

A 52-Year-Old Male with Bilaterally Duplicated Collecting Systems with Obstructing Ureteral Stones: A Case Report

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Key Words

Congenital abnormalities • Kidney tubules •
Ureteral obstruction • Vesico-ureteral reflux

Abstract

Collecting system duplication is marked by a variety of clinical syndromes. Bilateral and obstructed duplicated systems, particularly with asymmetric levels of duplication, are rare and typically due to ureteric bud development anomalies. The infrequency with which this condition exists makes it a formidable challenge for physicians and patients. To our knowledge, we present the first case report of bilateral obstruction of bilaterally duplicated collecting systems. In our case, a 52-year-old male complaining of low back pain, constipation, urinary urgency and hematuria was found to have bilateral obstructing stones as well as asymmetrical bilateral collecting system duplication. We discuss the natural history of this condition, its consequences and identification.

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Introduction

Bilateral collecting system duplication is a rare congenital genitourinary abnormality involving splitting of the ureteric bud. It presents with a variety of complaints. In our knowledge, we report the first known case of bilateral obstructing stones in a bilaterally duplicated system

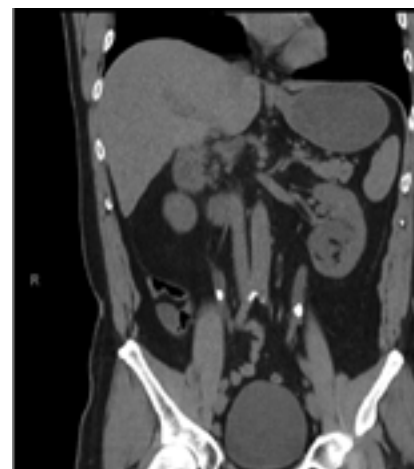


Fig. 1. CT scan of abdomen and pelvis which identified multiple bilateral renal collecting system calculi.

and present its characteristic urological and radiological findings.

Case Report

A 52-year-old Caucasian man presented to the emergency department with increasing left low back pain with radiation into his leg, hematuria, urinary urgency and constipation. He denied flank pain. His past medical history is significant for nephrolithiasis, lumbar radiculopathy, coronary artery disease, multiple sclerosis, hyperlipidemia, hypertension and a Meckel's diverticulum. His

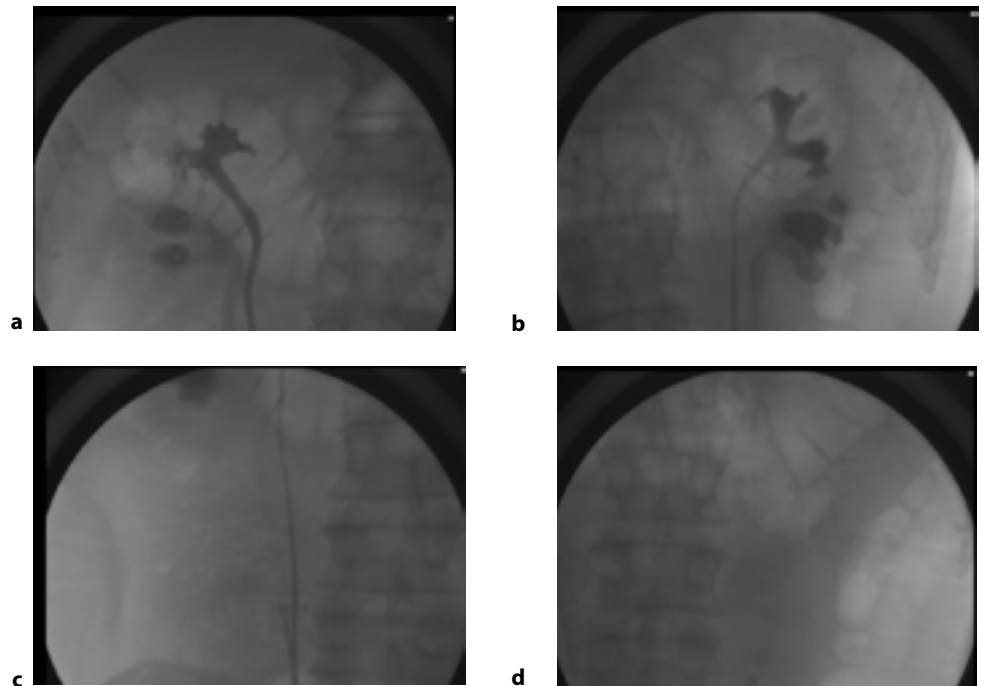


Fig. 2. Bilateral duplicated collecting systems with fully duplicated ureters on the left (**b, d**) and partially duplicated ureters on the right (**a, c**).

family and social history is non-contributory. He was found to be afebrile with a heart rate of 125 bpm, blood pressure of 148/98 mmHg, respiratory rate of 20/minute and urine output of 1 l/d. His abdomen was unremarkable and no costovertebral angle tenderness was appreciated. However, he exhibited low back pain and reproducible sciatica. Laboratory analysis revealed a white blood cell count of 10,000/ μ l, hemoglobin of 13.9 g/dl, creatinine of 1.29 mg/dl, BUN of 21 mg/dl, white blood cells 3–5/hpf and red blood cells 0–2/hpf with a negative nitrite in urine test. He underwent a non-contrast CT scan of his abdomen and pelvis which identified multiple bilateral renal collecting system calculi (fig. 1), the largest of which were 12 x 10 mm in the right upper pole and 10 mm in the left lower pole. The scan was also significant for bilateral duplicated collecting systems with fully duplicated ureters on the left (fig. 2b, 2d) and partially duplicated ureters on the right (fig. 2a, 2c). Furthermore, the radiologist reported moderate right lower pole hydronephrosis and hydroureter with an obstructing calculus measuring 6 mm and moderate to severe left kidney lower pole hydronephrosis and hydroureter with an obstructing 8 mm calculus in the mid-ureter (fig. 1, 2). Bilateral retrograde pyelograms were done and bilateral ureteric stents were placed in the operating room. Calculi were noted in the distal ureters bilaterally with accompanying hydronephrosis, especially on the left. During cystoscopy, 2 ureteral orifices were noted on the left and one on the right. Following the operation, excessive low back pain continued. He was admitted and placed on 0.4 mg Flomax at bedtime.

Upon discharge, he was placed on ciprofloxacin and narcotic analgesics and scheduled for lithotripsy. His pain improved but his creatinine remained 1.29 mg/dl.

Discussion

A duplex system refers to a kidney with two pelvicaliceal regions, each of which has a descending ureter. These ureters may join to form a partially duplicated ureter or remain fully separated, representing a complete duplication. Unilateral ureteral duplication has been found in up to 0.8% of the United States population at autopsy while bilateral duplication represents 20–40% of duplications, or 0.16–0.32% of the population [1]. Both subtypes are most commonly the result of premature splitting of the ureteric bud, a remnant of the Wolffian duct [2]. Because partial duplication is typically observed in metanephric tissue that has not separated fully, such systems share several lobes and have overlapping collecting tubules [2]. Rarely, complete duplication may result from having 2 distinct ureteric buds [3]. A complete duplication is one

third as common as partial duplication and has no shared aspects [3]. Duplication is believed to be autosomal dominant in inheritance with incomplete penetrance [4] and have highest prevalence in Caucasian females [5].

Presentation of duplication is variable and highly dependent on age. Approximately half of patients with duplication identified prenatally will endure repeated urinary tract infections and about 10% of children with such infections have this condition [5]. Accordingly, recurrent pediatric urinary tract infections may suggest duplication. Adults, similarly, are prone to obstruction, vesicoureteral reflux and recurrent infections. They may also experience hematuria, abdominal or flank pain. Complete duplication may result in an ectopic ureter implanted into the vagina, urethra, epididymal region or vestibule and can cause ureteroceles [6]. In complete duplication, normalized pelvicaliceal systems typically indicate that the ureteral orifice will be in the anatomically correct position [7]. Insertion of the upper pole moiety's ureter follows the Weigert-Meyer rule and is inferior and medial as compared to the lower pole moiety's ureter. However, incomplete duplication is associated with ureteroureteral reflux, vesicoureteral reflux and ureteropelvic junction obstruction with regards only to the lower pole [7]. Vesicoureteral reflux can alleviate with age [7].

With the exception of ureteropelvic junction obstruction, all complications are more common in females and must be identified [7].

Prenatal duplications are commonly identified by their dual pelvices or hydronephrosis, particularly in the lower pole if due to obstruction of the ureteropelvic junction [7]. After birth, ultrasound remains an excellent modality. Unilateral duplication may be evident through asymmetry [8]. Segmentally isolated hydroureter should raise suspicion for incomplete duplication [7] while hydronephrosis that is not uniform between upper and lower poles strongly indicates a complete duplication and uniformity is inconclusive [8]. Complete duplication without obstruction may evade ultrasound detection [7]. Voiding cystourethrography may identify lower pole reflux, an abnormal axis or 'drooping lilly sign', or decreased number of lower pole calices [7]. Longer fluoroscopic studies should be considered if reflux is transient. On excretory urography, reflux will be seen and the upper pole may be displaced [7]. Ureteroceles can be visualized by noting dilation of the upper pole ureter and calices above it with a filling defect in the bladder [7]. If a diagnosis of complete versus partial duplication remains elusive, the level of convergence between the ureters may be enunciated by the use of intravenous pyelography [7].

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