Bilateral Ureteral Leiomyoma With Bilateral Ureteropelvic Junction Obstruction

Hsun-Shuan Wang,[1] Chun-Hsiung Huang,[1,2] Ming-Tan Chen,[1] and Wen-Jeng Wu[1,2]
[1]Department of Urology, Kaohsiung Medical University Hospital, and
[2]Department of Urology, Faculty of Medicine, College of Medicine,
Kaohsiung Medical University, Kaohsiung, Taiwan.

Leiomyomas are benign tumors characterized by overgrowth of visceral smooth muscle in the respiratory, gastrointestinal, and female reproductive tracts. They rarely develop in the urinary system and only 10 cases of unilateral ureteral leiomyoma (UL) have been reported since 1955. No cases of bilateral UL or ureteropelvic junction obstruction due to UL have ever been reported. We present a case of bilateral UL with bilateral ureteropelvic junction obstruction. To the best of our knowledge, this is the first such case report in the English literature.

Key Words: ureteral leiomyoma, ureteral tumor, ureteropelvic junction obstruction
(*Kaohsiung J Med Sci* 2010;26:150–3)

Leiomyomas are benign tumors characterized by overgrowth of visceral smooth muscle in the respiratory, gastrointestinal, and female reproductive tracts. They rarely develop in the urinary system, and only 10 cases of unilateral ureteral leiomyoma (UL) have been reported since 1955 [1]. We present a case of bilateral ureteral leiomyoma with bilateral ureteropelvic junction obstruction (UPJO). To the best of our knowledge, this is the first such case report in the English literature.

Case Presentation

A 24-year-old man was admitted to our hospital with a 1-month history of left flank pain. No history of urolithiasis or previous trauma was mentioned. He denied fever or lower urinary tract symptoms. Physical examination revealed bilateral knocking pain at the costal vertebral angle, which was greater on the left side than on the right side. Abdominal ultrasonography showed bilateral hydronephrosis. Bilateral retrograde pyelography demonstrated bilateral UPJO with bilateral hydronephrosis that was more pronounced on the left side than on the right side, with no evidence of a tumor lesion (Figure 1). Tc^{99m} diethylene triamine pentaacetic acid renal scintigraphy revealed mildly impaired bilateral renal function and bilateral hydronephrosis, especially on the left side. Blood examination and urinalysis indicated no specific abnormalities. Bilateral UPJO with obstructive uropathy was diagnosed, with no evidence of a cause. Bilateral partial ureterectomy with Anderson-Hynes pyeloplasty was performed unilaterally in sequence. A dilated renal pelvis above an apparently normal ureter, with no tumor-like or other anatomical lesions causing the UPJO, was found during the operation. The resected ureter showed a reddish and elastic appearance with a whitish cut surface and no gross, tumor-like lesions (Figure 2). Microscopy identified a

Received: May 11, 2009 Accepted: Jul 27, 2009
Address correspondence and reprint requests to: Dr Chun-Hsiung Huang, Department of Urology, Kaohsiung Medical University Hospital, 100 Shih-Chuan 1st Road, Kaohsiung 807, Taiwan.
E-mail: chhuang@kmu.edu.tw

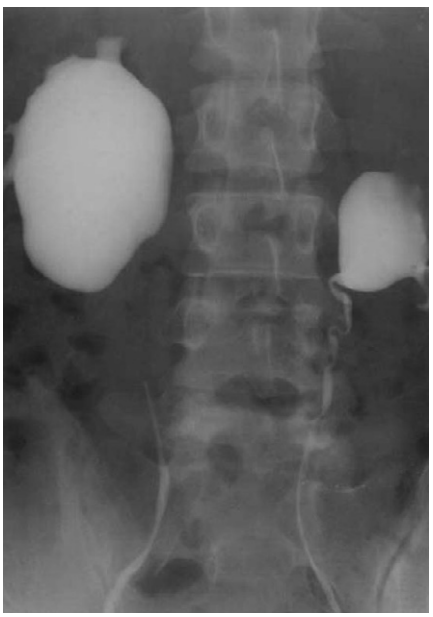

Figure 1. *Bilateral retrograde pyelography indicates bilateral ureteropelvic junction obstruction with bilateral hydronephrosis, with left side is greater in size than right side.*

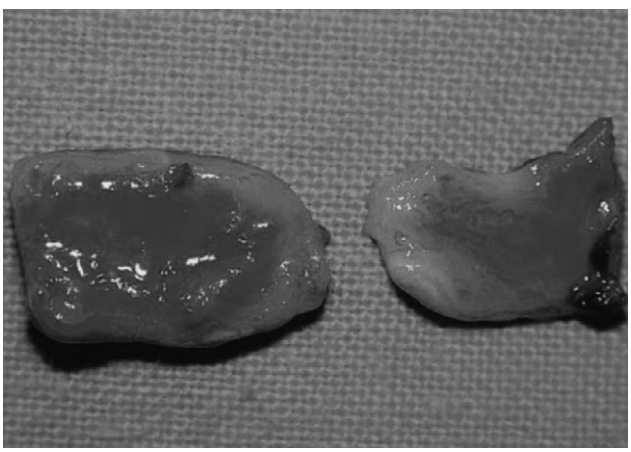

Figure 2. *The resected ureter was reddish and elastic in appearance with a whitish cut surface and no gross, tumor-like lesion.*

well-defined, non-encapsulated tumor in the muscular wall, which was composed of smooth muscle cells with usual cellularity, arranged in interlacing fascicles (Figure 3). Primary bilateral UL was diagnosed on the basis of pathologic evidence. Immunohistochemical analysis with smooth muscle actin further confirmed these findings. Double J stents were placed for 1 month following the pyeloplasties. Ultrasonography, retrograde pyelography, and blood examination at 1-year follow-up showed no signs of hydronephrosis, no evidence of tumor recurrence, and stable renal function.

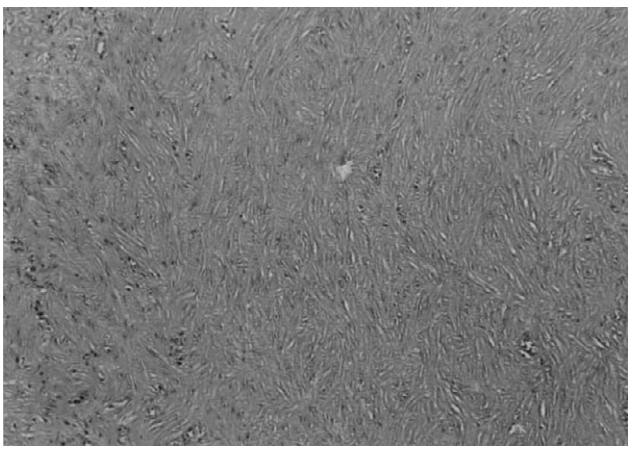

Figure 3. *Microscopy shows a well-defined, non-encapsulated tumor in the muscular wall, composed of smooth muscle cells with usual cellularity, arranged in interlacing fascicles.*

Discussion

Since the first report by Leighton et al in 1955 [1], only 10 cases of unilateral UL have been reported in the English literature [2–10]. No cases of bilateral UL or UPJO due to UL have ever been reported. The case presented here is believed to be the first reported case of bilateral UL with bilateral UPJO.

Of the 10 reported cases of unilateral leiomyoma, four were in Japan. Most of the patients were aged 30–40 years, with the exception of one case in an infant. The present case involved a 24-year-old man, who was significantly younger than most other reported cases. Six of the previous studies involved males and four involved females. In terms of the locations of lesions, six were right-sided and four were left-sided. Four lesions were in the upper ureter, one in the middle, and four in the lower, respectively. One case had lesions at multiple sites in both the upper and lower ureter. There were no significant differences in occurrences in relation to location of the lesion, sex, or site of development [10].

Although leiomyomas are benign tumors that develop mainly in the respiratory, gastrointestinal, and female reproductive tracts, they rarely develop in the urinary system. The mechanisms responsible for the development of ULs remain unclear. Inflammation, chronic stimulation, occlusion, trauma, urolithiasis, and multiple endocrine neoplasia type 1 have been documented. Ikota et al reported a case of multiple endocrine neoplasia type 1 with diffuse leiomyoma of the ureter [9]. Multiple endocrine neoplasia type 1 is

recognized in a variety of organs including the esophagus, stomach, lungs, uterus, and skin; however, the current case is the only reported case of UL. No tumors outside the ureter, and no prior history of ureteral trauma, such as urolithiasis, were noted.

The diagnosis of UL with bilateral UPJO is difficult, especially when the tumor is small and located in the muscular wall, because it can only be detected by microscopic examination, and it mimics other causes of UPJO. UPJO is usually caused by anatomic lesions or functional disturbances that restrict urinary flow across the ureteropelvic junction by compressing it and/or by interfering with peristalsis, resulting in hydronephrosis [11]. Diagnostic methods for UPJO included ultrasonography, intravenous urography/retrograde urography, computed tomography, and diuretic renography. Although tumors are one of the causes of UPJO, no obvious tumor lesion over the ureteropelvic junction was detected in this case, either by imaging or surgical exploration.

Concerns have been raised about the safety of the management of such ureteral tumors. Although ULs are benign tumors, five of the 10 reported cases were treated with nephroureterectomy due to the possibility of malignancy. With the advances in ureteroscopy, direct biopsy of the ureteral tumor and preservation of the kidney has become possible. However, radical excision is still recommended unless preoperative biopsy can confirm the benign nature of the ureteral tumor.

The development of leiomyomas in the urinary system is rare, and only a few unilateral ULs have been reported to date. No cases of bilateral ULs or UPJO due to ULs have been reported. This case of bilateral UL with bilateral UPJO is believed to be the first case report in the English literature. Urologists should keep these benign tumors in mind when investigating ureteral tumors, hydronephrosis, and even UPJO.

References

1. Leighton KM. Leiomyoma of the ureter. *Br J Urol* 1955; 27:256–7.
2. Kao VC, Graff PW, Rappaport H, et al. Leiomyoma of the ureter. A histologically problematic rare tumor confirmed by immunohistochemical studies. *Cancer* 1969; 24:535–42.
3. Mondschein LJ, Sutton AP, Rothfeld SH, et al. Leiomyoma of the ureter in a child. The first reported case. *J Urol* 1976;116:516–8.
4. Sekar N, Nagrani B, Yadav RV, et al. Ureterocele with leiomyoma of ureter. *Br J Urol* 1980;52:400.
5. Zaitoon MM. Leiomyoma of ureter. *Urology* 1986;28: 50–1.
6. Cussenot O, Teillac P, Billebaund T, et al. Leiomyomas of the urinary tract. *Ann Urol Paris* 1989;23:305–8.
7. Igarashi H, Onodera S, Nakada J, et al. A case of leiomyoma of the ureter. *Jpn J Clin Urol* 1994;48:328–30.
8. Yashi M, Hashimoto S, Muraishi O, et al. Leiomyoma of the ureter. *Urol Int* 2000;64:40–2.
9. Ikota H, Tanimoto A, Komatsu H, et al. Ureteral leiomyoma causing hydronephrosis in Type 1 multiple endocrine neoplasia. *Pathol Int* 2004;54:457–9.
10. Naruse K, Yamada Y, Aoki S, et al. A case of primary leiomyoma of the ureter. *Int J Urol* 2007;14:248–50.
11. Koff SA, Mutabagani KH. Anomalies of the kidney. In: Gillenwater JY, Grayhack JT, Howards SS, Mitchell ME, eds. *Adult and Pediatric Urology*, 4th edition. Philadelphia: Lippincott Williams and Wilkins, 2002:2129.

雙側輸尿管平滑肌瘤造成雙側腎盂輸尿管交接處阻塞

王巽玄[1] 黃俊雄[1,2] 陳明潭[1] 吳文正[1,2]

[1]高雄醫學大學附設醫院 泌尿科

[2]高雄醫學大學 醫學院醫學系 泌尿學科

平滑肌瘤為一良性腫瘤，好發於呼吸道、消化道以及女生之生殖道，發生於泌尿系統者則相當少見。從 1955 年至今，僅有十位單側輸尿管平滑肌瘤之病例被報導，並無雙側輸尿管平滑肌瘤之病例，也從無腎盂輸尿管交接處阻塞由此病因所致之文獻被提出過。我們呈現一位雙側輸尿管平滑肌瘤造成腎盂輸尿管交接處阻塞之病例，這是於英文文獻中之首例報告。

關鍵詞：輸尿管平滑肌瘤、輸尿管腫瘤、腎盂輸尿管交接處阻塞

（高雄醫誌 2010;26:150–3）

收文日期：98 年 5 月 11 日
接受刊載：98 年 7 月 27 日
通訊作者：黃俊雄教授
高雄醫學大學附設醫院泌尿科
高雄市 807 三民區十全一路 100 號