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Case Report

Solving the Diagnostic Challenge-Kidney Triplication with Ureterocele: The Indispensable Role of MR Urography

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ABSTRACT

Kidney triplication, also known as ureteral triplication, is a rare congenital anomaly affecting the upper urinary tract. While the precise embryological origin remains uncertain, it is thought to result from multiple ureteric buds arising from the Wolffian duct during fetal development.

We present the case of a young girl with a triplex right renal moiety with an associated ureterocele and Chiari malformation type 1. The diagnosis was established through a comprehensive multi-modality imaging approach, with MR Urography playing a pivotal role in solving the diagnostic challenge. While ultrasound and scintigraphy provided initial insights, only the high-resolution capability of MR Urography allowed for a definitive characterization of the triplex collecting system, offering unparalleled clarity that was essential for accurate diagnosis.

This case underscores the significance of advanced imaging modalities in the diagnosis and management of such complex anomalies to guide clinical decisions effectively.

Keywords: Kidney triplication; Ureteral triplication; MR Urography

Background

Kidney triplication, also known as ureteral triplication, is a rare congenital anomaly affecting the upper urinary tract. While the exact embryological cause remains unclear, it is believed to arise from multiple ureteric buds developing from the Wolffian duct during fetal development[1,2]. This case report of a 6-year-old girl with right renal triplication & ureterocele and Chiari malformation[1], highlights the importance of a thorough radiological workup for definitive diagnosis and guiding treatment decisions.

Clinical Presentation

The patient presented with a 2-year history of bed wetting and recurrent UTIs, which prompted further investigation. A multi-modality imaging approach was employed.

Ultrasound revealed a small sized right kidney with raised parenchymal echogenicity and loss of cortico-medullary differentiation. The right lower ureter was prominent and ended in an ureterocele.

Renal scintigraphy (EC scan) showed a marginally small

right kidney with optimal renal parenchymal function (40 %) and progressive excretion pattern. The left kidney was normal.

Micturating cystourethrogram (MCU) was normal and there was no vesico-ureteric reflux.

MR Urography confirmed a small right kidney with triplex collecting system, with ectopic opening of the right upper moiety ureter at the neck of the urinary bladder. The two lower moiety ureters on the right showed fusion of their draining ureters at the L4 vertebral level, draining into the right vesico-ureteric junction.

MR Brain and spine screening revealed peg-like tonsillar herniation with formation of a syrinx in the cervical cord was seen indicative of Chiari Type I malformation.

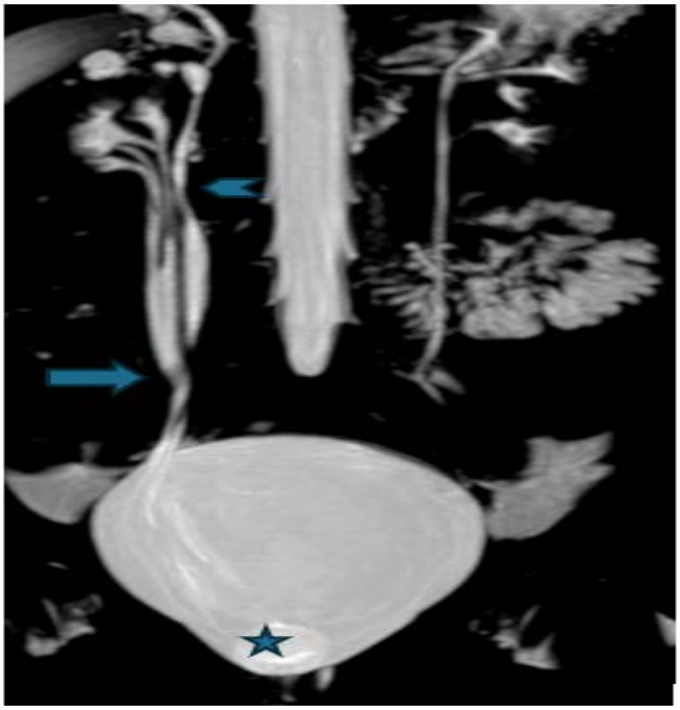

Figure 1: MR Urography coronal MIP Image shows right kidney with triplex collecting system, The lower two moieties on the right show fusion of the ureters at the L4 vertebral level (➡) draining normally into the right vesico-ureteric junction, terminating into a ureterocele (★). The upper moiety ureter is marked by ➤

Discussion

The understanding of ureteral triplication, which is a rare congenital anomaly, has evolved significantly since it was first described by Wrany in 1870. Despite the advancements in medical diagnostic techniques and imaging, the embryological basis and pathogenesis of ureteral triplication remain ambigious[1,4]. This article provides a thorough exploration of the significance of imaging in diagnosing ureteral triplication, the complexities involved in performing a tailor-made MR urography and the possible differential diagnoses & associated anomalies.

Classification

Smith's classification system categorizes ureteral triplication into four types based on the number of ureters and ureteral orifices[3]. Our case falls under type 2, with three ureters and two ureteral orifices (incomplete triplication).

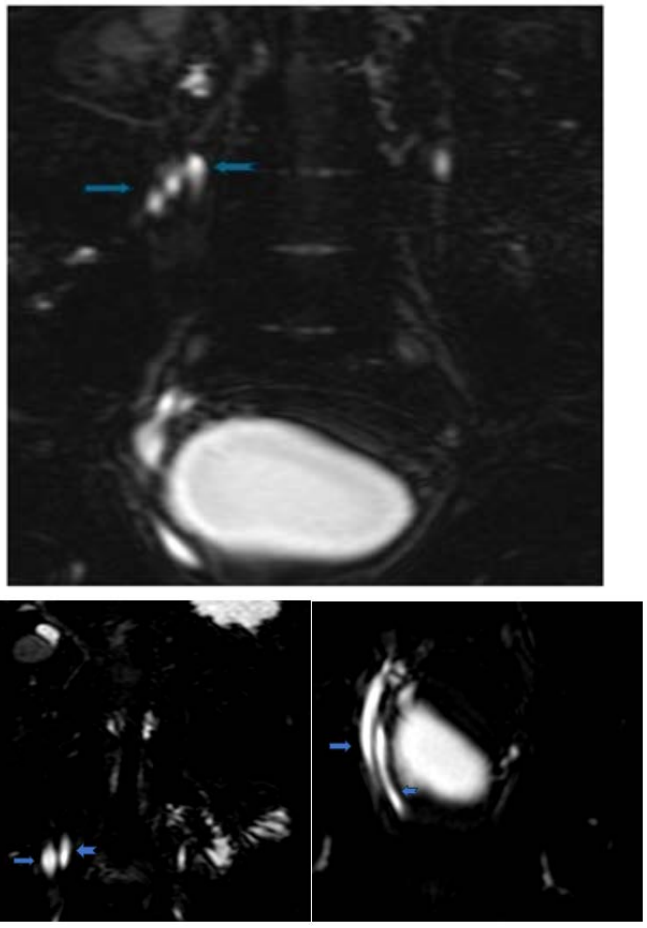

Figure 2 (a,b,c): MR Urography image shows the two laterally placed right lower ureters, fuse at the level of L4 vertebral body (➡) and terminate into the vesico-ureteric junction; whereas the single upper moiety ureter terminates ectopically into the neck of the bladder (➡)

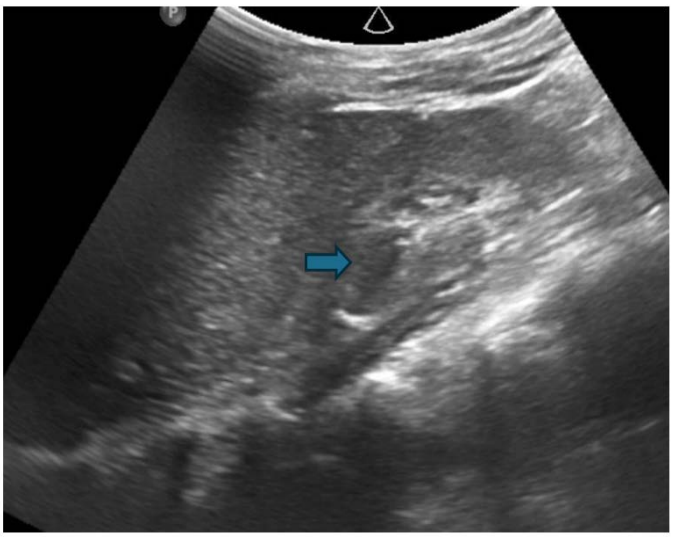

Figure 3: Ultrasound image of the right kidney showing a triplex collecting system (➡) with focal upper moiety caliectasis. An increased parenchymal echogenicity and loss of cortico-medullary differentiation.

Importance of imaging in diagnosis

A precise diagnosis of urinary tract anomalies requires a multimodality imaging approach, with each technique providing distinct structural and functional insights.

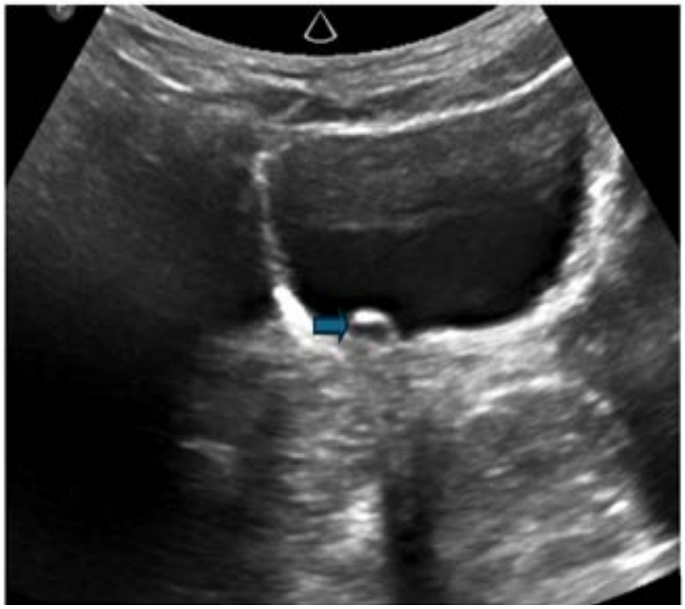

Figure 4: Ultrasound image of the urinary bladder reveals a prominent right ureter terminating in a ureterocele (➡)

Ultrasound is the first-line imaging tool due to its easy accessibility and non-invasive nature, identifying obvious anatomical abnormalities such as duplex moieties, dilated ureters with duplication and ureteroceles. Intravenous urography (IVU), has been particularly useful in detecting anomalies like Smith's type two ureteral triplication. Micturating cystourethrogram (MCU) plays a key role in evaluating the lower urinary tract and diagnosing vesico-ureteric reflux (VUR), a frequent complication of ureteral anomalies. Renal scintigraphy provides functional assessment of the kidneys, helping to identify impairments in the collecting system and overall renal function[1].

MRU emerges as the gold standard for detailed anatomical and functional evaluation of the urinary system, offering superior and unparalleled visualization of complex urinary tract anomalies. MRI, due to its non-ionising nature, has proven valuable in detecting concurrent embryological and developmental conditions, particularly in the pediatric age group; highlighting its indispensable role in a comprehensive diagnostic workup[1,4]. However, it does require patient cooperation and sometimes sedation in pediatric patients to minimize movement during the scan[2,6].

Discussion

Understanding the clinical significance of ureteral triplication is vital for proper management. While incontinence, recurrent urinary tract infections (UTIs) and abdominal pain are prominent presenting signs, the illness often manifests asymptomatically. In some cases, patients may develop complications such as obstruction, reflux and renal dysfunction, which necessitate prompt and appropriate intervention[1,6].

Management strategies for ureteral triplication depend on the severity of symptoms, the presence of complications and the overall functional status of the kidneys. Observation may be sufficient for asymptomatic patients, while those with recurrent UTIs require appropriate antibiotic management to prevent complications. Minimally invasive laparoscopic techniques are indicated for patients with ongoing obstruction or deteriorating renal function[1,2,4].

This 6-year-old girl with right renal triplication and a ureterocele is a complex and rare congenital anomaly which requires expeditious diagnosis. The integration of various imaging modalities, as demonstrated in this report, aligns with the findings of previous studies, highlighting the evolution of diagnostic techniques over time.

Ureteral duplication is the primary differential for triplication, as both anomalies involve multiple ureters draining a single kidney and share complications like reflux, obstruction and ureteroceles. However, distinguishing between them is critical for management. Ultrasound and IVU have limitations, often failing to detect an additional ureter due to poor resolution or contrast filling issues. MRU is the gold standard, offering high-resolution, multiplanar visualization, superior tissue contrast and functional assessment. MRU allows accurate differentiation between duplication and triplication, facilitating optimal treatment planning[1,6].

Another important differential diagnosis is renal dysplasia, a condition characterized by abnormal kidney development, leading to cyst formation and impaired renal function. Where modalities like USG and IVU fail, MRU plays a crucial role in differentiating renal dysplasia from triplication by identifying structural abnormalities and assessing the presence of multiple collecting systems[1,2].

Ureteral triplication often occurs alongside other congenital anomalies, further complicating their diagnosis and management. Contralateral duplication is the most frequently associated anomaly, reported in 37% of cases[2]. Additionally, ureteral ectopia is observed in 28% of cases, while renal dysplasia is noted in 8%[5]. Another significant concern is vesicoureteral reflux (VUR), which can arise as a consequence of obstruction, particularly in the presence of a ureterocele[2]. Recognizing these coexisting anomalies is essential for comprehensive evaluation, as they influence both clinical presentation and treatment strategies.

Conclusion

The discussion emphasizes how crucial it is to use a multidisciplinary approach when diagnosing and treating ureteral triplication, involving radiologists, urologists and pediatric surgeons. Using sophisticated imaging methods such as MRU, can provide comprehensive anatomical and functional data that is critical for informing therapy choices, thereby allowing medical practitioners to proficiently handle this uncommon condition and improving the well-being of those impacted.

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