Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Urology

Coexistence of Right Renal Ectopia and Contralateral Renal Agenesis: A Case Report

Kabamba K. Serge[1*], Mpawenayo J[1], M. Taibou, Kimwanga. S[1], Ngangu. N[1], M. Bennani[1], Y. Daghdagh[1], A. Kbirou[1], A. Moataz[1], M. Dakir[1], A. Debbagh[1], R. Aboutaib[1]

[1]Department of Urology, Ibn Rochd University Hospital, Faculty of Medicine, Hassan II University, Casablanca

DOI: https://doi.org/10.36347/sjmcr.2025.v13i08.012 | **Received:** 02.06.2025 | **Accepted:** 29.07.2025 | **Published:** 07.08.2025

***Corresponding author:** Kabamba K. Serge
Department of Urology, Ibn Rochd University Hospital, Faculty of Medicine, Hassan II University, Casablanca

Abstract **Case Report**

The coexistence of renal agenesis with contralateral renal ectopia is extremely rare: to date, no similar case has been reported in the literature. The ectopic kidney may show dilation of the collecting system. We report here the case of a 17-year-old patient with a dilated and destroyed pelvic ectopic right kidney (hydronephrosis due to ureteropelvic junction obstruction) associated with contralateral renal agenesis, discovered incidentally on ultrasound. Initial management involved close monitoring, but progression to end-stage renal disease due to the destruction of the solitary kidney led to definitive treatment with laparoscopic right nephrectomy. The patient was placed on peritoneal dialysis. Surgical exploration revealed a right renal artery originating from the anterior surface of the terminal aorta.

Keywords: Renal ectopia, renal agenesis, ureteropelvic junction obstruction.

INTRODUCTION

Congenital uropathies (CUs) are the leading cause of chronic kidney disease in children worldwide, highlighting the need for further research to better preserve and improve renal outcomes in children affected by such malformations [1].

Various anomalies can affect the number, position, shape, size, rotation, and migration of the kidney, calyces, ureter, orbladder.

Renal ectopia is a migration anomaly that occurs during embryogenesis. Normally, the kidneys begin to develop in the pelvis and migrate to their final anatomical position in the upper abdomen. This ascent precedes the descent of the gonads into the pelvis. The caudal growth of the embryo appears to contribute to the migration of the kidneys out of the pelvis toward their eventual retroperitoneal location in the renal fossa. They reach their adult position by the 9th week of gestation.

Renal ectopia may be high, low, crossed, fused or unfused, but is rarely bilateral. When isolated, it is not life-threatening. Some urinary tract anomalies may be associated with it, most commonly vesicoureteral reflux and ureteropelvic junction obstruction [2,3].

Renal agenesis results from the failure of the ureteric bud to develop, which leads to the absence of induction of the metanephric blastema. It is a rare malformation, occurring in approximately 1 in 10,000 births. It can be caused by mutations in several genes, including

RET, **BMP4**, **FRAS1**, **UPK3A**, and **PAX2**, among others. When bilateral, renal agenesis is incompatible with life. The clinical implications of renal agenesis vary depending on whether the contralateral kidney is present and on the presence of associated anomalies, which may be urological, genitourinary, or of other systems. Common anomalies associated with renal agenesis include vesicoureteral reflux and megaureter [4,5].

CASE REPORT

A 17-year-old male presented 4 years prior with abdominal pain and dysphagia. Abdominalultrasound revealed a right ectopic kidney and left renal agenesis. He was referred to a pediatric nephrologist and placed under conservative surveillance.

Unfortunately, his condition deteriorated, leading to end-stage renal disease due to a destroyed pelvic ectopic kidney with reduced cortical thickness

(UPJO). He was started on peritoneal dialysis in 2020 and later referred for further management.

The patient had no history of trauma or penetrating injuries. Vital signs were stable on admission. Laboratory findings showed no inflammatory signs (WBC: 57,500/mm³, CRP: 0.5 mg/L). Renal function was impaired (Creatinine: 144 mg/L, Urea: 1.17 g/L).

IMAGING:

CT angiography revealed a right renal artery originating anteriorly from the terminal abdominal aorta, feeding a malrotated ectopic pelvic kidney with significant pyelocaliceal dilatation and reduced cortical index. The left kidney was absent (agenesis).

Figures:

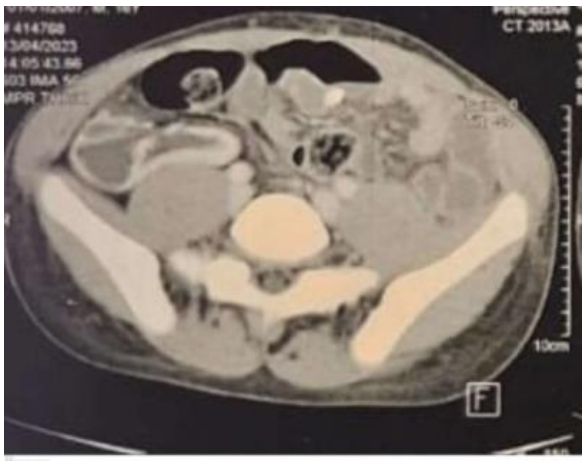

Figure 1: Pelvic CT showing a dilated ectopic kidney with reduced cortical index

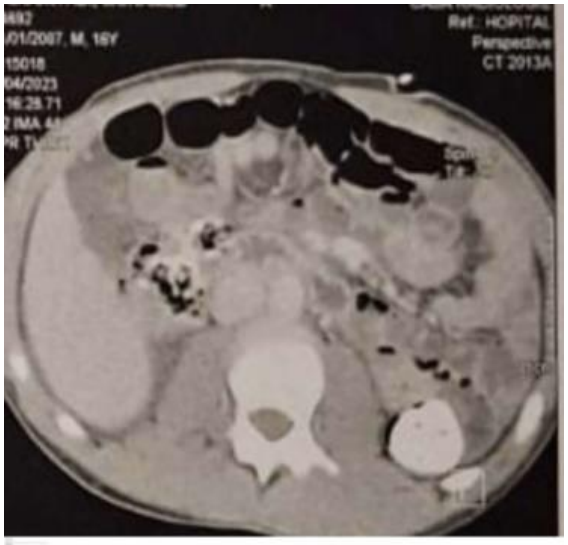

Figure 2: CT scan showing absence of the left kidney (renal agenesis)

Definitive treatment included right laparoscopic ilio-pelvic nephrectomy. Surgical exploration confirmed the presence of a right renal artery originating from the anterior aspect of the terminal aorta.

DISCUSSION

Our 17-year-old patient presented with a non-functional right ectopic kidney located in the pelvis due to ureteropelvic junction obstruction (UPJO), along with an incidentally discovered contralateral renal agenesis. Initial monitoring was implemented; however, as the condition progressed to end-stage renal disease (ESRD) due to destruction of the solitary kidney, the patient was diagnosed with ESRD and initiated on peritoneal dialysis. Definitive treatment consisted of a right nephrectomy via ilio-pelvic laparoscopic approach. Surgical exploration revealed a right renal artery originating from the anterior surface of the terminal aorta.

The combination of renal ectopia with contralateral renal agenesis is exceptionally rare. S. Agbakou *et al.,* reported that 3.4% of UPJO cases occur in ectopic kidneys [6]. However, there is a paucity of literature on the coexistence of renal ectopia and agenesis.

Patients with renal agenesis present with variable clinical features. However, 66.9% remain asymptomatic [4]. Only 3.4% of patients with renal agenesis develop urinary tract infections, while 25.4% eventually progress to renal failure [4].

Ultrasound is the first-line diagnostic tool, highlighting the extent of dilation and cortical atrophy. Its main strength lies in prenatal diagnosis [7]. Magnetic Resonance Imaging (MRI) is an excellent diagnostic tool, allowing for precise morphological and functional assessment [8]. Although it can be used as a first-line modality, in our context its limited availability means it is reserved for cases where CT is contraindicated [9].

A DMSA renal scan (technetium-99m labelled dimercaptosuccinic acid) can be used to assess relative renal function. It also helps to locate an ectopic kidney, particularly when atrophic and poorly functional even with a relative function as low as 5% [10].

The therapeutic management of pathologic pelvic ectopic kidneys depends on the underlying renal pathology and its impact, rather than on the malposition itself [11]. Qian Xu et al. reported in their study that 11.9% of patients had additional anomalies [12].

A conservative therapeutic approach is increasingly recommended, especially for children with a long-life expectancy, for whom preserving nephron mass is crucial. Currently, it is considered acceptable to preserve a kidney with a relative function greater than 10%, or even less, according to studies by Aziz et al., as there is no strict correlation between the degree of dilation and the severity of obstruction [13,14]. Surgical correction of obstruction, such as pyeloplasty, can be considered for giant hydronephrosis due to UPJO. Studies report functional improvement in over 90% of such kidneys following pyeloplasty [14,15]. In a series of 53 patients, Nerli even proposed a laparoscopic approach—better suited to paediatric patients—with particularly encouraging results [15].

Most children with ESRD begin dialysis and later undergo kidney transplantation.

CONCLUSION

This case highlights the importance of early diagnosis, comprehensive management, and long-term follow-up in individuals with renal ectopia and agenesis.

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