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Case Report

Renal agenesis associated with contralateral ectopic ureter and hydroureteronephrosis

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ABSTRACT

Congenital anomalies of the kidney and the urinary tract such as renal agenesis and ectopic ureter have complex development. These anomalies have variable presentations and associations. In this report, we highlight the case of a young man with congenital renal agenesis presenting for a urinary tract infection. Abdominal and pelvic computed tomography imaging revealed the rare association of renal agenesis with contralateral ectopic ureter and subsequent hydroureteronephrosis. A urinary tract infection can be the presenting complication of such association, and a long follow-up is needed to anticipate the management.

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Introduction

The spectrum of congenital anomalies of the kidney and urinary tract is extremely broad and ranges from asymptomatic malformations such as renal agenesis and ectopic ureteral insertion to life-threatening pathologies [1]. Each malformation can have associations with other malformations potentially leading to complications. Several genetic and environmental factors might play a role during kidney embryogenesis contributing to the development of malformations.

Embryologically, failure of ureteric bud activity has been proposed to cause abnormal formation of the ureteric orifice, either as an ectopic ureter or as a laterally displaced ureter with inadequate incorporation into the bladder wall, resulting in vesicoureteral reflux, and thus hydroureteronephrosis [2]. The prevalence of hydroureteronephrosis in adults is poorly reported in the literature. Notably, it often stays silent in adults [3].

Renal agenesis is a missing kidney which results from embryologic lack of initiation of development. The incidence of unilateral renal agenesis is one in 1000 to one in 3000 live births, with male and the left side predominance [4]. In patients with renal agenesis, additional abnormalities of the urinary tract may be present. For example, vesicoureteral reflux has been found in approximately 24% of individuals with unilateral renal agenesis [5].

In this case report, we present a 16-year-old male who presented for a urinary tract infection to be diagnosed with unilateral renal agenesis and contralateral ectopic ureter

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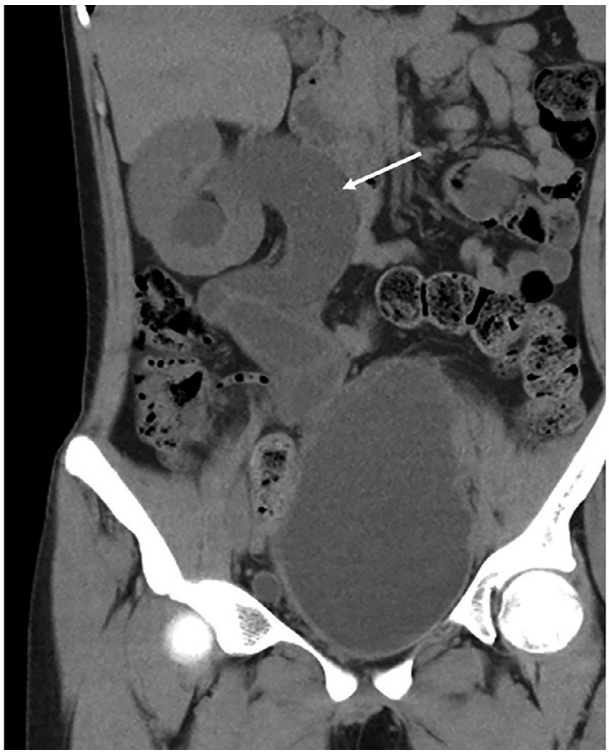

Fig. 1A – Coronal view of a noncontrast abdominal and pelvic CT scan showing an absent left kidney and severe right hydroureteronephrosis (White arrow).

Case report and/or case presentation

A 16-year-old healthy male with congenital left renal agenesis presented to the emergency room with one day history of acute onset right flank pain radiating to his right lower abdominal quadrant. He denied dysuria, fever, chills, nausea, or vomiting. There was no history of trauma or penetrating injuries. The patient stated that his left renal agenesis was diagnosed at around 1 year of age with no known abnormality of the right kidney.

Vital signs were stable on admission. Studies revealed leukocytosis with a WBC count of 15,000 (82% neutrophils) and a serum creatinine of 1.1 mg/dL. Urinalysis was positive for pyuria, leukocyte esterase, nitrite, proteinuria, and hematuria. Urine culture was positive for *Escherichia coli* colonies. The patient was started on 100 mg twice daily for 5 days.

Abdominal and pelvic noncontrast CT scan revealed severe right hydroureteronephrosis consistent with either obstructive process or significant reflux (Fig. 1A). Moreover, it showed that the right ureter was inserting ectopically in a more anterior position than usual (Fig. 1B).

A renal MAG3 scan was not performed. The patient was planned to be observed after recovering from the urinary tract infection.

Discussion and/or conclusion

An interplay of genetic and environmental factors can lead to congenital anomalies of the kidney and urinary tract. For instance, the absence of transcription factor Islet1 can lead to ectopic branching of the ureteric bud out from the nephric duct or to the formation of accessory buds, both of which could lead to obstruction of the ureter-bladder junction and consequent hydroureteronephrosis [6].

and hydroureteronephrosis by a noncontrast abdominal and pelvic computed tomography (CT) scan.

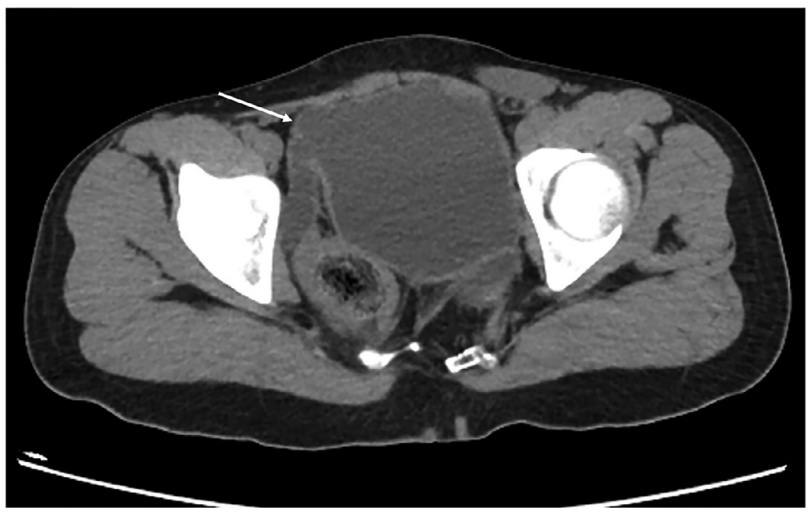

Fig. 1B – Axial view of a noncontrast abdominal and pelvic CT scan showing severe right hydroureter, with the ureter inserting ectopically in a more anterior position than usual (White arrow).

The association between renal agenesis and contralateral ectopic ureter and hydronephrosis is uncommonly described in the literature. In 1988, Muren and Wikstad described case histories of 6 children with absence of functioning renal parenchyma on 1 side and dilatation of the contralateral pelvis [7]. A comparative study assessing 19 cases of single system ectopic ureters found that 26.3% of the cohort had unilateral or contralateral renal agenesis [8]. Two other cases of children with unilateral single ectopic ureters and contralateral renal agenesis were also reported [9].

Other anomalies that are commonly associated with renal agenesis include vesicoureteral reflux and megaureter [10]. Less commonly, renal agenesis can be associated with ureterocele and posterior urtethral valve [10]. Interestingly, one thrid of patients of patients with renal agenesis can undergo a urinary tract infection [10].

Patients with renal agenesis present in variable ways. However, 66.9% of patients are asymptomatic [4]. Only 3.4% of patients with renal agenesis present for a urinary tract infection [4]. Subsequently, 25.4% of patients develop renal insuficiency [4].

Contrast enhanced CT abdomen with arterial, portal, and excretory phase images could identify different kinds of urinary tract anomalies, such as renal agenesis, as well as other associated intra-abdominal anomalies [11]. Besides, renal agenesis can be confirmed by ultrasound, with empty renal fossae and absent bladder filling [12]. In addition, the lack of recognisable renal arteries on color Doppler would strongly support the suspicion of renal agenesis [13]. Regarding the work-up of ectopic ureter, a renal ultrasound and/or computerized tomography is essential for the diagnosis [11].

This case report draws attention to the association of renal agenesis with contralateral ectopic ureter and the complication of hydroureteronephrosis. The exact genetic and environmental factors leading to such associations are still to be defined.

Patient consent statement

A consent has been obtained from the patient.

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