Ectopic Megaureter Opening to Urethra and Leading to Pyonephrosis and Urinary Incontinence: Case Report

Üriner İnkontinans ve Piyonefroza Neden Olan Üretraya Açılımlı Ektopik Megaüreter

Osman ERGÜN, MD,[a]
Sedat SOYUPEK, MD, Assoc.Prof.[a]

[a]Department of Urology,
Süleyman Demirel University
Faculty of Medicine, Isparta

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Yazışma Adresi/*Correspondence:*
Osman ERGÜN, MD
Süleyman Demirel University,
Faculty of Medicine,
Department of Urology, Isparta,
TÜRKİYE/TURKEY
osmanergun77@mynet.com

ABSTRACT Ectopic ureter in adult female patients usually presents with urinary incontinence. Clinical manifestations include incontinence and urinary tract infections. Here we report a case of ectopic megaureter opening to urethra which caused incontinence and obstructive urinary symptoms in a 19 years old woman. When we reviewed the literature, we found several cases regarding ectopic ureter associated with a single (nonduplicated) collecting system opening to the urethra. Typically, the diagnosis is made after a long delay. Surgical reconstruction of an ectopic ureter should be planned according to the degree of renal function. Most patients are cured with ureteroneocystostomy or nephroureterectomy. However, following these surgical procedures, some incontinent women may have persistent urinary incontinence due to mal-development of the bladder neck and sphincter or Gartner's duct cyst.

Key Words: Ureteral diseases; urinary incontinence; urinary tract infections

ÖZET Yetişkin kadın hastalardaki ektopik üreter sıklıkla üriner inkontinansla kendini göstermektedir. Klinik bulgularında inkontinans ve üriner sistem enfeksiyonları bulunmaktadır. Makalemizde ektopik megaüreterin neden olduğu inkontinans ve tıkayıcı idrar yolu bulgularının olduğu 19 yaşındaki kadın hastayı sunmaktayız. Literatürü gözden geçirdiğimizde ektopik üreterin üretraya açıldığı, tek toplayıcı sistemin eşlik ettiği az sayıda olgu sunumu bulduk. Tipik olarak çok geç tanı konulmaktadır. Ektopik üreterin cerrahi olarak yeniden yapılandırılması böbrek fonksiyonlarının seviyesine göre planlanmalıdır. Hastaların büyük bir kısmı üreteroneosistostomi ve nefroüreterektomi ile tedavi edilmektedir. Ancak, cerrahi işlemler sonrasında bazı inkontinent kadınların üriner inkontinansı mesanenin boyun ve sfinkterinin ya da Gartner kanal kistinin kötü gelişimi nedeniyle tekrarlayabilmektedir.

Anahtar Kelimeler: Üreter hastalıkları; üriner inkontinans; üriner kanal enfeksiyonları

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Ectopic ureter (EU) is more common in adult females than the males and usually presents with urinary incontinence. The common presenting symptoms of EU include urinary tract infection and incontinence.[1] Ureteral abnormalities that are related EU directly affect kidney functions and they are one of the most important urogenital abnormalities. EU can be asymptomatic in childhood and remain undiagnosed until adulthood. About half of female patients with EU present with urinary incontinence. It is due to relaxation of urogenital diaphragm, through which the ureter travels.[2] Classical radiological methods such as ultrasonography (US), voiding cystourethrogram (VCUG) and intravenous pyelogram (IVP) are

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usually performed to determine EU. It is typically associated with a duplicated collecting system. There is a duplicated collecting system in 0-80% of EU and approximately 15% of EU have a single collecting system.[3-5] EU opens to urethra extremely rarely. It frequently results with the dysplasia of associated renal segment in girls with ureteral duplication. However, single system ureteral ectopia is uncommon and is seen more frequently in boys. When we reviewed the literature, we found several cases regarding to EU associated with a single (nonduplicated) collecting system opening to the urethra. To our knowledge, this is the first case in the literature reporting an ectopic megaureter opening to urethra and causing pyonephrosis without renal function disturbance in a woman with a single collecting system. The aim of this case report is to present and discuss diagnostic and therapeutic problems of EU opening to urethra that causes pyonephrosis and urinary incontinence.

CASE REPORT

A 19-year-old female was referred with intermittent urinary incontinence starting from her childhood and recurrent left renal colic throughout the previous year. She had a normal micturition pattern and had delivered a baby 5 months previously. Her complaints increased after the childbirth. She applied to a medical center in June 2007 for a pregnancy control. At that time, left pyonephrosis was diagnosed. A percutaneous nephrostomy catheter (PNC) was inserted and antibiotics were administered. A second PNC was inserted after the pregnancy. When she was admitted to our clinic, grade 2 hydroureteronephrosis and a megaureter in left kidney (Figure 1A; distal ureter wide: 13 mm) were seen on US. The IVP confirmed the left hydroureteronephrosis and megaureter. The ureter appeared to open to the bladder (Figure 1B) on IVP images. A possible vesicovaginal fistula was excluded using the methylene blue test through the PNC. Then, percutaneous nephrostography done and it was suggested that the megaureter was opening into the bladder (Figure 1C). Urodynamic examination revealed normal findings. On cystoscopy, left ureter orifice and left hemi-trigone could not be seen. The left ureteral orifice was observed in urethra with methylene blue test. Left kidney urine creatinine concentration and osmolality were normal. Following the disappearance of the acute symptoms and preoperative studies yielding normal results, the informed consent was obtained from the patient and she underwent an ureteroneocystostomy with tailoring (Figure 2A). The postoperative course was uncomplicated, and the patient had a satisfactory urinary continence. The follow-up US was performed one month after surgery showed a mild dilatation at the left kidney. A technetium-99m mercaptoacetyl triglycine (MAG-3) scan performed after six weeks demonstrated satisfactory drainage of all functioning systems.

DISCUSSION

Congenital abnormalities of the kidney and urinary tract are a group of diseases with a diverse anatomical spectrum, including kidney anomalies such as multicystic dysplastic kidney, hypoplastic kidney, ureteropelvic junction obstruction, vesicoureteral reflux and megaureter. The ureteric bud is formed by an out-pouching that arises from distal mesonephric duct at the fourth gestational week. Any variation in the origin of the ureteric bud will result in an anomalous position of the ureteral orifice. Therefore, the ureteral orifice is considered ectopic when it drains in a location different to its normal trigonal position. Megaureter is a generic term indicating the presence of an enlarged ureter with or without concomitant dilatation of upper collecting system. In practice, a ureter with a diameter of 7 mm or more should be considered as a megaureter.[6] Megaureter may be primary or secondary, refluxing or nonrefluxing, obstructed or unobstructed, and nonrefluxing unobstructed.[5] In our case, there was an ectopic megaureter (13 mm) draining into urethra and a single collecting system with pyonephrosis.

The cause of pyonephrosis and megaureter associated with EU opening to the urethra is the stricture at the ectopic ureteral orifice on the urethra. It is a fundamental difference between female and male ureteral ectopia and the probability of in-

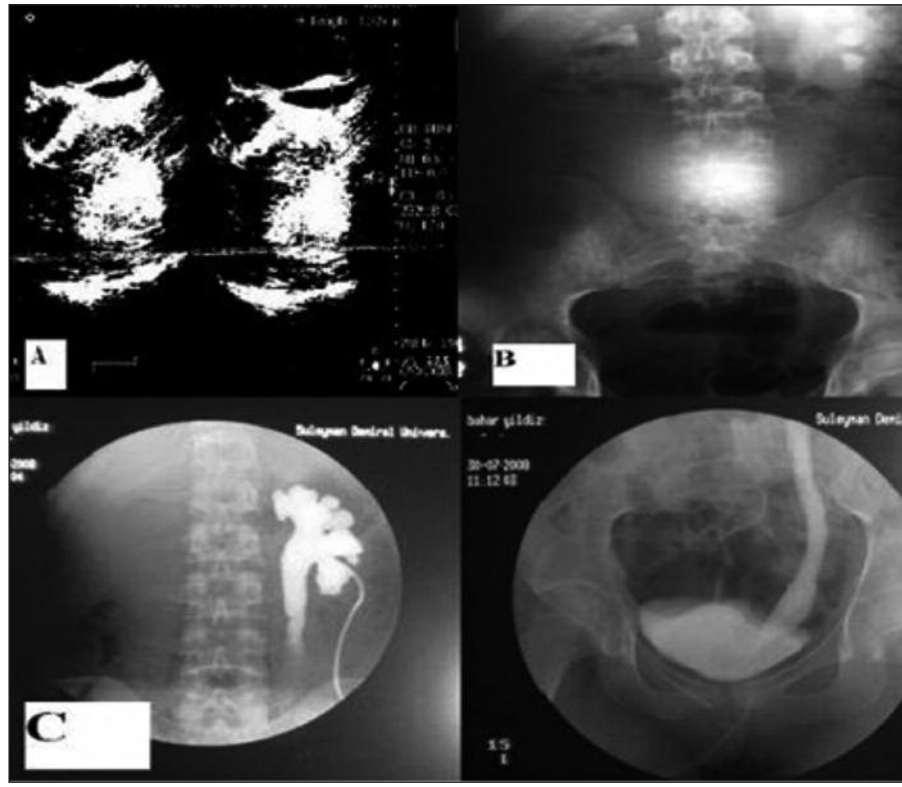

FIGURE 1: A Transverse US image through the bladder shows dilatation of left ureter (distal ureter width: 13 mm). **B** IVP revealed a hydronephrosis at left kidney with single collecting system. **C** Nephrostography shows the megaureter with hydroureteronephrosis at left kidney and it seems to be open the bladder.

continence is more common in females, because EU may more likely terminate at a level distal to continence mechanisms consisting of bladder neck and external sphincter. Similarly in our case, when we performed cystourethroscopy, we observed that EU orifice was opening to the mid-urethra. An EU may present with a single collecting system, however, about 70-80% of them are associated with complete ureteral duplication.[7] In our case, the patient had dribbling incontinence since her childhood. Classic clinical presentations of this condition include urinary infection as the main symptom, hydronephrosis and pyonephrosis. Pyonephrosis is the end-result of purulent material concentrating at pyelocaliceal level due to the ectopic meatus being obstructed. This may subsequently cause renal parenchyma destruction and urinary incontinence.[8] As in localization, clinical presentation varies in each gender, as well. Girls with EU usually suffer from recurrent urinary tract

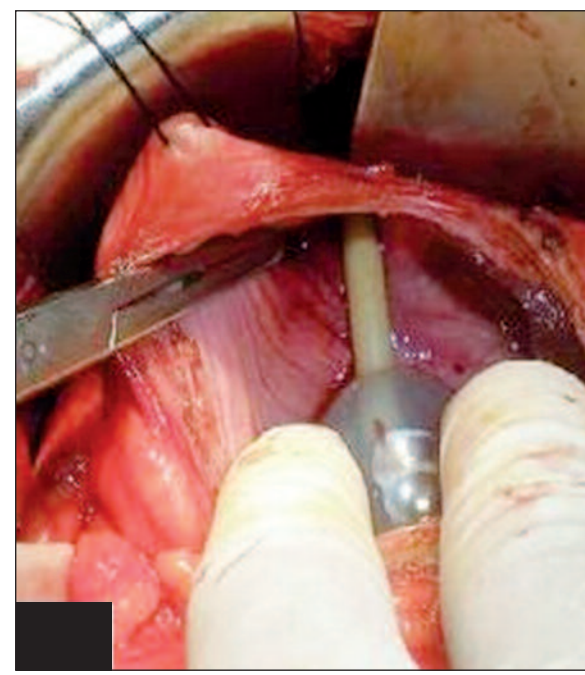

FIGURE 2: Photograph of the ectopic mega ureter that drain into the urethra. A right angle clamp is inserted in ectopic ureter orifice.
(See for colored form http://tipbilimleri.turkiyeklinikleri.com/)

infections, permanent urinary incontinence, vaginal discharge or colic pain. Treatment options depend on the degree of impairment of the renal function. If renal functions are normal, more conservative techniques, such as ureterovesical reimplantation, pyelo-pyelic anastomosis with ligation of the redundant ureter, uretero-ureteral anastomosis or endoscopic dilatation of obstructed ureteral orifice can be attempted. The result of surgery is very gratifying in unilateral EU as the chance to achieve complete dryness is very high. The same is not true for bilateral cases due to abnormal trigone, small bladder, and insufficiency of the bladder neck. Transient residual symptom of wetting after surgery for a single system EU has been reported.[9] In our case, postoperative course was uncomplicated, and the patient had a satisfactory urinary continence. The most important determinant recognized for continence is the integrity of the bladder neck. If the ureter is draining a functioning kidney, reimplantation is the treatment of choice, like our case. A MAG-3 scan six weeks after surgery demonstrated satisfactory drainage of all functioning systems.

CONCLUSION

Persistent incontinence after toilet training in young girls and recurrent urinary tract infections should raise suspicion of an EU. Appropriate imaging studies should be obtained and carefully interpreted. If an EU is found, surgical reconstruction should be planned depending on the degree of the renal function.

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