Journal of Pediatric Surgery Case Reports 89 (2023) 102561

Contents lists available at ScienceDirect

Journal of Pediatric Surgery Case Reports

journal homepage: www.elsevier.com/locate/epsc

A rare association between crossed fused renal ectopia, urethral stricture, bilateral cryptorchidism, and sub-coronal hypospadias in a non-syndromic 6-year-old child

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ARTICLE INFO

Keywords:
Kidney
Urinary tract
Nephrology
Urology
Congenital kidney anomalies

ABSTRACT

Introduction: From asymptomatic ectopic kidneys to potentially fatal renal agenesis, congenital abnormalities of the kidney and urinary tract (CAKUT) encompass a wide variety of anomalies (bilateral). With varied degrees of severity, many of them are found in the antenatal or immediately postnatal period, with a sizable fraction also found in the adult population. On the basis of embryology, CAKUT can be divided into three categories: defects in the formation of the renal parenchyma, aberrant embryonic migration, and abnormalities of the collecting system. Multiple cystic dysplastic kidneys, renal hypoplasia, number (agenesis or supernumerary), form, and cystic renal illnesses are examples of renal parenchymal abnormalities.

Case presentation: A 6-year-old male patient was referred to the hospital complaining of painful and difficult urination. Past medical history was significant for recurrent urinary tract infections, with temporary improvement with antibiotic therapy. Physical assessment and imaging evaluation revealed a constellation of genitourinary anomalies, including sub-coronal hypospadias, penile urethral stricture, bilateral cryptorchidism, and a crossed fused ectopic left kidney.

Discussion and conclusion: Congenital kidney and urinary tract anomalies are a group of several diseases that affect the kidney and/or urinary tract. Despite substantial differences in phenotypic and clinical consequences, CAKUT has a similar genetic foundation and shares molecular signals that influence kidney development. The prognosis and quality of life for affected families have improved thanks to advancements in prenatal diagnostics, imaging, genetic testing, laboratory surveillance, and medical management, as many of these congenital defects are inherited.

1. Introduction

Congenital anomalies of the kidney and urinary tract (CAKUT) encompass a wide range of conditions brought on by abnormalities in the development of the lower urinary tract, the urine collecting system, the kidney(s)' embryonic migration, or the development of the kidney parenchyma. About 50% of the individuals with the condition have lower urinary abnormalities, which include vesi-

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https://doi.org/10.1016/j.epsc.2022.102561
Received 11 December 2022; Accepted 20 December 2022
Available online 26 December 2022

coureteral reflux (25%), ureteropelvic junction blockage (11%), and ureterovesical junction obstruction (11%). 20–30% of all detectable defects are kidney malformations, which are frequently found in the prenatal period [1,2]. These problems affect about 2% of all pregnancies and are frequently linked to other developmental defects or hereditary syndromes. Severity can range from moderate prenatal pelvictasis to bilateral renal agenesis [3]. According to data from sizable registries, CAKUT accounts for 30–50% of end-stage renal disease and is a significant cause of morbidity in the patient populations of children and adolescents [2]. Children with CAKUT are diagnosed and treated in a variety of clinical settings, such as prenatal obstetrics, pediatric primary care offices, pediatric nephrology clinics, and pediatric urology centers. CAKUT can cause kidney issues in adults if it is not caught in childhood. These include renal insufficiency, proteinuria, and hypertension, which frequently necessitate referral to adult nephrology [4].

2. Case presentation

A 6-year-old male patient who was referred to our hospital complaining of painful and difficult urination. The child's medical history revealed significant recurrent urinary tract infections, which improved temporarily with antibiotic treatment for each episode. The patient's mother mentioned that her child had a noticeable poor stream. He had no prior surgeries. Upon admission, the physical assessment was unremarkable except for suprapubic fullness with mild tenderness, sub-coronal hypospadias, and an empty scrotum. Laboratory evaluation, including a complete blood count, urinalysis, and urine culture, was normal. The patient underwent a retrograde urethrography, which showed a urethral stricture at the level of the penile portion of the urethra (Fig. 1). Ultrasonography was performed and revealed undescended testes along with right testicular hypoplasia compared to the left one (Fig. 2). He also had computed tomography with and without contrast, which revealed a crossed fused ectopic left kidney with normal parenchymal thickness; both ureters are united anterior to the kidney level, seen crossing the midline posterior to the right common artery and inferior vena cava, and inserted into the urinary bladder on the right side (Figs. 3 and 4). The genetic analysis and metabolic panel of the patient were normal. The patient underwent pediatric cystoscopy under general anesthesia with urethral dilatation and insertion of a ure-

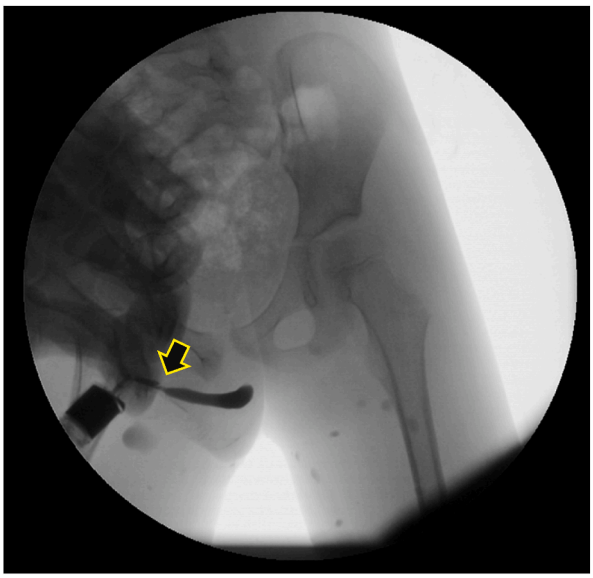

Fig. 1. A retrograde urethrogram showing a penile urethral stricture (**arrow**).

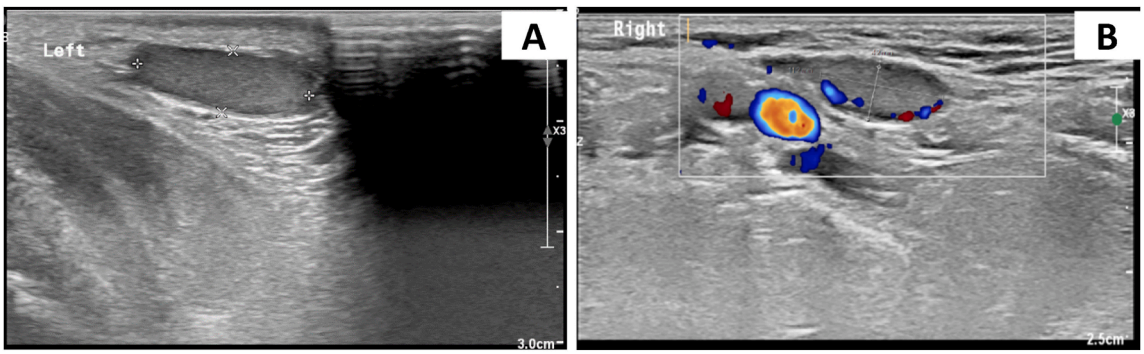

Fig. 2. Shows an ultrasound of **(A)** the left testicle, which is located in the left pre-scrotal region and has normal texture, and **(B)** the right testicle, which is located high in the right inguinal canal and is small in size (1.2 * 0.5 cm) and also has normal texture.

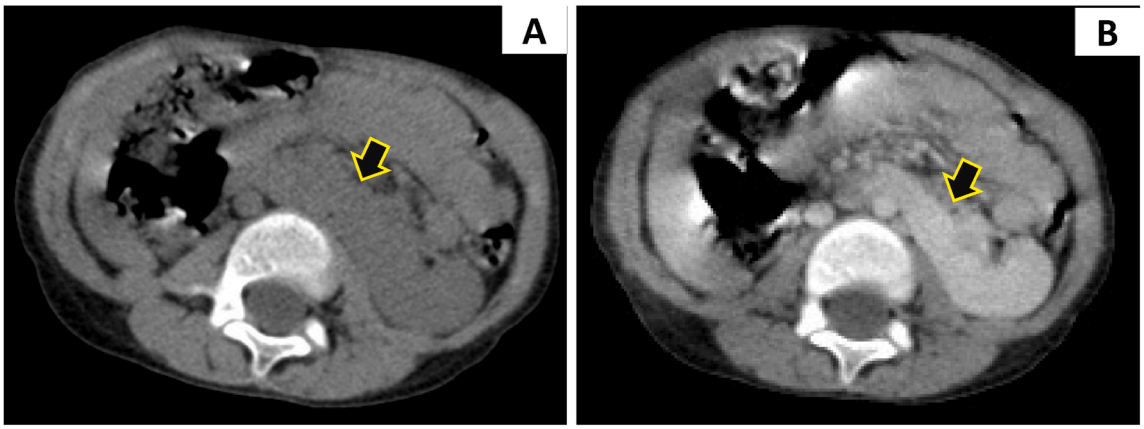

Fig. 3. (A): is an axial non-enhanced computed tomography (CT), **(B):** is an axial enhanced CT, showing a crossed fused ectopic left kidney (**arrows**).

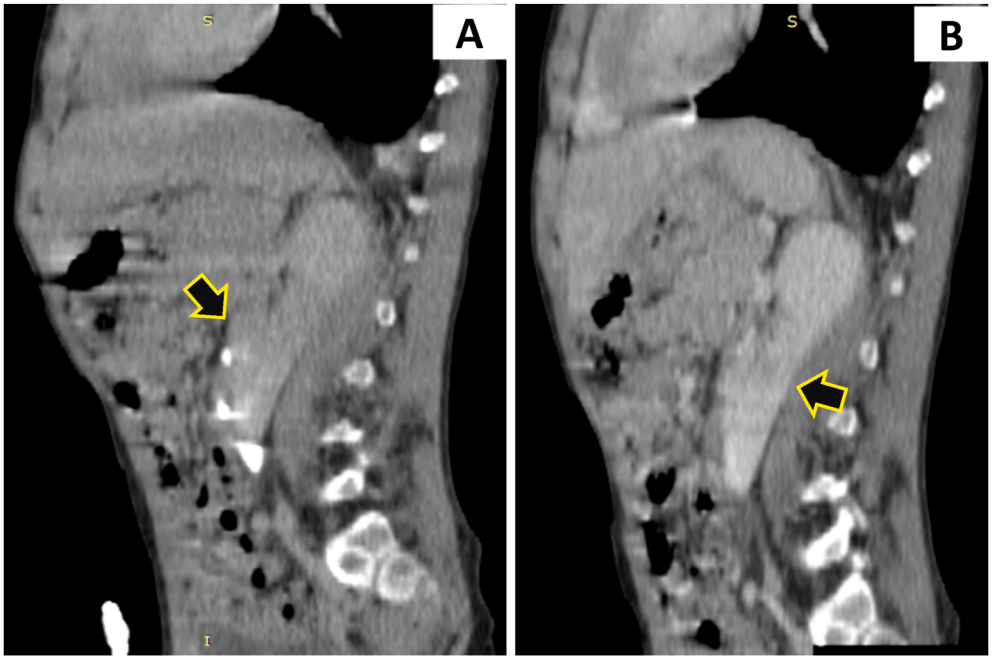

Fig. 4. (A): is a sagittal CT scan with delayed venous phase. **(B)** is a sagittal CT scan with venous phase, showing a crossed fused ectopic left kidney (**arrows**).

thral catheter. The operation went smoothly. Post-operatively, he was transferred to the surgical ward with stable vital signs; he has been started on IV antibiotics and analgesia, and his Foley catheter was clear. The Foleys catheter was removed, the patient voided normally, and he was discharged home in stable condition with recommendations for additional urethroplasty and orchiopexy follow-ups.

3. Discussion

The ascent errors that cause ectopic kidneys, which are frequently located in the pelvis, are. Thoracic kidneys have only a few documented cases [5]. You can have unilateral or bilateral ectopic kidneys. The term “pancake kidney” refers to a midline mass of renal tissue formed when the bilateral pelvic kidneys combine. This mass has two different renal pelvises and a variable number of ureters. An ectopic kidney whose ureter crosses the midline is referred to as a “cross-fused” ectopia. Hypoplastic and smaller ectopic kidneys are common. Thus, compensatory hyperfiltration and hypertrophy may occur in the contralateral kidney [6].

An abnormal constriction of the urethra known as a urethral stricture is caused by fibrosis in the area's corpus spongiosum [7]. Idiopathic causes, such as transurethral resection, urethral catheterization, prostate cancer treatments, and prior hypospadias surgery, are the most frequent causes of urethral strictures, which are then followed by iatrogenic causes [8]. Trauma to the pelvis and perineum is known to result in urethral strictures [7,8]. Patient morbidity, such as urinary tract infections (UTIs), incontinence, and stricture recurrence, increases the expenditures related to urethral strictures [8]. The second most frequent genital birth abnormality in

boys, after cryptorchidism, is hypospadias, a disease where the urethra opens on the bottom of the penis with concomitant ventral penile curvature. Hypospadias treatment is one of the most common surgical operations carried out by pediatric urologists, occurring in one in every 200 live male births [9].

A critical developmental stage on the way to successful human reproduction is the descent of the testes into the scrotum's temperate environment. One of the most prevalent congenital abnormalities in boys is cryptorchidism, or the failure of one or both testes to permanently descend, also known as undescended testis. According to statistics, 1–9% of all males are born with at least one cryptorchid testes [10,11]. While almost half of these cases may spontaneously descend within the first three months of life, towards the conclusion of their first year, only 1% of all men will still be cryptorchid [12]. in absolute terms, only about 2% of males with cryptorchidism who are born will also have hypospadias [13]. Testicular Regression Syndrome, often known as "vanishing testis," is a rare disorder in which the testis disappears as a result of torsion or some other unidentified reason [14].

Depending on the kind, severity, and laterality of the renal abnormality, clinical characteristics can vary greatly. Many of them, including hypoplasia, ectopic kidneys, and form anomalies, can be seen in adults incidentally and without any symptoms. Others, including oligohydramnios, renal agenesis, MCDK, bilateral PUJO, and cystic renal disorders, can manifest early, either antenatally or later, with kidney stones, hematuria, proteinuria, renal impairment, an abdominal mass, or a urinary tract infection [15].

Early in the second trimester of the second trimester, antenatal screening ultrasonography (US) permits the demonstration of fetal kidneys and urine bladder and the detection of a number of significant congenital defects of the urinary system. The presence, location, and size of both kidneys, as well as their structure and echogenicity, are all assessed during an ultrasound examination of the normal urinary tract [16]. The examination is completed by assessing the fetal bladder, external genitalia, and the amount of amniotic fluid. Hydronephrosis, hydroureter, thickened bladder, cystic kidney, tiny or dysplastic kidney, and missing kidney are among the abnormalities found during prenatal ultrasound. A postnatal physical examination and postnatal renal ultrasound should be performed soon after birth and once more at 4–6 weeks of age [16] if renal abnormalities were detected during pregnancy.

4. Conclusion

Ectopic kidneys with no symptoms and life-threatening renal agenesis are only two examples of the large range of disorders that fall under the umbrella of congenital anomalies of the kidneys and urinary tract (bilateral). Due to its accessibility, non-invasive nature, and lack of radiation, ultrasound is frequently utilized as the initial imaging procedure both throughout pregnancy and after delivery. CT and MRI are helpful for confirming an abnormality found on ultrasound, finding complicated abnormalities, showing the anatomy of the collecting system and arteries, and, most critically, for the early diagnosis of problems such as renal calculi, infections, and cancers. Radiologists must be knowledgeable about the imaging characteristics of a wide range of renal abnormalities and their consequences in order to make an accurate diagnosis and choose the best course of treatment.

Author contribution

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Guarantor

Oadi N. Shrateh.

Patient consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Funding

No funding or grant support.

Authorship

All authors attest that they meet the current ICMJE criteria for authorship.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

Acknowledgments

None.

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