"Let's get it straight": the story of the spiral ureter

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A "corkscrew deformity" of the proximal ureter is a rare congenital anomaly that typically lacks any postnatal clinical significance. The rarity of this entity, however, has not allowed the clarification of its natural history and the ideal approach to its management.
We herein present a case of a 27-year-old female patient who presented with right flank pain and significant hydronephrosis. On retrograde ureterography, a typical spiral configuration of the proximal ureter was noted. The patient underwent successful reconstruction by laparoscopic transperitoneal dismembered pyeloplasty.
We report the first use of laparoscopic reconstruction for the management of "corkscrew deformity" of the proximal ureter and we focus on the imaging findings, technical details, advantages and limitations of this technique.

Key Words: ureter, corkscrew, spiral, laparoscopy, congenital

Introduction

A spirally twisted or "corkscrew" ureter is a very rare anomaly beyond the neonatal age. In a study of 12,080 children autopsies, this anomaly was observed only twice.[1] The presence of a spirally twisted ureter is not considered clinically significant, unless it causes obstruction and secondary hydronephrosis. The rarity of this entity, however, has not allowed the clarification of its natural history and the ideal approach to its management.

We herein report the unique case of a female patient who presented in adulthood with upper urinary tract obstruction secondary to a spirally twisted ureter and was managed successfully by laparoscopic reconstruction.

Case report

A 27-year-old female patient presented in the Emergency Department with acute right flank pain. The patient's past history was unremarkable for problems related to the urinary tract such as urolithiasis, recurrent urinary tract infections or vesicoureteric reflux.

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An abdominal computed tomography (CT) scan revealed the presence of a grossly dilated right pelvicalyceal system and upper ureter. A subsequent 99mTc-mercaptoacetyltriglycine (MAG3) scan confirmed the presence of right upper tract obstruction and showed a relative function of 27% of the right kidney. The left kidney exhibited good uptake of tracer material, no obstruction and a relative function of 73%, Figure 1. Retrograde ureterography suggested the presence of a spirally-shaped proximal ureter, as well as secondary hydronephrosis, Figure 2.

After appropriate consent, the patient elected to undergo reconstruction by laparoscopic transperitoneal approach. Access was achieved through three 5 mm ports. The right colon and mesentery were deflected medially, allowing identification of the right ureter near the lower pole of the right kidney. The dilated renal pelvis was identified and dissected along with the proximal ureter, which followed a spiral configuration. In fact, the proximal ureter was enclosed by thick fibrous bands and two complete spirals were noted along its course, Figure 3. Sharp dissection of the investing tissue led to the identification of a patent PUJ and a persistent spiral twist of the proximal ureter, in the absence of any crossing vessels. The classical approach of Anderson-Hynes dismembered pyeloplasty was used, with excision of the spiral part of the ureter and lateral spatulation of the ureter and renal pelvis. A watertight tension-free anastomosis between

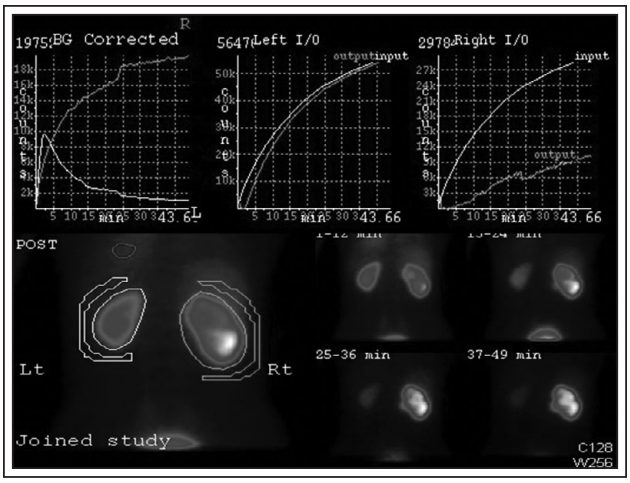

Figure 1. 99mTc-mercaptoacetyltriglycine (MAG3) scan. Confirmation of right upper tract obstruction and a relative function of 27% of the right kidney. The left kidney exhibited good uptake of tracer material, no obstruction and a relative function of 73%.

the renal pelvis and the ureteric stump was achieved with running 4-0 polyglactin sutures, intracorporeal freehand suturing and in situ knot-tying, Figure 3. Prior to the completion of the anastomosis, a double-J stent was inserted in an antegrade manner. The total operative time was 92 minutes

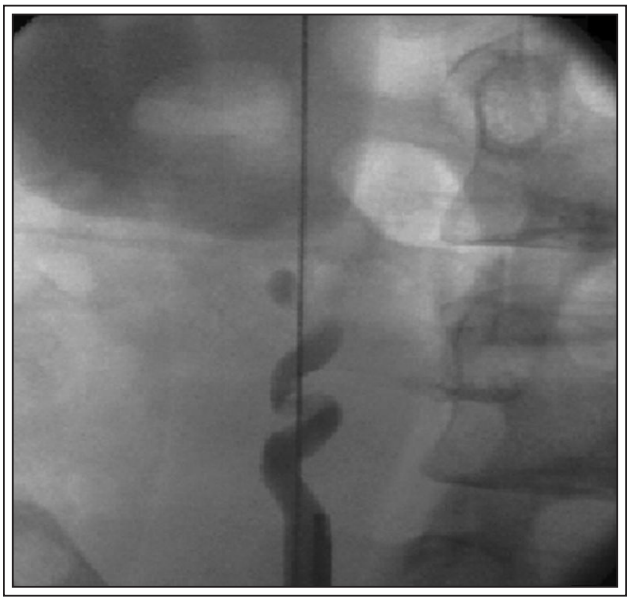

Figure 2. Retrograde ureterography. Findings consistent with the presence of a spirally-shaped proximal ureter, as well as secondary hydronephrosis.

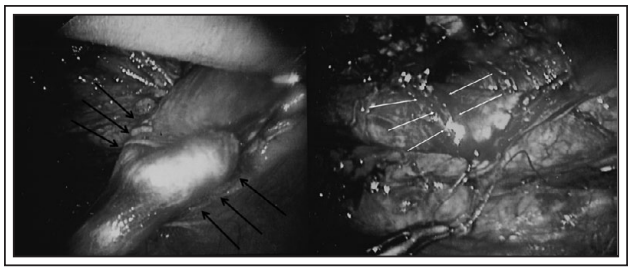

Figure 3. Intraoperative images. The mobilized proximal ureter is enclosed by thick fibrous bands and two complete spirals are noted along its course (left image, black arrows). A watertight tension-free anastomosis between the renal pelvis and the ureteric stump was achieved with running 4-0 polyglactin sutures (white arrows), over a double-J stent inserted in an antegrade manner.

The early postoperative course was uneventful. The urethral catheter was removed on the morning of the second postoperative day and the abdominal drain was removed 6 hours later. The patient was discharged on postoperative day 3. The double-J stent was removed 6 weeks later.

A MAG-3 renogram, performed 3 months later, confirmed the absence of right kidney obstruction, while the patient remained asymptomatic at last follow up.

Discussion

The embryological development of the upper urinary tract has been extensively described and represents the basis for understanding the pathogenesis of congenital anomalies in number, structure, position and course of the pelvicalyceal system and ureter. At 4 weeks' gestation, the ureteric bud arises as an outpouching from the distal mesonephric duct and interacts with the mesenchyme of the metanephric blastema. This interaction induces branching of the ureteric bud and development of the pelvicalyceal system and the ureter, while the metanephric blastema forms all elements of the nephron.[2] The interaction between the ureteric bud and the metanephric blastema is of critical importance and any alteration or disruption of this interaction may potentially lead to anomalies of the renal parenchyma, collecting system and ureter.[3] Congenitally abnormal ureteric development is considered clinically significant if it causes proximal obstruction. Clinical presentation may be acute or insidious and therefore some patients first present in adulthood with upper urinary tract dilatation secondary to congenital abnormalities of ureteric structure or course.

According to Kirks et al,[4] infants frequently have a "corkscrew" appearance of the proximal segment of the ureter on intravenous urography, but this is considered an imaging finding of no postnatal clinical significance. It may represent persistence of normal fetal developmental structures, such as folds (as described by Östling in 1942), mucosal redundancy and spiral convolutions.[5] Persistent mucosal folds, however, are associated with a smooth external appearance of the ureter.[4] In the case presented herein the external surface of the proximal ureter had a spiral configuration. Another possible explanation is the failure of the ureter to rotate with the kidney,[1] but this hypothesis does not explain the presence of two complete spirals. Corkscrew configuration of the ureter may also be the result of ureteric varicosities [6] or extrinsic ureteric obstruction.[7] Obstruction secondary to spiral deformity of the ureter appears to result from the ensheathment of that part of the ureter by dense fibrous bands.[8]

Because of its high efficacy and low morbidity, laparoscopic dismembered pyeloplasty is considered today the gold standard for the surgical management of ureteropelvic junction obstruction.[9,10] The use of laparoscopy in adults presenting with less common congenital anomalies of the ureter has not been extensively studied, due to the rarity of these entities, but successful laparoscopic management of congenital strictures involving the proximal or middle segment of the ureter has been described in the past.[11,12] On the other hand, the laparoscopic approach, either retroperitoneal or transperitoneal, has emerged as the first-line option for the surgical management of retrocaval ureter.[13,14]

From a technical standpoint, the principles of dismembered pyeloplasty were applied in this case in a relatively straightforward manner. Tension-free anastomosis was achieved, despite the need to excise the spiral portion of the ureter. Extensive distal mobilization of the ureter was not necessary, thus avoiding ischemia secondary to extensive dissection of the ureteric sheath. Prior ureteric stenting may be technically challenging due to the tortuous course of the proximal ureter.[14] The presence of a preoperatively inserted stent may also hinder posterior anastomotic suturing. We prefer intraoperative insertion of a double-J stent in an antegrade manner after completing the posterior part of the anastomosis and before proceeding to suturing the anterior part.

Newer approaches to minimally invasive reconstructive surgery of the upper urinary tract, such as robotically assisted laparoscopy[15] and laparoendoscopic single-site surgery (LESS),[16] may emerge as safe and effective alternatives to standard laparoscopy, but the available evidence in regards to these techniques are limited by the short follow up and small sample size of the relevant studies.

In conclusion, upper urinary tract obstruction secondary to a spiral deformity of the proximal ureter is a rare, yet significant, clinical entity. Laparoscopic reconstruction by dismembered pyeloplasty, due to its minimal morbidity, quick convalescence and relative technical ease, should be considered the first-line option for the surgical management of adult patients presenting with this rare congenital anomaly. □

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