KJU
Korean Journal of Urology

www.kjurology.org
http://dx.doi.org/10.4111/kju.2014.55.10.687

Images in Urology

A Rare Cause of Ureteropelvic Junction Obstruction

Chan Ho Lee[1], Sang Don Lee[1,2]

[1]Department of Urology, Pusan National University College of Medicine, Busan, [2]Department of Pediatric Urology, Pusan National University Children Hospital, Yangsan, Korea

Corresponding Author:
Sang Don Lee
Department of Pediatric Urology, Pusan National University Children Hospital, 20 Geumo-ro, Mulgeum-eup, Yangsan 626-700, Korea
TEL: +82-55-360-2671
FAX: +82-55-360-2164
E-mail: lsd@pusan.ac.kr

Article History:
received 19 August, 2013
accepted 16 October, 2013

WHAT IS THE CAUSE OF LEFT URETEROPELVIC JUNCTION OBSTRUCTION?

An 8-year-old boy was referred to Pusan National University Yangsan Hospital with left ureteropelvic junction (UPJ) obstruction. He had experienced episodes of intermittent gross hematuria that had first presented at the age of 5 years. He was otherwise healthy and had no previous medical problems. He had no history of urinary tract infections or stones. A physical examination showed no left flank pain. A complete blood count with differential and the results of renal function tests were within normal limits. Urinalysis revealed microscopic hematuria, whereas the urine culture was negative. Urine cytology showed no evidence of malignancy.

Ultrasonography (USG) showed grade 3 left hydronephrosis according to the Society of Fetal Urology classification, with renal pelvis dilatation of a maximum diameter of 6.4 cm. Longitudinal USG showed a mildly echogenic mass at the dilated upper ureter (Fig. 1A). The right side was normal. The intravenous pyelogram (IVP) revealed hydronephrosis and a dilated renal pelvis that had corkscrew narrowing on the left side. Below the dilated renal pelvis, an S-shaped ureteral fold was observed, with an intraluminal nonopaque and smooth filling defect of about 3 cm in length (Fig. 1B). An obstructive pattern in the left kidney was revealed by ^{99m}Tc diethylenetriamine pentaacetic acid (DTPA) renography. A serial excretion film demonstrated isotope retention in the left kidney of up to 59 minutes (Fig. 1C).

THE DIAGNOSIS: URETEROPELVIC JUNCTION OBSTRUCTION CAUSED BY A FIBROEPHITHELIAL POLYP

An open pyeloplasty with upper ureteral segmental resection was performed. In the intraoperative findings, the S-shaped ureteral fold was observed below the dilated renal pelvis and no aberrant vessels were found. We carefully dissected the fascia around the UPJ, ureteral folding, and renal pelvis. We transected between the ureteral fold and the UPJ area. Upon opening the ureter, we found multiple slender, cylindrical ureteral polyps with smooth surfaces (Fig. 2A, B). Our pathologic findings demonstrated fibroepithelial polyps (FEPs) with chronic inflammation (Fig. 2C).

FEPs of the ureter in children are rare benign tumors that can occur throughout the urinary tract [1]. The current literature indicates that FEPs can occur in children and adults but commonly present in adults in the third through the fifth decades of life, with a male-to-female ratio of 3:2 [2,3]. A review of the English-language literature up to 2007 revealed a total of 28 cases of FEPs causing UPJ obstruction in patients less than 14 years of age [2]. Although the etiology of FEPs is unclear, various theories have been proposed, including developmental defects and acquired conditions [2]. Because of the relatively higher incidence of FEPs in younger children, developmental defects are

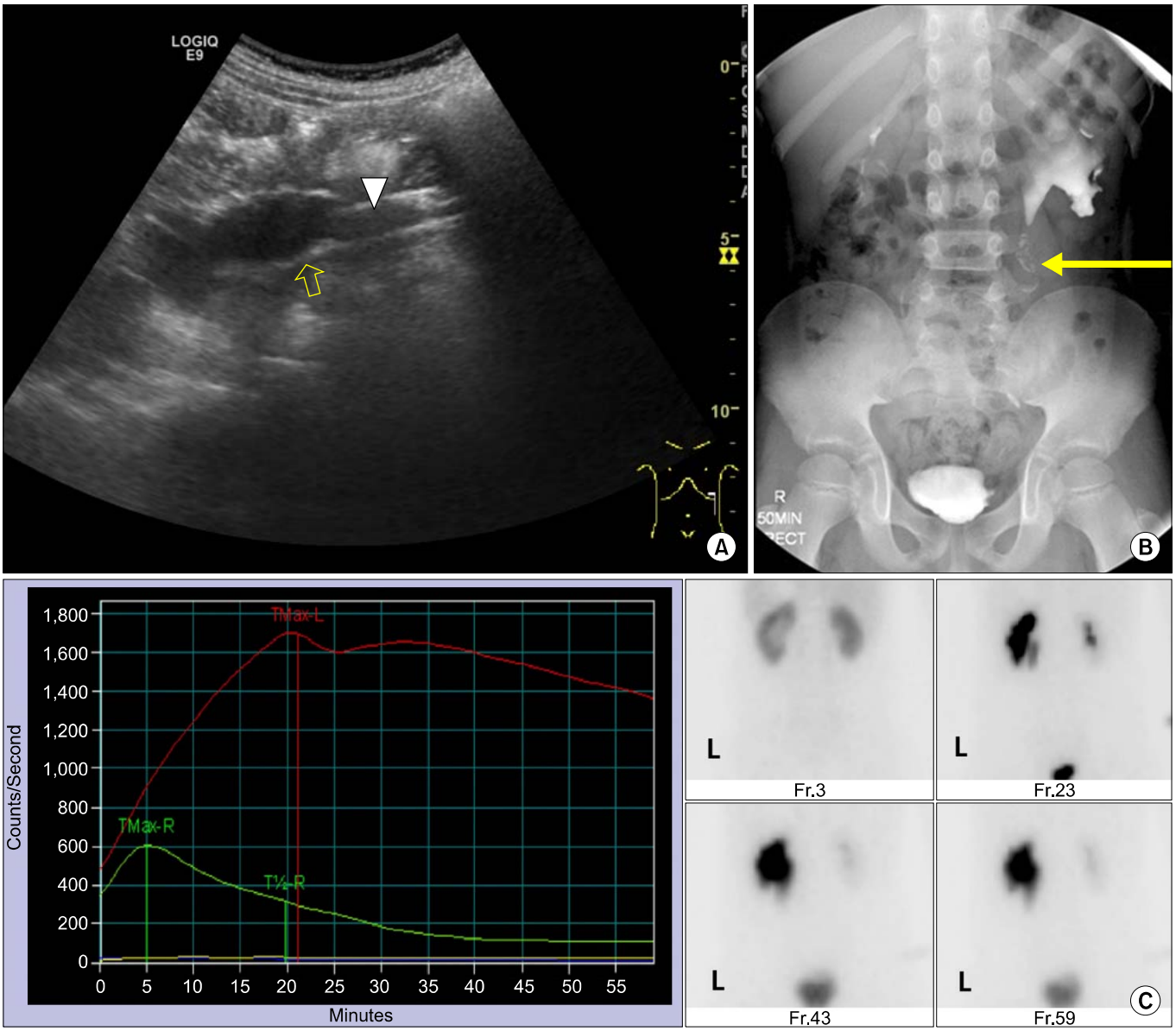

FIG. 1. (A) Longitudinal ultrasonography showed a mildly echogenic mass (arrowhead) and dilated upper ureter (arrow). (B) Intravenous pyelogram showed left hydronephrosis with a dilated renal pelvis with corkscrew narrowing and intraluminal nonopaque and smooth filling defects (arrow). (C) ^{99m}Tc diethylenetriamine pentaacetic acid renography showed the obstructive pattern of the left kidney on the renogram after furosemide administration. A serial excretion film demonstrated isotope retention in the left kidney of up to 59 minutes.

suspected as a main cause. Acquired conditions, such as allergic factors, trauma, and chronic irritation, have also been proposed. The usual location of an FEP of the ureter is in the UPJ or the upper ureter [2]. FEPs are also found in the renal pelvis, and a small number develop in the posterior urethra or bladder [3,4]. Most polyps are single and are shorter than 5 cm. However, some studies report long, multiple, and bilateral polyps [3]. The most common symptom resulting from urinary tract obstruction by FEPs is flank pain [2]. Other presenting features, such as urinary frequency, hematuria, and dysuria, have also been reported. However, because of the slow growth and nonmalignant nature of FEPs, some patients with FEPs may be asymptomatic. FEPs have smooth surfaces; are tan-gray, grayish-white, or pink in color; and can be single long, slender cylindrical masses or multiple finger-like projections attached to a single base [1,2]. In most cases, FEPs are longer than they are wide [2]. They are usually distal to the base as a result of the urinary flow and the peristalsis of the ureter. Histologically, FEPs are benign tumors of mesodermal origin [1]. FEPs are covered with normal urothelium and consist of thick, loose, and fibrous connective tissues that are rich in smooth muscle cells, blood vessels, and dense collagen [1,2].

The preoperative diagnosis of ureteral polyps is difficult. The USG can show hydronephrosis secondary to UPJ

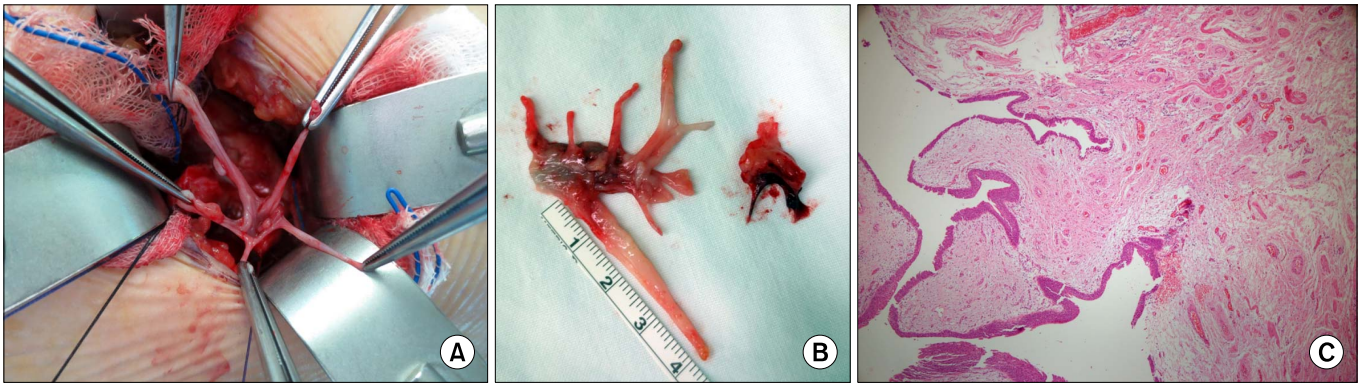

FIG. 2. Intraoperative images and histologic findings. (A) Fibroepithelial polyps (FEPs) were exposed via ureterotomy. Multiple slender, cylindrical ureteral polyps with smooth surfaces were observed. (B) Morphology of the multiple excised long FEPs. (C) Histologic examination showing FEPs composed of loose fibrovascular connective tissue covered with a layer of normal urothelium (H&E, ×40).

obstruction. In the USG, FEPs are shown as mildly echogenic and nonshadowing, with well-defined margins outlined by a urine-distended pelvis [5]. If a USG suggests the presence of UPJ obstruction or other ureteral problems, IVP may be indicated. In the IVP, FEPs can appear to be long, smooth ureteral filling defects with various degrees of hydronephrosis. However, the detection rates of FEPs through IVP vary from 36% to 100% according to current literature [1]. When a filling defect is found in the IVP, other conditions, such as nonopaque renal calculi, blood clots, or mucosal folds, should be excluded [4]. The use of ureteroscopy for the diagnosis and endoscopic removal of FEPs has been reported, but ureteroscopy is not useful in young children because of the small working space available [1,6].

Treatment of ureteral FEPs includes segmental resection with ureteroureterostomy, nephroureterectomy, local coagulation by laser, and polypectomy by ureteroscopy [2,7]. In the past, FEPs within the ureter were treated by nephroureterectomy because of potential malignancy [1,2]. However, because FEPs are benign lesions, a conservative approach is now taken in most cases [2]. In children, therefore, the operation of choice for FEPs causing UPJ obstruction is resection of the tumor-bearing ureteral segment, with or without dismembered pyeloplasty [1,2]. Laparoscopic surgery is also performed as a minimally invasive treatment and can achieve complete resection of the polyp in children [7]. Recently, many reports have described success with the use of ureteroscopy and with holmium: yttrium-aluminium-garnet laser excision and coagulation [3,6]. Although ureteroscopic excision is a less invasive and widely available alternative to an open procedure, it presents difficulties in patients with multiple long polyps [7]. Before endoscopic procedures are carried out, surgeons should consider the possibility of incomplete resection. In our case, we performed the upper ureteral segmental resection with open pyeloplasty because the IVP showed a dilated renal pelvis. Although we did not perform ureteroscopy, we would pick open surgery again because of the high possibility of ureteral stricture after endoscopic procedures in a case such as this with multiple FEPs.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

ACKNOWLEDGMENTS

This work was supported by Pusan National University Yangsan Hospital Research Grant.

REFERENCES

1. Adey GS, Vargas SO, Retik AB, Borer JG, Mandell J, Hendren WH, et al. Fibroepithelial polyps causing ureteropelvic junction obstruction in children. J Urol 2003;169:1834-6.
2. Cassar Delia E, Joseph VT, Sherwood W. Fibroepithelial polyps causing ureteropelvic junction obstruction in children--a case report and review article. Eur J Pediatr Surg 2007;17:142-6.
3. Lam JS, Bingham JB, Gupta M. Endoscopic treatment of fibroepithelial polyps of the renal pelvis and ureter. Urology 2003;62:810-3.
4. Williams TR, Wagner BJ, Corse WR, Vestevich JC. Fibroepithelial polyps of the urinary tract. Abdom Imaging 2002;27:217-21.
5. Wang XM, Jia LQ, Wang Y, Wang N. Utilizing ultrasonography in the diagnosis of pediatric fibroepithelial polyps causing ureteropelvic junction obstruction. Pediatr Radiol 2012;42:1107-11.
6. Carey RI, Bird VG. Endoscopic management of 10 separate fibroepithelial polyps arising in a single ureter. Urology 2006;67:413-5.
7. Bian Z, Liu X, Hua Y, Liu F, Lin T, He D. Laparoscopic management of multiple ureteral polyps in children. J Urol 2011;186:1444-9.