

## Primary leiomyoma — A rare tumour of ureter

Ali Akbar Zehri,<sup>1</sup> Athar Ali,<sup>2</sup> Farah Iqbal,<sup>3</sup> Muthoni Jessca<sup>4</sup>

### Abstract

A case of huge primary leiomyoma of the ureter in which nephroureterectomy was performed is presented. To the best of our knowledge, this case is a unique form of leiomyoma of the ureter due to its large size. There have been only twelve cases of primary leiomyoma of the ureter reported since 1955 and eleven of them were very small and one was big in size but smaller than the present case. Our case is considered to be the thirteenth.

**Keywords:** Leiomyoma, Ureteral neoplasm, Hydronephrosis.

### Introduction

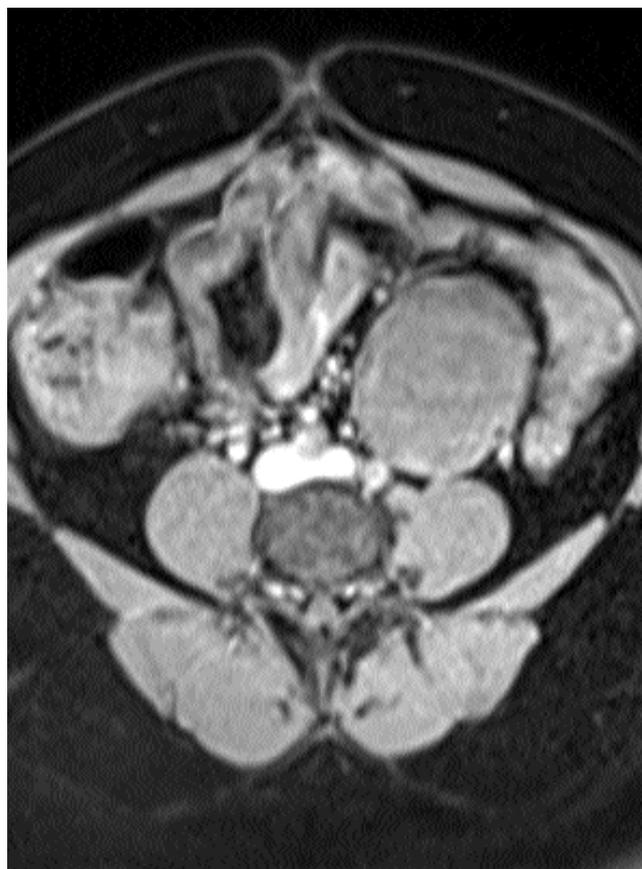
Leiomyomas of the genitourinary tract are rare, and the ureter is an uncommon site for them.<sup>1</sup> Renal leiomyoma, a slow growing benign neoplasm, can arise from the capsule or peripelvic tissues and less often from the renal vein. Since 1955 Leighton et al.<sup>2</sup> reported leiomyoma of the ureter, and now 12 cases have been reported to date. This case will be the 13th. This is a unique form of leiomyoma of the ureter due to its large size. Ureteral leiomyoma often causes hydronephrosis, making it difficult to diagnose. Therefore, total nephroureterectomy is performed because of the possible diagnosis of malignancy. As the technology of ureteroscopy has advanced in recent years, the possibility of a malignant tumour can be excluded by preoperative examination, increasing the number of reports of cases in which the kidney was preserved.<sup>3</sup> We experienced a huge primary leiomyoma of the ureter in which nephroureterectomy was performed. The present report describes this case together with a review of published literature.

### Case Presentation

The patient was a 32 year old female with a history of dull left flank pain and a palpable abdominal mass since 2 years. On physical examination, a soft, mobile and well defined mass was palpated in left lumbar region with no tenderness. Routine lab tests including complete blood

count, liver function tests, renal function studies and urine analysis were all within normal limits.

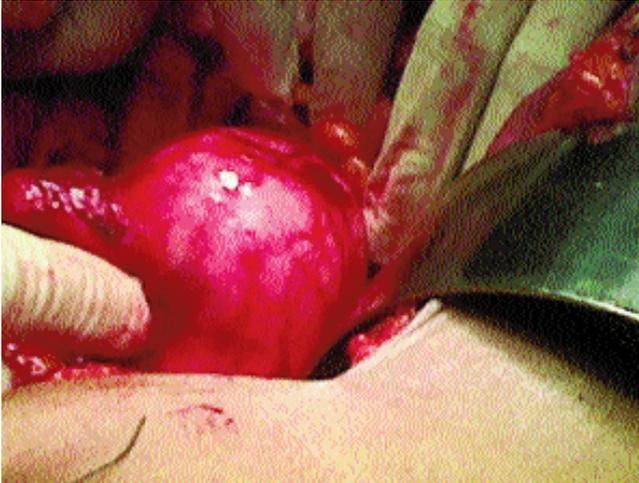
Ultrasound study showed a large heterogeneous intraabdominal mass with compression effect on the left ureter and causing hydronephrosis. Further work up with abdominopelvic Magnetic resonance imaging (MRI) (Figure-1) revealed a well defined encapsulated retroperitoneal mass with lateral tail seen on left side obstructing the mid ureter to a degree that it has resulted in severe parenchymal loss of the left kidney with paper thin parenchyma and gross hydronephrosis. This mass measuring about 135×95mm anteroposterior and transverse dimension was not infiltrating the surrounding structures. However it displaced the mesentery anteriorly.



**Figure-1:** MRI revealed well defined encapsulated retroperitoneal mass measuring about 135×95mm anteroposterior and transverse dimension.

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<sup>1</sup>Department of Surgery, Urology Section, <sup>2</sup>Department of General and Laparoscopic Surgery, <sup>3</sup>Department of Radiology, <sup>4</sup>Aga Khan Health Services, Aga Khan Hospital, Dar es Salaam, Tanzania.

**Correspondence:** Ali Akbar Zehri. Email: [aliakbar.zehri@akhst.org](mailto:aliakbar.zehri@akhst.org)



**Figure-2:** Gross appearance of the mass at the time of surgery.

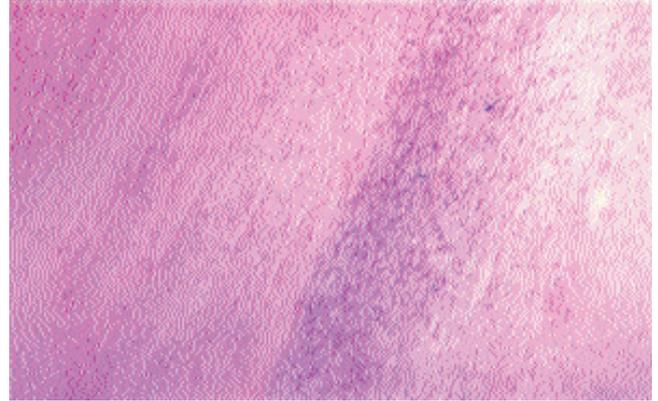


**Figure-3(a):** Macroscopic appearance of the leiomyoma in the left ureter.

It was seen originating at the level of aortic bifurcation and in close proximity to the left iliac artery medially. The mass reached the gonadal artery on the left side which was passing inferiorly. No significant retroperitoneal lymphadenopathy was seen associated with this mass. No pelvic or inguinal lymph nodes were noted. Renal scan revealed non functioning left kidney.

Since there was a very low probability of a ureteral tumour, we did not perform any endoscopic evaluation of left ureter before surgery.

The surgery was performed through a midline incision. There was a soft, mobile and well defined encapsulated mass in left retroperitoneal region (Figure-2). An



**Figure-3(b):** Leiomyoma of the ureter, IHC staining for smooth muscle actin , clone 1A4, 400 xs.

approximately 10x7 cm length of left ureter was completely involved by the tumour. The left kidney was shrunken and preoperatively diagnosis of nonfunctioning left kidney was made on nuclear scan so left nephroureterectomy was done. IHC staining of the mass was positive for alpha smooth muscle actin and desmin (Figure-3).

After removing the Foley's catheter and closed suction drain on 3th and 4th postoperative day, the patient was discharged from hospital with no early complications.

### Discussion

Ureteral leiomyoma as Non-epithelial benign ureteral tumour of mesodermal origin is a rare disease.<sup>1</sup> According to our research, only three cases in Japan and nine cases in other countries have been reported since the case report by Leighton et al. in 1955,<sup>2,4-10</sup> making the present case the 13th (Table 1). Most of the cases were in patients aged 24-60 years except for one case, in an infant, including the present case 32 years. Location of the lesion in six cases was left side of ureter and six on the right side. Eight cases were in men and five in women. The sites of the lesion were in the upper, middle and lower parts in six, three and four cases, respectively. There were no significant differences in the location of the lesion, sex and site of development.

Exact mechanism of development of ureteric leiomyoma is not clear, however inflammation, chronic stimulation, occlusion and trauma are suspected reasons. In a couple of cases, there was history of ureterolithiasis, but not in the present case. Ikota et al. reported a diffuse leiomyoma of the ureter as a complication of multiple endocrinoma (MEN) type 1.<sup>10</sup> Shailesh et al, reported that if preoperative studies and intraoperative findings strongly suggest a benign lesion, some urologists recommend a ureterotomy and biopsy of the lesion for frozen section

Table: Reports of ureteral leiomyoma after 1955.<sup>2</sup>

Case	Reference	Age	Sex	Side	Size (mm)	Location	Treatment
1	Leighton <sup>2</sup>	45	F	Rt	Quite small	Upper	Nephroureterectomy
2	Kao et al. <sup>4</sup>	34	F	Lt	50	Upper	Nephroureterectomy
3	Mondschein <sup>5</sup>	4	M	Lt	20x15x10	Upper	Nephroureterectomy
4	Sekar et al. <sup>6</sup>	35	F	Rt	Micronodule	lower	Ureterctomy
5	Zaitoon <sup>7</sup>	48	M	Lt	25x20x15	Lower	Partial ureterectomy
6	Cussenot et al. <sup>8</sup>	48	M	Rt	2	Lower	Endoscopic biopsy
7	Igarashi et al. <sup>9</sup>	60	M	Lt	10x5x5x	Middle	Nephroureterectomy
8	Yashi et al. <sup>1</sup>	40	F	RT	7	Upper	Partial ureterectomy
9	Ikota et al. <sup>10</sup>	40	M	Lt	15x13x12.2	Upper	Nephroureterectomy
10	Naruse et al. <sup>3</sup>	38	M	Rt	35x13	Lower	Partial ureterectomy
11	Shailesh et al. <sup>11</sup>	43	M	Rt	45x15x5	Middle	Nephroureterectomy
12	Nouralizadeh et al. <sup>12</sup>	24	M	Lt	120x110x75	Middle	Partial ureterectomy
13	Present case	32	F	Lt	15x112x70	Upper/Middle	Nephroureterectomy

with either excision of the tumour or segmental ureteral resection.<sup>11</sup> However, the decision to violate strict principles of cancer operation in an effort to save the involved kidney is a difficult one. Such conservative approach is probably justified in a child if the ipsilateral kidney function is normal. In adults with a poorly functioning kidney, as was the present case who also had non functioning kidney on nuclear scan, nephroureterectomy was the logical option Nouralizadeh et al. reported the second largest leiomyoma of ureter which was diagnosed preoperatively and partial ureterectomy was done.<sup>12</sup>

In MEN type 1, the complication of multiple leiomyoma is recognized in a variety of organs including the oesophagus, stomach, lungs, uterus and skin, but this case was the only one to develop in the ureter. It has been suggested that the MEN type 1-associated gene may have a causal relationship to multiple leiomyoma. In most of the cases diagnosis was made by diagnostic imagings such as excretory urography, retrograde urography or computed tomography (CT) and urinary cytology, as for other ureteral tumours, but no characteristic findings were encountered.<sup>3</sup> Surgical treatment was performed in twelve out of 13 cases. Seven cases were treated by Nephroureterectomy even without defferential diagnosis of benign or malignant tumour before surgery including the present case. Partial ureterectomy preserving the kidney was performed in only four cases and ureterectomy was done in one case. It has become possible to diagnose benign tumours by preoperative examination as a result of advancements in ureteroscopic technology. It is important to pay careful consideration to the preservation of renal function but in the present case,

nuclear scan showed non functioning left kidney.

## Conclusion

Leiomyomas of the genitourinary tract are rare and the ureter is an uncommon site for them. Ureteral leiomyoma often causes hydronephrosis, making diagnosis difficult. As the technology of ureteroscopy has advanced in recent years, the benign nature of the mass can be excluded

If the preoperative suspicion of ureteric tumour is made otherwise, the nephroureterectomy is the logical option.

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