also been reported to involve an upper urinary tract, including leiomyosarcomas, plasmacytomas, angiosarcomas. The sarcomas are rapid growing tumors, invade the adjacent structures. Leiomyosarcomas have early metastasis to mesentry, lungs, liver and regional lymph nodes.[3, 5, 8, 9]
The number of reported cases are few yet there is a predominance in women. [8]

About 1% Of the upper urinary tract tumors arise in the ureter. Patients present with pain in flank region, microscopic or gross hematuria, a flank mass.[4] In most cases hematuria is absent may be due to non-involvement of ureteral mucosa.[6,7] In two third cases involves distal ureter resulting in an ureteric obstruction. Synchronous bladder involvement found in 40% of cases.[2, 7, 8]

On immunohistochemistry leiomyosarcoma is negative for epithelial markers, but positive for Desmin and SMA showing its smooth muscle origin. A better prognosis is achieved by wide local excision and adjuvant radiation and chemotherapy [1, 3, 5, 8, 9]

At exploratory laparotomy a retroperitoeal neoplasm infiltrating the left ureter was kept in mind but histopathological examination, immunohistochemical staining report was positive for smooth muscle, confirmed it to be leiomyosarcoma.[6,7] We recommended adjuvant radiotherapy of tumor bed to our patient hoping for a better prognosis though radiosensitivity of leiomyosarcoma is doubtful. Due to rarity of the condition nothing

definite can be told regarding management and prognosis.[8] Usual treatment followed is nephro-ureterectomy with a bladder cuff resection as done in ureteral carcinoma. This does not imply that leiomyosarcoma is multicentric or has intraluminal metastasis but accurate preoperative diagnosis is not possible. Prognosis is also unclear due to paucity of data. Not all ureteral leiomyosarcoma are fast growing.[4]

In the present case a complete surgical removal of the tumor with safe margin was done. Patient has completed 16 months of disease free life and is without any signs of recurrence inspite of being noncompliant for chemo-radiotherapy.

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