Case Report

Unilateral Renal and Spermatic Cord Agenesis Accompanying to the Ureteral Extraperitoneal Inguinal Herniation in An Infant

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ABSTRACT

Ureteroinguinal herniation is a rare event, usually diagnosed during the surgical repair of inguinal hernias and most reported cases have occurred in obese men during the fifth and sixth decades of life. Here, we describe the first case of unilateral renal and spermatic cord agenesis accompanying to the ureteral inguinal herniation in an infant.

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Introduction

Inguinal herniation of the ureter is a rare event even in infants and is mainly observed in obese middle-aged men. It is important to recognize in order to prevent accidental ureteral damage during inguinal hernia repair, which is a known complication [1]. Furthermore, symptomatic hydronephrosis has been reported [1]. Here, we report the case of a male infant with renal and spermatic cord agenesis caused by this condition. Consent from the parents has been obtained.

Case Report

A 2-month-old boy, antenatally diagnosed with left hydronephrosis and right renal agenesis, was seen firstly at the pediatric nephrology polyclinic. The right kidney was not observed (agenesis?) with ultrasound (USG), and the left unit was reported as hydroureteronephrosis with AP diameter: 9 mm and distal ureter diameter: 10 mm. A dilated (depending on grade IV-V vesicoureteral reflux (VUR)) right ureter, extending to the right anterolateral side of the bladder, was seen with the Voiding cystourethrography (VCUG) (Figure 1). MR showed that left kidney is in normal localization with a mild hydronephrosis (AP: 10 mm, ureter: 11mm), and the right kidney is not visible probably due to atrophy prenatally (Figure 2). Despite all investigations, the anatomy could not be clearly demonstrated. Exploration for the right inguinal canal was planned, and after treatment of the pyelonephritis he was operated. We performed cystoscopy and saw that the right ureteral orifice very broad and in out of the normal localization. Conversely, urethra, verumontanum and bladder were normal, and the left ureteral orifice settled in normal but very narrow to guide. Then, the right inguinal canal was observed via a right inguinal incision. Hernia and dilated (measured 2.5cm in diameter) ureter were seen. High ligation hemiectomy was performed. We gave some contrast agent to the dilated and thick-walled tissue to confirm that it is ureter or not and we proved it by seeing the contrast passed to the bladder. It was identified that there was not any kidney tissue by following the ureter. The ureter was ending at epididymis without any spermatic cord that was evaluated as cord agenesis. After separating the ureter from the epididymis, we resected the tract totally from the entrance of the bladder (Figure 3). Pathologic evaluation revealed no kidney parenchyma, no spermatic cord tissue but ureter with squamous metaplasia. The postoperative course was uneventful.
Figure 1: Voiding cystourethography (VSUG).
A tubular construct compatible with the atrophic kidney and ureter, extending to the right inguinal canal, was found to be filled with contrast material.

a) continuous arrow inguinal canal, intermittent arrow atrophic kidney, thick short arrow bladder.
b) continuous arrow ureter, cut arrow atrophic kidney, thick short arrow bladder.

Figure 2: Mr Urography.
The left kidney is in normal localization and mild hydroureteronephrosis is mentioned. Right kidney is not visible.

a. left kidney
b. left ureter
c. bladder
d. right ureter
e. atrophic / pouch right kidney

Discussion
Renal agenesis, spermatic cord agenesis (ureter ending as epididymis), extraperitoneal inguinal ureter herniation, age of the patient (diagnosed at 1 month, operated as 2-month-old), and pathology (squamous metaplasia that is known to develop cancer in the long-term period) were the different characteristics that enable us to present our patient. Two types of uretero-inguinal hernias were identified from an anatomical and pathogenic standpoint: 1- paraperitoneal (more frequent, acquired, and always presenting with a peritoneal hernia sac, frequently associated with other herniated abdominal structures), 2- extraperitoneal (very uncommon, congenital, never associated with a true peritoneal sac, always composed only of the ureter) [2]. Paraperitoneal variants (80%) are more frequent in obese men during fifth to sixth decades of life and can rarely include the ureter alone [3]. In contrast, the extraperitoneal type is considered a congenital embryonic defect that results in fusion between the ureter and the genitourinary ligaments due to failure of separation of the ureteric bud from the Wolffian duct. Both of which are then drawn down to the scrotum to form the epididymis and vas deferens [4]. The ureter is dragged into the inguinoscrotal region during physiologic descent of the testes. Age at diagnosis is variable, and on physical examination, extraperitoneal ureteroinguinal hernias are usually small in size, difficult to palpate, and irreducible [3]. Although ureteral herniation in infants is very rare, VUR is fairly common. Preventing pyelonephritis and subsequent renal scarring is essential [1]. Our patient was diagnosed while investigating urinary tract in the postnatal period because of prenatal right renal agenesis. Despite all the detailed investigations, he could not diagnose accurately. Finally, we operated our patient for two reasons: precise diagnose and recurrent infections. Development of the renal system is closely integrated with the development of genital system [5]. Forty-six percent of the patients have an associated anatomical urinary tract malformation, the most common being crossed fused ectopia and nephrophtosis [6]. The unilateral renal agenesis, usually diagnosed incidentally during a radiological examination, occurs in 0.93 to 1.8 per 1000 autopsies [7]. Genital anomalies associated with unilateral renal agenesis occur in 37-60% of women and 12% of men. Congenital absence of the vas deferens can be either unilateral or bilateral. The exact pathophysiology still remains poorly understood [5]. We detected ipsilateral renal and spermatic cord agenesis in our patient.

Conclusion
Even though ureteral herniation in infants occurs sporadically, it is important to recognize it to prevent both recurrent infections and ureteral damage (during inguinal hernia repair). If the ureter has no deformity, the herniation can be corrected with retroperitoneal replacement, but as in our case when renal dysfunction or agenesis detected, totally ureter resection is necessary.

REFERENCES