Idiopathic bilateral ureteral stenosis presenting as bilateral hydronephrosis

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ABSTRACT

Ureteral stenosis is typically seen in the setting of genitourinary tract manipulation or nephrolithiasis. Bilateral idiopathic ureteral stenosis is an exceedingly rare clinical entity that has been described in only a small number of cases. More specifically, we describe a case of idiopathic bilateral, distal ureteral stenosis resulting in hydronephrosis and deterioration of renal function in an adolescent patient. The patient was successfully treated with robotic bilateral ureteral implants. This case brings to light a rare, but clinically relevant, cause of hydronephrosis, highlights the importance of early intervention in minimizing renal dysfunction, and describes a novel treatment option.

1. Introduction

Bilateral ureteral stenosis is a rare clinical entity. When pediatric and adolescent ureteral stenosis occurs, it is usually a unilateral, mid-ureteral stricture and often considered secondary to congenital anatomic anomalies. Other hypotheses suggest they may be exaggerated physiological narrowing at the ureteropelvic junction (UPJ), ureterovesical junction (UVJ), and pelvic brim. Prior reports describe a small number of patients with bilateral ureteral stenosis, most of whom presented within their first year of life with mid-ureteral strictures.

Herein, we report a case of a 19-year-old boy who presented with bilateral hydronephrosis and impaired kidney function. After a lengthy workup, the patient was diagnosed with idiopathic, bilateral, distal ureteral stenosis. He was successfully treated with robotic bilateral ureteral reimplantation.

2. Case presentation

An otherwise healthy, 19-year-old boy presented with one-month of bilateral flank pain. Physical exam was within normal limits. Laboratory measurements revealed an elevated serum creatinine of 1.38 mg/dL (<1.30 mg/dL); otherwise, the patient’s complete blood count, basic metabolic panel, and urine analysis were within normal limits. Renal bladder ultrasound revealed bilateral hydronephrosis with possible bilateral ureteroceles. Non-contrast computed tomography revealed bilateral severe hydroureteronephrosis without the presence of nephrolithiasis or urolithiasis (Fig. 1).

Due to suspicion of vesicoureteral reflux, a voiding cystourethrogram (VCUG) was obtained. The VCUG failed to detect any reflux and showed normal bladder anatomy. Nuclear medicine renography revealed normal bilateral renal uptake and delayed excretion with possible high-grade obstruction, with a split renal function of 47% on the right and 53% on the left. Following multidisciplinary rounds with pediatric nephrology, urology, and radiology, a presumptive diagnosis of bilateral hydronephrosis secondary to possible obstructive ureteroceles was made. The patient was brought to the operating room for cystoscopy, stent placement, and ureteroceles incision. Cystoscopy revealed a bulging and stenosis of both ureteral orifices, though without clear evidence of ureteroceles. The right ureteral orifice could not be located and, therefore, was not cannulated. Meanwhile, the left ureteral orifice was found but stenosis in the distal ureter prevented stent placement. Retrograde pyelography revealed stenosis of the left distal ureter (Fig. 2). Following surgery, he endorsed worsening left flank pain and a subsequent CT abdomen and pelvis with and without contrast revealed persistent moderate bilateral hydronephrosis and hydroureret with incomplete filling of the left-sided tract (Fig. 3).

Given the failure to dilate and stent the patient’s ureters during cystoscopy, a robotic-assisted bilateral ureteral reimplantation was performed. Intraoperatively, there was noted to be significant J-hooking of the distal bilateral ureters and bilateral stenosis of the distal ureter 3...
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2 cm proximal to the UVJ. Resection of the stenotic segments of the ureters and successful non-refluxing reimplantation of the ureters onto the anterior bladder in a lich-Gregoir fashion was performed. Bilateral ureteral stents were placed.

Post-operatively, the patient’s serum creatinine decreased to 0.81 mg/dL. The surgical pathology specimen demonstrated marked luminal narrowing along with focal smooth muscle disorganization and mural fibrosis without evidence of malignancy. One-week post-operation, the patient passed a trial of void following foley catheter removal. At 6-weeks post-operation, office cystoscopy was performed with subsequent removal of the bilateral stents without complication. The bladder was noted to be normal. Repeat metabolic panel revealed sustained normal renal function, with a creatinine of 0.74 mg/dL. A post-operative VCUG was not obtained given his clinical improvement and lack of concern for an infectious process.

At 2-months post-operation, a repeat renal-bladder ultrasound revealed improved mild left hydronephrosis and moderate right hydronephrosis, which was reported to be unchanged compared to pre-operative sonography. At 4 months post-operation, a nuclear medicine renal scan revealed normal kidneys bilaterally without evidence of obstruction and with a split quantitative function of 48% on the right and 52% on the left. At 10 months post-operation, a repeat renal scan again revealed normal kidneys bilaterally without evidence of obstruction. He continues to endorse feeling well without urinary symptoms.

3. Discussion

Herein we discuss a rare case of idiopathic, bilateral, distal ureteral stenosis in an adolescent patient successfully managed with robotic bilateral ureteral reimplantation.

Typically, pediatric and adolescent hydronephrosis is due to congenital collecting system abnormalities, such as vesicoureteral reflux, UPJ obstruction, or posterior urethral valves. In our case, the patient presented without a significant medical history or atopic symptoms, and thus, we suspect his stenosis is likely congenital in nature. Past literature includes only a handful of cases that were primarily localized to the mid-ureter. However, the presented case is localized to the distal ureter rather than the mid-ureter. Furthermore, the presented case is distinct from other ureteral pathologies, such as a mega-ureter, as the obstruction was found to be at the level of the distal ureter rather than at the ureterovesical junction. With regard to diagnosis, our case further confirms that retrograde pyelography, as opposed to renal ultrasound or radionuclide renography, is crucial for accurate diagnosis. Accurate and prompt diagnosis is paramount since ureteral stenosis, and resultant hydronephrosis, can lead to renal function deterioration, as seen in our patient.

Historically, ureteral stenosis was — and in many cases still is — treated with an open surgical ureteral reimplantation or ureteroureterostomy. That said, a robotic-assisted laparoscopic approach to ureteral stenosis repair has gained popularity. Given the decreased morbidity seen with robotic-assisted laparoscopic procedures compared to open surgeries, we opted for, and successfully completed, our patient’s ureteral reimplantation via a robotic approach. At almost one-
year post-operation, he showed no evidence of surgical complication.

4. Conclusion

Here, we present a rare case of idiopathic, bilateral, distal ureteral stenosis presenting with hydronephrosis and renal deterioration. Our case highlights the importance of early recognition and treatment of hydronephrosis to prevent kidney injury. While a rare cause of hydronephrosis, patients being evaluated for hydronephrosis should have ureteral stenosis considered if other etiologies have been ruled out. Ureteral dilation and stent placement may serve as an appropriate first-line therapy; however, urologists should be prepared to perform ureteral reimplantation should initial therapeutic measures fail, with a robotic approach serving as a viable and effective treatment option.

Ethics

Patient consent was obtained prior to writing of this case report.

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Declaration of competing interest

The authors have no conflicts of interest, financial or otherwise, to declare.

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None.

Fig. 3. (A) Axial and (B) coronal CT Urogram revealing persistent moderate bilateral hydrourerter with incomplete filling of the left sided tract.