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A rare form of crossed nonfused renal ectopia, multicystic dysplasia: A case report

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ABSTRACT

We report a 6 months-old male who presented a rare phenomenon of nonfused crossed renal ectopia associated with multicystic dysplasia. Male, prenatally diagnosed with cystic lesion in right hypocondria, presented as palpable mass in right upper abdominal quadrant at birth. Detailed radiologic evaluation including laparotomy confirmed a diagnosis of multicystic dysplasia in a crossed non-fused left ectopic kidney and leading to partial right ureteral obstruction, treated by surgical removal of the lesion.

Key Words: Crossed non-fused renal ectopia; multicystic dysplasia.

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Introduction

Crossed renal ectopia is a congenital anomaly consisting of the transposition of a kidney to the opposite side [1], simple renal ectopia represents a defective ascension of the affected kidney during embryogenesis [2]. Less than 10% of the crossed ectopic kidneys are non-fused [2]. Additionally, multicystic dysplasia in a crossed, fused or non-fused, ectopic kidney is rare [3].

The majority of pediatric patients are asymptomatic but non-specific symptoms such as abdominal pain, palpable mass, hematuria and dysuria can be present [4]. In some reported cases the patient develops additional symptoms such as; infection, renal calculi or urinary obstruction [1]. Up to 30% of cases can be incidentally diagnosed [5].

Herein is reported a rare form of nonfused crossed left renal ectopia where the patient

presented a multicystic dysplastic ectopic kidney.

Case report

A 6 months-old male was referred to the Pediatric Urology Department for evaluation of a multicystic dysplastic left kidney. The past story of the patient was as follows: He was diagnosed with a cystic lesion in the left abdomen during 22 weeks gestational ultrasound. As he presented with no other associated anomaly and normal amniotic fluid level, gestation was taken to term. Physical examination after birth was demonstrated a palpable mass in right abdomen, painless and with low mobility. Urinary volume during the first 48 hours of life was normal, as well as renal function laboratory evaluation.

Ultrasound scan of the Urinary Tract was confirmed a multicystic lesion in all right hemiabdomen associated with minimal right pyelocaliceal dilatation. The left kidney was not identified. In order to clarify the anatomy as well as to plan any surgical intervention, magnetic resonance (MR) urography was performed. MR urography showed multicystic left kidney associated with rotation anomaly crossing the midline plan. The left kidney was functionally excluded and was also compressing the proximal right ureter causing a partial obstructive renogram pattern.

At laparotomy, the right kidney was normal and the left kidney was accessed via transperitoneal. The retroperitoneal cystic mass, which was not fused with the right kidney, was resected [**Fig. 1**]. The procedure presented uneventful and the patient was discharged within 24 h. Histopathological examination of the mass revealed multicystic dysplasia of the kidney. Six months after surgery he is asymptomatic, with a control ultrasound showing no dilatation in the right kidney or ureter.

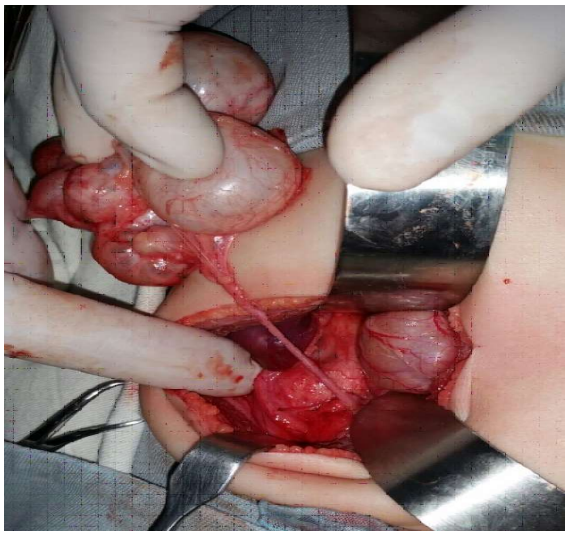

Fig. 1. Multicystic ectopic kidney and ortothopic ureter presenting discrete dilation cause by multicystic crossed kidney.

Discussion

Crossed renal ectopia can be anatomically differentiated into four groups: A) Crossed renal ectopia with fused kidneys (the most common type – 90%), B) Crossed renal ectopia without fusion (rare), C) solitary Crossed renal ectopia (very rare) and D) unfused bilateral Crossed renal ectopia (also very rare) [5]. The ectopic kidney is usually located below the orthotopic kidney and occurs more frequently in males. Left-to-right ectopia represent 50-75% of cases [6].

Diverse associated urological malformations have been described, most commonly vesicoureteral reflux (VUR), but also ureteropelvic junction obstruction, megaureter, hypospadias, cryptorchism, urethral valve and cystic dysplasia [2]. So far, the presence of an ectopic kidney demands investigation for other anomalies, specially VUR [1,4].

Multicystic displasia in a crossed-fused or non-fused ectopic kidney has been rarely described in the literature [3,7]. In this particular case, not only this rare association was detected, but also a partial obstruction of the orthotopic pyelouretheral system.

Imaging investigation is extremely important during the investigation and surgical planning of complex anomalies [8,9]. In this very case, ultrasound study was initially used, followed by voiding urethrocystogram, in order to exclude VUR. A DMSA scintigraphy was able to detect renal exclusion of the multicystic kidney. The use of MR imaging was considered to clearly define anatomy and improve surgical planning. Not used so frequently, especially because of mandatory general anesthesia and contraindication in patient with compromised renal function [4,9], in our opinion this imaging modality was very helpful for the case.

Besides the lack of consensus about surgical treatment of multicystic dysplastic kidney [10], the patient required resection because of the risk for the ortothopic kidney.

Conclusion

Crossed renal ectopia is a rare malformation, varying from 1:20000 to 1:7000 live births. Its association with dysplastic multicystic kidney is even rarer. This paper presents, in our view, the first case with association of both malformations leading to contralateral partial ureter obstruction.

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