JOMH
Journal of Men's Health

J. Mens. Health **2022**; 18(2): 031
http://doi.org/10.31083/jomh.2021.088

Case Report

Renal agenesis with ureterocele, duplicated megaureter and translocation of seminal vesicle: a case report and review of the literature

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Submitted: 19 April 2021 Accepted: 10 June 2021 Available online: 6 August 2021 Published: 8 February 2022

Abstract

Background: Renal agenesis is a congenital malformation that occurs due to the inhibition of metanephric blastema induction due to a decrease in ureteric bud activity. Although renal agenesis is not very rare, unilateral renal agenesis with ureterocele occurs rarely, and the coexistance of unilateral renal agenesis, ureterocele, and blind ended proximal ureter is very rare. Recently, we experienced a case of left renal agenesis with huge ureterocele, blind ended proximal ureter, and duplicated ureter on Computed tomography (CT) of a 17-year-old man who visited our emergency department with hematuria. Ureterocelectomy and nephrectomy were performed, and a translocation of seminal vesicle was also observed. This case is a very rare case, so we judged that it may be helpful in making treatment decisions in similar cases later. **Case summary**: A 17-year-old man without specific medical history visited our emergency department with hematuria and voiding difficulty. CT showed left ectopic kidney, megaureter and the blind ended proximal ureter. After ureterocelectomy and nephrectomy, pathological examination revealed seminal vesicles in the periphery of the kidney. After one year, the patient has no complications and no complaining symptoms complaints without any abnormal finding of follow up imaging test. **Conclusions**: This case report focuses on the treatment of renal agenesis with ureterocele, blind ended proximal ureter, duplicated megaureter and translocation of seminal vesicle. This rare case of treatment will be helpful in the determination of treatment for similar cases in the future. To establish standard treatment, data accumulation and well-designed studies are required.

Keywords: Urogenital abnormalities; Renal agenesis; Ureterocele; Seminal vesicles; Case report

1. Introduction

Renal agenesis is a congenital malformation that occurs due to the inhibition of metanephric blastema induction because of a decrease in ureteric bud activity [1,2]. Unilateral renal agenesis is seen in 0.03–0.1% of live births, and is more common on the left side and in males [2]. Unilateral renal agenesis is often discovered accidentally in cases with compensatory hypertrophy of the contralateral kidney [1,3]. In the presence of renal agenesis, there may be additional urologic abnormalities such as vesicoureteral reflux (24–28%), obstructive megaureter (11%), and ureteropelvic junction obstruction (3%) [1,2].

Although renal agenesis is not particularly rare, unilateral renal agenesis with ureterocele is uncommon. The coexistence of unilateral renal agenesis, ureterocele, and a "blind-ended proximal ureter" is very rare; in fact, it has only been reported together with megaureters once in the international literature, according to our literature search [1,4].

Recently, we experienced a case of left renal agenesis with a large ureterocele, blind-ended proximal ureter, and "duplicated megaureter", as revealed by computed tomography (CT) of a 17-year-old male who visited our emergency department with hematuria. We performed ureterocelectomy and nephrectomy. In addition to the other anomalies, a seminal vesicle was also found adjacent to the removed kidney, associated with the developmental abnormalities of this patient. This case report was conducted in accordance with the Declaration of Helsinki. Informed written consent was obtained from the patient for publication of this report and any accompanying images.

2. Case presentation

2.1 Patient information and clinical findings

A 17-year-old male with no specific medical history visited our emergency department with hematuria and voiding difficulty. The patient complained of abdominal disten-

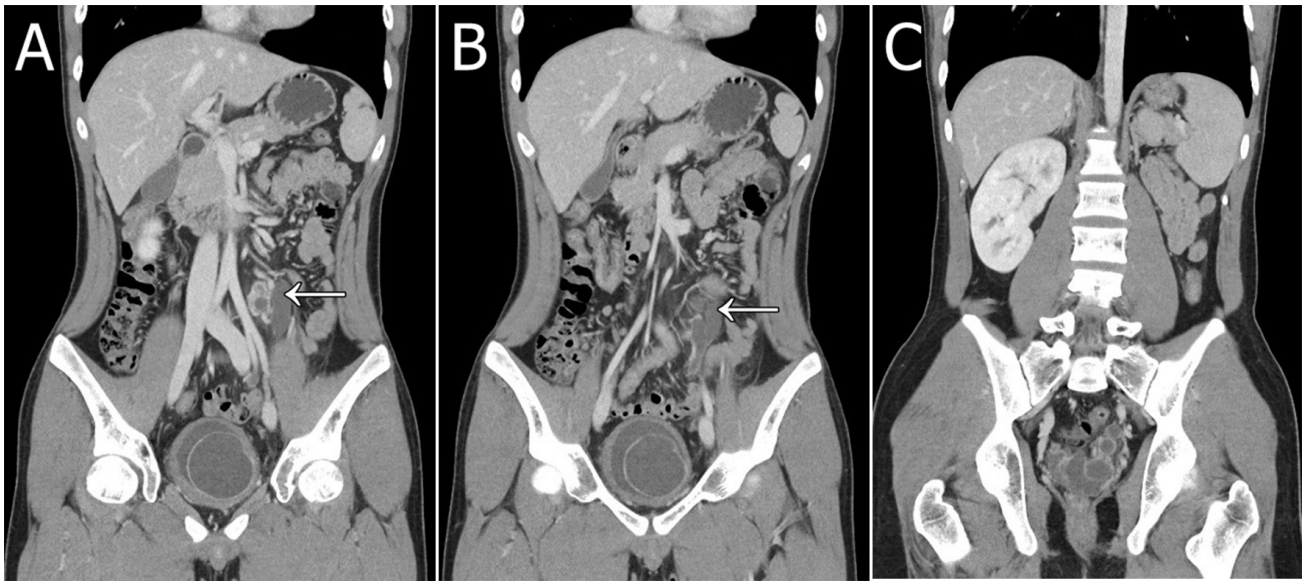

Fig. 1. Preoperative computed tomography. (A,B) A megaureter, duplicated ureter and blind-ended proximal ureter accompanying the left atrophied kidney. (C) The right kidney showed compensatory hypertrophy.

tion and pain. Transurethral catheterization was attempted to relieve the urinary retention, and 400 cc of urine was released. Bladder irrigation was performed and the patient's discomfort and pain improved after a blood clot had been removed. The urology department decided to further evaluate his symptoms.

2.2 Diagnostic assessment

In the complete blood cell count, the red blood cell (RBC) count was 5,170,000/uL (Normal range: 4,000,000–5,300,000/uL), the white blood cell (WBC) count was 14,820/uL (Normal range: 4,000–10,000/uL), and hemoglobin was 15.3 g/dL (Normal range: 13–17 g/dL). Urinalysis showed hematuria. CT revealed a left ectopic kidney with an associated large left ureterocele. The left kidney was not in the normal position; an atrophied kidney was observed in the left lower quadrant. In addition, a dilated left megaureter was found, which was incompletely duplicated with a blind proximal end (Fig. 1), and a 5.7 cm ureterocele was found in the bladder (Fig. 2). The right kidney had compensatory hypertrophy, with no abnormal findings such as hydronephrosis.

2.3 Surgical procedure

Under general anesthesia, the patient was placed in the lithotomy position. The pelvic and inguinal areas were prepped and draped in a conventional sterile manner. A resectoscope was introduced to assess the size and location of the ureterocele in the bladder, and the ureterocele was resected using an electroresection loop (Fig. 3). After electrocoagulation at the bleeding site, a size 18 Fr. Foley catheter was inserted.

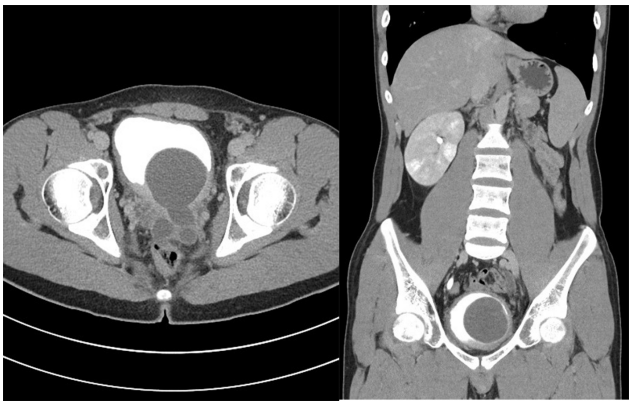

Fig. 2. Preoperative computed tomography showed a 5.7-cm left-sided ureterocele.

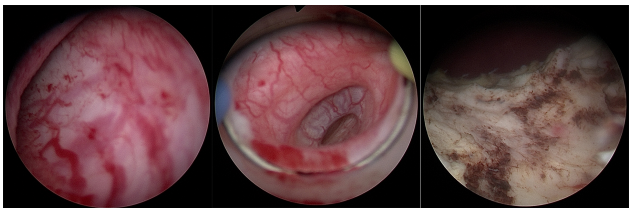

Fig. 3. The ureterocele was resected using a resectoscope.

The patient was then placed in the right lateral position. Following the re-sterilization of abdominal and pelvic area, a laparoscope was inserted into the abdomen through a camera port via a 1-cm supraumbilical incision. After placing the port in the mid-axillary line 5 cm below the xyphoid process, the atrophied left kidney was identified in the left lower quadrant of the abdomen. After dissection of the tissue around the kidney, a cystic mass, thought to

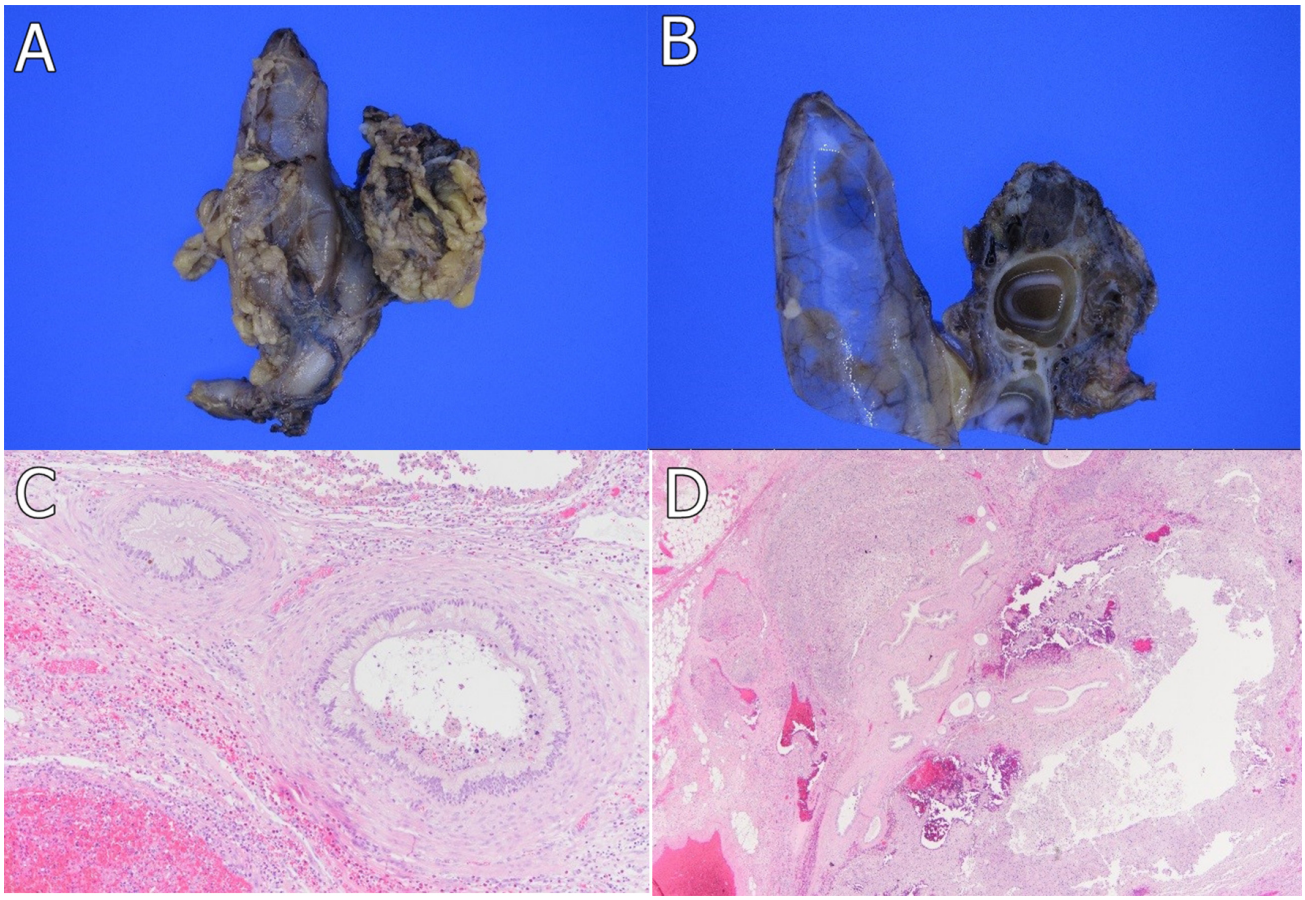

Fig. 4. Pathologic findings. (A,B) Convoluted tubular structures with cystic dilation were observed. (C) Low magnification showed cystic and glandular structures of various sizes. (D) High magnification showed pseudostratified columnar epithelium with semen, including sperm cells.

be atrophied kidney, and the dilated and duplicated ureter were identified, sufficiently dissected, and then clamped and excised. The specimen was removed through an opening extending 5 cm below the umbilical incision. The bleeding site was electrocoagulated. After a Jackson-Pratt drain (200911, Cardinal Health, Waukegan, IL, USA) had been positioned and anchored, all ports were removed. Finally, the skin wound was closed layer by layer.

2.4 Pathologic findings

The transurethral resected ureterocele and removed atrophied kidney were obtained as specimens. The ureterocele specimen showed chronic inflammation with fibrosis, congestion, and hemorrhage. In the nephrectomy specimen, the duplicated ureter and a cystic mass abutting the ureter were found. Histopathology revealed cystic and glandular structures of various sizes under low magnification, and a pseudostratified columnar epithelium with semen, including sperm cells, was observed under high magnification, confirming the presence of seminal vesicle tissue (Fig. 4).

2.5 Outcome and follow-up

The surgical duration was 150 min, and the estimated blood loss was 100 mL. The Jackson-Pratt drain was removed on postoperative day 4, the Foley catheter on day 6, and all stitches on day 12. The patient was discharged on postoperative day 13 with no complications or symptoms. Follow-up CT performed 1 year after surgery showed no specific findings other than thickening of the bladder wall around the left ureterovesical junction where the ureterocele was removed (Fig. 5). We continue to follow the patient.

3. Discussion

A ureterocele is a cystic dilation of the intravesical submucosal ureter; it is common in children, and the prevalence is 4–6 times higher in females. The incidence rate of ureterocele has been reported as 0.08–0.2% [5,6]. Although ureterocele is mostly associated with a duplex kidney, it is occasionally seen in association with a multicystic dysplastic kidney or single urinary collecting system with hydronephrosis. In ureteroceles with complete pyeloureteral duplication, the upper moiety is usually involved [5,7]. If the ureterocele is completely contained within the bladder

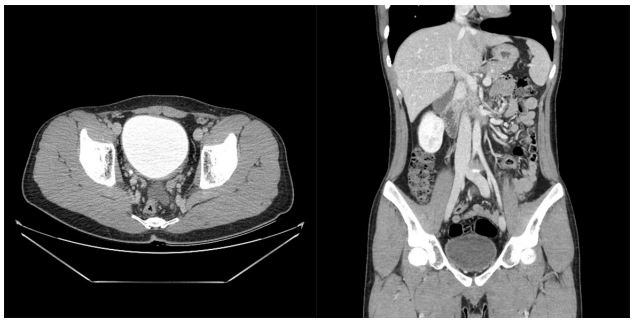

Fig. 5. Computed tomography at 1 year postoperatively. There were no specific findings other than thickening of the bladder wall around the left ureterovesical junction where the ureterocele had been removed.

trigone, it is defined as intravesical, and if part of the ureterocele extends to the posterior urethra or bladder neck, it is defined as ectopic [6,8].

The incidence of ectopic ureterocele in children was reported to be four times higher in girls, left side-dominant, and open to the seminal vesicles in 28% of boys. Cystic dilation of the seminal vesicle is seen occasionally, and the kidney is often non-functioning symptoms [9]. The ureteroceles associated duplex system is usually manifested as a urinary tract infection that develops at a young age, while ureteroceles in adults are usually less obstructive, intravesical, and associated with a single system [9,10]. Endoscopic treatment is effective in relieving the symptoms [9,11].

Treatment of ureterocele aims to preserve maximum renal function while preventing infection, vesicoureteral reflux, and bladder outlet obstruction. Depending on the type, treatments such as simple endoscopic incision, upper pole partial nephrectomy, and complete reconstruction at the bladder level are options. The rate of additional surgery after endoscopic intervention was reported as 7–23%, and 48–100% of ectopic ureteroceles required surgical treatment such as upper pole partial nephrectomy [5].

During embryonic development, the mesonephric blastema remains as a mesonephric duct, which extends to the ureteric bud. Later, the ureteric buds grow toward the metanephric mesenchyme under the influence of mutual induction. The metanephric mesenchyme develops into glomeruli and renal parenchyma, while the collecting system arises from the ureteric bud [12]. In addition, male reproductive organs such as the seminal vesicles, epididymis, and vas deferens develop from the mesonephric duct [13]. Since the urinary and male reproductive systems go through similar developmental processes, it is not uncommon for abnormalities to occur together in both systems [12]. Renal agenesis, one of various urinary abnormalities, is generally thought to occur due to a lack of any induction effect between mesonephric blastema and ureteric buds, caused by a developmental problem of the mesonephric duct during embryonic development. Due to their embryological similarity, unilateral renal agenesis may be associated with ipsilateral urogenital anomalies [1].

Alcaraz *et al*. [14] suggested through experiments using human embryos and rat embryos that the ureter undergoes a process of transient ureter luminal obstruction and recanalization during ureteral development before and after 40 days of pregnancy. In addition, among several genes causing kidney and ureteral anomalies, EYA1 was associated with a dominantly inherited disorder associated with the duplex collecting system, renal dysplasia, and renal agenesis [15]. Based on these theories, it can be inferred that the renal agenesis, duplicated ureter and blind-ended proximal ureter in this case were caused by a problem in the development process. Also, since the seminal vesicle sprouts from the nephric duct from which the ureter is derived, it can be seen that the ureteral anomaly accompanied by the dislocation of the seminal vesicle is due to a developmental problem [16].

Merlini *et al*. reported that the goal of ureterocele diagnosis was confirmation of the ureterocele, vesicoureteral reflux, and any contralateral malformation. Intravenous pyelography (IVP), CT scan, ultrasound of the urinary tract, and voiding cystourethrogram are used for diagnosis. Simple endoscopic puncture is advocated as emergency treatment for a ureterocele, with the possibility of additional surgery in the future [5]. Hasbani *et al*. reported a case of renal agenesis associated with a contralateral ectopic ureter in a healthy 16-year-old male complaining of sudden right flank pain. Renal agenesis was confirmed through abdominal pelvic CT. Infection control and follow-up were planned, and the etiology was not defined [2]. Ahmed *et al*. reported an ipsilateral renal agenesis with megaureter and blind-ended proximal ureter and ureterocele in a 30-year-old male, evaluated by IVP and CT. In this renal agenesis case, it was reported that early evaluation was necessary to reduce morbidity, and surgery to remove the left ureter was performed [1].

Although similar cases have been reported, in our patient, renal agenesis was accompanied by a ureterocele, blind-ended proximal ureter, duplicated megaureter, and translocated seminal vesicle. The translocation of the seminal vesicle was thought to be due to an abnormality in the ascent of the kidney, a process occurring during embryonic development in which the kidney rises from the level of the sacral vertebrae to the level of the first lumbar vertebra. However, the translocation of seminal vesicles was unexpected and further discussion is needed on the etiology. This case report focuses on the evaluation and treatment of a rare renal agenesis combination, which may facilitate treatment decisions in similar cases. The patient has experienced no complications during follow-up. The ejaculation disorder was explained, and semen analysis is planned in the future. Further studies and more data will be needed to establish standard treatments for future cases.

4. Conclusions

This case report focuses on the treatment of a patient with renal agenesis accompanied by a large ureterocele, blind-ended proximal ureter, duplicated megaureter, and translocation of the seminal vesicle. The embryological factors that induced the occurrence of this rare combination of conditions remain to be elucidated. However, this report should aid treatment decision-making for similar cases in the future. To establish a standard treatment, additional data are required.

Abbreviations

CT, computed tomography; RBC, red blood cell; WBC, white blood cell; IVP, Intravenous pyelography.

Author contributions

JJP: Conceptualization, Data curation, Methodology, Validation, Investigation, Writing-original draft. WBK, KWL, JMK, YHK, AM, JHK, SHK: Data curation, Validation, Investigation. SWL: Conceptualization, Data curation, Methodology, Validation, Investigation, Writing-original review & editing. All authors read and approved the final manuscript.

Ethics approval and consent to participate

This study was conducted as a retrospective chart review. Informed written consent was obtained from the patient for publication of this report and any accompanying images.

Acknowledgment

This manuscript was prepared with the assistance of Textcheck, an English-language scientific editing company.

Funding

This work was supported by Soonchunhyang University Research Fund.

Conflict of interest

The authors declare no conflict of interest.

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