Primary leiomyosarcoma of the middle ureter: A rare case report with literature review

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Abstract

Leiomyosarcoma is a rarely seen neoplasm of the ureter. Malignant tumors of smooth muscle of the ureter are extremely rare, and about 22 cases of leiomyosarcoma of ureter have been reported to date. A 57-year-old diabetic Pakistani man presented with a dull ache pain in the right flank. Past surgical history was three ureteroscopic surgeries for a ureteric stricture. Computed tomography showed a stricture with a peri-ureteral soft tissue mass of 11 mm x 5 mm at the middle third of the ureter at the level of common iliac vessels. Laparoscopic excision with safety margin and right ureterovesical reimplantation is performed. Diagnosis of leiomyosarcoma of the right ureter was made, and one iliac lymph node was excised and was positive for tumor by pathologic examination. Although leiomyosarcoma is rarely seen in urinary tract, it should be considered in the differential diagnosis of ureteral stricture disease and retroperitoneal tumors.

KEYWORDS

immunohistochemistry, leiomyosarcoma, tumor, ureter

1 | INTRODUCTION

Transitional cell carcinoma is the commonest malignancy in urinary tract and was found in more than 90% of upper urinary tract tumors. In non-transitional cell carcinoma, the most common is squamous cell carcinoma (0.7 to 7%) and adenocarcinoma (1%). Multiple types of sarcomas have also been reported in the upper urinary tract, including leiomyosarcoma, plasmacytomas, and angiosarcomas. The sarcomas are rapidly growing tumors, invade the adjacent structures. Leiomyosarcomas have early metastasis to mesentery, lungs, liver, and regional lymph nodes.1-4

Hematuria is absent in most cases due to non-involvement of ureteral mucosa.5,6 In two-third cases involves distal ureter resulting in a ureteric obstruction. Synchronous bladder involvement was found in 40% of cases.4 Leiomyosarcoma is a rare neoplasm of the ureter with less number of reports in the literature. We report the clinical features, histology, imaging, and treatment of ureteral leiomyosarcoma in a 57-year-old male Pakistani patient.7-10

1.1 | Case report

A 57-year-old Pakistani man presented to our hospital with a dull pain in the right flank, which had persisted for 4 years. He complains of right flank pain and clinically diagnosed as ureteric stricture. The patient is diabetic but controlled by medical treatment. Past surgical history of
the patient was three ureteroscopic surgeries and double J stenting three times for the stricture and one laser cut for the same stricture with stenting for 6 weeks in different institutions in the last few years. Past history of appendectomy when he was a twenty-year-old with ugly scar in the right iliac fossa was seen. The scar was due to complicated appendectomy wound with incisional hernia and repaired with mesh. The patient has unexplained polyuria, and he passes 5 liters of urine every day without consultation. Our team work formed of urologist, nephrologist, endocrinologist, and oncologist.

1.2 | Laboratory investigation

Urine examination identified microscopic hematuria; Blood sugar is normal, renal function tests are within normal. Urine PH is normal and repeated many times with normal results. Anti-diuretic hormone is evaluated with normal results. Potassium results are borderline results and when repeated, postoperatively, he had hypokalemia associated with ileus in the postoperative course.

1.3 | Radiological findings

Ultrasonography identified right moderate hydronephrosis with dilation of the proximal ureter. Computed tomography abdomen and pelvis (CT-KUB) (Figure 1A, B) showed the presence of a stricture of 11 mm x 5 mm at the middle third of the ureter at the level of common iliac vessels with dilation of the proximal ureter and renal pelvis. The peri-ureteral enhanced soft tissue mass was seen 11 mm x 5 mm as reported by CT-KUB with contrast. Furthermore, the lumen of the affected ureter appeared to be irregular in shape. After injection of contrast medium, the mass was greatly enhanced, with a clear portion in the nearby tissues. The distance of the mass from the renal pelvis was about 15 cm.

1.4 | Procedure

Diagnostic ureteroscopy revealed a narrowing of the lumen of the ureter without any mass seen in the ureteric lumen. Under general anesthesia, diagnostic laparoscopy revealed the ureteric stricture at the area of crossing iliac vessels, and proximal ureter is dilated. The ureteric stricture area is resected with safety margin 1 cm above. The proximal tortuous ureter is straightened and re-implanted in submucosal tunnel in the bladder with Boari flap using 4/0 Vicryl. A double J stent 5 Fr/24 cm is fixed for 6 weeks. Postoperatively, the patient had distended abdomen with urine output exceeding 7 liters every day associated with hypokalemia. Potassium is corrected, and bowel movements are resumed. The patient is investigated for diabetes insipidus, and nephrogenic type is diagnosed. Diabetes mellitus is controlled well in the postoperative course. The patient is discharged after 10 days of the surgery with oral potassium and medicines of diabetes mellitus.

1.5 | Pathologic examination

Pathologic examination showed that leiomyosarcoma of the ureter. The cancer was composed of fascicles of interlacing, moderately large, spindle-shaped cells, with abundant eosinophilic cytoplasm. Immunohistochemistry showed strong staining for smooth muscle actin (SMA) (Figure 2A-D). A diagnosis of leiomyosarcoma of the right ureter was made, and one iliac lymph node was excised and was positive for tumor by pathological examination. The cutting margin of the ureter near the bladder was negative, and the kidney was intact. Adjuvant chemotherapy
was suggested after surgery. The patient’s convalescence was uneventful at 6 months after surgery.

1.6 | Follow-up

PET CT scan is performed after diagnosis of leiomyosarcoma to find out any metastasis, but the result was negative. The patient did not have recurrence at 3 and 6 months CT scan examination (Figure 3A, B). We recommended adjuvant radiotherapy or chemotherapy of tumor bed to our patient hoping for a better prognosis though radio-sensitivity of leiomyosarcoma is doubtful.

2 | DISCUSSION

Leiomyosarcoma is a highly malignant tumor with an extremely poor prognosis. The 5-year overall survival rate in patients with soft-tissue sarcoma of all stages remains poor, at only 50–60%, and 5-year disease-free survival is rare. To our knowledge, a literature review has revealed only less than 22 cases of primary leiomyosarcoma of the ureter. We did PubMed search for all leiomyosarcoma cases in urogenital system. Sixty-one cases are reported in humans in the last 46 years. From 1975 to 2021, they reported 22 cases in the ureter and 21 cases in the urinary bladder. Leiomyosarcoma was reported in kidney in 31 cases. Moreover, three cases were reported in the prostate and two cases in the urethra (Table 1).

In our case report, the metastasis to iliac nodes is positive leiomyosarcoma at short-term follow-up for 6 months after surgical excision is good.

An important aspect of the management of leiomyosarcoma is to differentiate the lesion from rhabdomyosarcoma and other spindle-cell neoplasms. This is especially difficult with poorly differentiated high-grade lesions. On immunohistochemistry, leiomyosarcoma is negative for epithelial markers (cytokeratins

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**FIGURE 2** (A). Immunohistochemical staining and microscopic study showing periureteric mass positive for smooth muscle actin (40 X, IHC). (B) was positive for vimentin confirmed the diagnosis of Leiomyosarcoma. (C) Interlacing bundles of pleomorphic spindle cells with storiform pattern in periureteric mass (10x, H & E). (D) Periureteric mass showing elongated, pleomorphic spindle cells (40 x, H & E)

**FIGURE 3** (A, B) Postoperative CT-KUB revealed the double J in situ and high level of urinary bladder due to Psoas hitch.
TABLE 1 PubMed search for previous publications for primary leiomyosarcoma of ureter in humans

| DATE OF PUBLICATION | TITLE | JOURNAL | MAIN AUTHOR |
|---------------------|-------|---------|-------------|
| 1 1975              | Primary sarcoma of the ureter. Study of literature apropos of a personal case | J Urol Nephrol (Paris) | Giraud B et al |
| 2 1979              | Primary leiomyosarcoma of the ureter in a dog. | J Am Vet Med Assoc | J L Berzon et al |
| 3 1980              | Primary leiomyosarcoma of ureter. | Urology | Roemer CE et al |
| 4 1981              | Primary neoplasms of the ureter. | J. Urol | Werth DD et al |
| 5 1983              | Primary leiomyosarcoma of the ureter: a case report with electron microscopy. | J Urol. | Rushton HG et al |
| 6 1984              | Primary ureteral leiomyosarcoma | Scand J Urol Nephrol | Gislason T et al |
| 7 1987              | Primary leiomyosarcoma of the ureter | Actas Urol Esp. | Chesa PN et al |
| 8 1988              | Leiomyosarcoma of the ureter | Eur Urol. | Madgar I et al |
| 9 1993              | Non-urothelial tumors of the urinary tract | Verh Dtsch Ges Pathol | Mikuz G et al |
| 10 1994             | Primary leiomyosarcoma of the ureter | Urol Int | Nakajima F et al |
| 11 1996             | Primary leiomyosarcoma of the ureter. | J Surg Oncol | Griffin JH et al |
| 12 2003             | Primary ureteral leiomyosarcoma: a rare cause of obstructive uropathy | Arch Esp Urol. | Márquez-Moreno AJ et al |
| 13 2006             | A case of primary leiomyosarcoma of the ureter | Hinyokika Kiyo | Shirotake S et al |
| 14 2007             | Poorly differentiated transitional cell carcinoma versus leiomyosarcoma of the ureter: different defects in tumor suppressor genes. | Histopathology | Tzen CY et al |
| 15 2008             | Primary leiomyosarcoma of the ureter. | Asian J Surg. | Chen Lv et al |
| 16 2011             | Prognostic factors and clinical outcomes of urological soft tissue sarcomas. | Korean J Urol. | Lee G. |
| 17 2012             | Leiomyosarcoma of the ureter: a rare case | Diagn Interv Imaging | Aubert E et al |
| 18 2014             | Carcinosarcoma of the Ureter with a Small Cell Component: Report of a Rare Pathologic Entity and Potential for Diagnostic Error on Biopsy | Case Rep Pathol | Newsom K et al |
| 19 2014             | Retroperitoneal Sarcoma Involving Unilateral Double Ureter: Management, Treatment and Psychological Implications | Case Rep Oncol | Lanza V et al |
| 20 2016             | Bladder chondrosarcoma plus urothelial carcinoma in reconverted transitional cell carcinoma of the upper urinary tract: a case report and literature review | World J Surg Oncol. | Cho M.H et al |
| 21 2019             | Primary carcinosarcoma of the ureteropelvic junction associated with ureteral duplication: A case report | Medicine (Baltimore) | Tsuji K et al |
| 22 2021             | Sarcomatoid urothelial carcinoma of the ureter with heterologous elements of chondrosarcoma and osteosarcoma, and concurrent divergent squamous differentiation: A rare case report | Urol Case Rep. | Attia A et al |

and epithelial membrane antigens). Positivity for desmin and SMA shows the smooth muscle origin of the tumor and gives a definitive diagnosis. Negative myoglobin, cytokeratin, and S-100 help to rule out rhabdomyosarcoma, sarcomatoid carcinoma, and melanoma, respectively.16,17 Leiomyosarcoma shows nodular aggregates and densely packed, interlacing bundles of smooth muscle cells in the center. At the margin, strands of smooth muscle cells extend between collagen bundles. Areas of lesser differentiation are present in all tumors
to varying degrees, including irregularly shaped, anaplastic nuclei, and atypical giant cells with bizarre nuclei. A high mitotic rate remains the main criterion for leiomyosarcoma, and other histopathological features of the tumor include atypical smooth muscle cells with peripheral extension into the surrounding tissue, pleomorphism, giant cells, and necrosis of the tumor and peripheral tissue. Only some rhabdomyosarcomas contain cells that are recognizable as rhabdomyoblasts, by virtue of their cytoplasm exhibiting cross-striation, which is best revealed by phosphotungstic acid-hematoxylin staining. Round cells of the embryonal variant tend to have cytoplasmic rims that are stained red by trichrome. The botryoid type is a variant of the embryonal type and is found mainly in mucosa-lined hollow organs. Cells with elongated eosinophilic cytoplasm may be suggestive but not necessarily diagnostic of rhabdomyosarcoma and may be found in the pleomorphic type. A sclerosing variant has cords of small, round malignant cells embedded in a densely hyalinized matrix that has a chondroid and osteoid appearance. Immunohistochemical staining of deparaffinized sections shows expression of desmin and muscle-specific actin in the great majority of leiomyosarcomas. Anti-desmin staining is found in 47–85% of superficial leiomyosarcomas and is less common in higher-grade tumors. Immunohistochemical studies of deparaffinized tissue may show rhabdomyosarcoma cells to be positive for vimentin, which indicates a mesenchymal phenotype. More differentiated cells may be positive for desmin and myoglobin, which is indicative of muscular differentiation. Wide local excision is considered the treatment of choice with radiation and chemotherapy being offered to patients with positive margins or nodes, or those with bulky disease. Radiotherapy can be considered for larger tumors and/or positive margins, if anatomically feasible. Adjuvant chemotherapy has not been proven to be effective and remains investigational. Treatment for metastatic disease is palliative. Active agents include doxorubicin, ifosfamide, gemcitabine, and docetaxel. We recommended adjuvant radiotherapy of tumor bed to our patient hoping for a better prognosis though radio-sensitivity of leiomyosarcoma is doubtful. Due to rarity of the condition, nothing definite can be said regarding management and prognosis. The authors declared no potential conflict of interest with respect to the research, authorship, and/or publication of this article.

**CONCLUSION**

Although leiomyosarcoma is rarely seen in urinary tract, it should be considered in the differential diagnosis of ureteral stricture disease and tumors.

**CONFLICT OF INTEREST**

The authors declared no potential conflict of interest with respect to the research, authorship, and/or publication of this article.

**AUTHOR CONTRIBUTIONS**

HA wrote the manuscript, performed the operation, and managed the patient's perioperative course. HA, YE treated the diabetes insipidus and PK examined the tumor histopathology. YE and PK gave the final approval of this manuscript and read and approved the final manuscript.

**ETHICAL APPROVAL**

All procedures performed in this study were in accordance with the ethical standards of the Institution and/or National Research Committee and with the 1964 Declaration of Helsinki and its later amendments or comparable ethical standards.

**CONTENT**

Written informed consent was obtained from the patient for the publication of this case report and any accompanying images.

**DATA AVAILABILITY STATEMENT**

The data that support the findings of this study are available from the corresponding author upon reasonable request.

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**REFERENCES**

1. Abeshouse BS. Primary benign and malignant tumors of the ureter. *Am J Surg*. 1956;91:237-271.
2. Griffin JH, Waters WB. Primary leiomyosarcoma of the ureter. *J Surg Oncol*. 1996;62:148-152.
3. Roeme CE, Pfister RC, Brodsky G, Sacknoff EJ. Primary leiomyosarcoma of the ureter. *Urology*. 1980;16(5):492-495.

4. Spiess PE, Leibovici D, Pisters LL. PART X: Neoplasms of the upper urinary tract. *Campbell-Walsh Urology*, 11th edn. 2016;1404-1412.

5. Gislason T, Arnarson O. Primary ureteral leiomyosarcoma. *Scand J Urol Nephrol*. 1984;18:253-254.

6. Mehraban D. Primary leiomyosarcoma of the ureter. *Med J Islam Repub Iran*. 1988;2(1):67-69.

7. Aubert E, Millet I, Serre I, Taourel P. Leiomyosarcoma of the ureter: a rare case. *Diagn Interv Imaging*. 2012;93(1):60-63.

8. Shirotake S, Sumitomo M, Asakuma J, Asano T, Aiko S, Hayakawa M. A case of primary leiomyosarcoma of the ureter. *Hinyokika Kiyo*. 2006;52(1):41-45.

9. Márquez- Moreno AJ, Julve-Villalta E, Corral-García P, Gutiérrez-Chacón P, Blanes-Berenguel A. Primary ureteral leiomyosarcoma: a rare cause of obstructive uropathy. *Arch Esp Urol*. 2003;56(2):169-172. Spanish.

10. Madgar I, Goldwasser B, Czerniak A, Many M. Leiomyosarcoma of the ureter. *Eur Urol*. 1988;14(6):487-489. Review.

11. Lv C, Chen N, Zhu X, Zhang X, Zhong Z. Primary leiomyosarcoma of the ureter. *Asian J Surg*. 2008;31(4):191-194. doi:10.1016/S1015-9584(08)60084-6

12. Tzen CY, Wu CJ, Huang ZD, Wu TY. Poorly differentiated transitional cell carcinoma versus leiomyosarcoma of the ureter: different defects in tumour suppressor genes. *Histopathology*. 2007;51(2):271-273.

13. Griffin JH, Waters WB. Primary leiomyosarcoma of the ureter. *J Surg Oncol*. 1996;62(2):148-152.

14. Kasper B, Gil T, Awada A. Treatment of patients with advanced soft tissue sarcoma: disappointment or challenge? *Curr Opin Oncol*. 2007;19(4):336-340. Review.

15. Madgar I, Goldwasser B, Czerniak A, Many M. Leiomyosarcoma of the ureter. *Eur Urol*. 1988;14:487-489.

16. Miettinen M. Antibody specific to muscle actins in the diagnosis and classification of soft tissue tumors. *Am J Pathol*. 1988;130(1):205-215.

17. Ikeda J, Fletcher CD. Immunohistochemical detection of cytokeratin and epithelial membrane antigen in leiomyosarcoma: a systematic study of 100 cases. *Pathol Int*. 2000;50(1):7-14.

18. Lee GH, Kim DS, Chung MJ, Chae SW, Kim HR. Chae H. J.L ysoyl oxidase-like-1 enhances lung metastasis when lactate accumulation and monocarboxylate transporter expression are involved. *Oncol Lett*. 2011;2(5):831-838. Epub 2011.

19. Maruyama E, Azuma H, Yamamoto K, Katsuoka Y. Retroperitoneal leiomyosarcoma found 5 cm in size: a case report. *Hinyokika Kiyo*. 2000;46(9):615-617. Japanese.

20. Kamba T, Kawakita M, Noguchi T, et al. Neoadjuvant CYVADIC (cyclophosphamide, vincristine, Adriamycin and dacarbazine) therapy for retroperitoneal leiomyosarcoma: a case report. *Hinyokika Kiyo*. 1997;43:577-580.

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