Co-existing obstructed and refluxing lower moiety megaureter in a female without incontinence: a perplexing myriad in a duplex system with ectopic lower moiety ureter

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Abstract

Ectopic ureter is a rare congenital anomaly that predominantly affects females and typically manifests as urinary incontinence due to atypical ureteral openings in structures such as the vagina or cervix. This condition is estimated to occur in approximately 1 in 4000-10,000 female births. An exceptional variant, the intrasphincteric ectopic ureter, poses significant diagnostic challenges due to its atypical presentation, as it may cause dynamic obstruction and reflux phenomena without resulting in incontinence.

A 28-year-old female presented with severe left flank pain, fever, and chills persisting for 10 days. Her medical history included intermittent dull, aching left flank pain over 7 years, exacerbated during voiding, but no urinary incontinence. Physical examination revealed marked tenderness in the left flank, and laboratory investigations showed elevated leukocyte count and normal serum creatinine levels. Computed tomography urography revealed a left duplex moiety with upper moiety infected hydronephrosis, necessitating percutaneous nephrostomy placement. Subsequent nephrostogram and micturating cystourethrogram identified a grossly dilated and tortuous upper moiety ureter with intrasphincteric ectopic ureteral opening, confirming the diagnosis of obstructed and refluxing megaureter. Following infection resolution, the patient underwent laparoscopic extravesical non-refluxing reimplantation of the upper moiety ectopic ureter with DJ stenting. The postoperative course was uneventful, and the patient reported no flank pain or fever upon stent removal. Periodic evaluations showed no increase in hydronephrosis, indicating a successful intervention. This case underscores the diagnostic complexity of intrasphincteric ectopic ureter, a rare anomaly presenting with obstructed and refluxing megaureter without incontinence. Heightened clinical vigilance and comprehensive radiological evaluation are imperative for timely diagnosis and intervention. Preservation of continence further complicates the clinical picture, necessitating early intervention to prevent renal function deterioration and improve long-term outcomes.

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Introduction

Ectopic ureter, a rare congenital anomaly, predominantly affects females and often results in urinary incontinence due to its atypical ureteral opening in structures such as the vagina or cervix. This condition is estimated to occur in approximately 1 in 4000 to 10,000 female births.[1] The clinical presentation of ectopic ureter is typically characterized by continuous dribbling incontinence. However, an exceptional and intricate variant exists where the ectopic ureter opens in the urethra at the level of the sphincter, known as intrasphincteric ectopic ureter, resulting in patients who are continent. This uncommon presentation introduces significant diagnostic challenges due to the dynamic obstruction and reflux phenomena it incites.[2]

During the bladder filling phase, the sphincter remains contracted, causing functional obstruction at the ectopic ureteral orifice. Conversely, during the voiding phase, the relaxation of the sphincter opens the ureteric orifice, yet the high detrusor pressure generated leads to urine reflux into the ectopic ureter. This cyclical

interplay of functional obstruction and reflux cumulatively impairs renal function over time. Patients often present with flank pain on the affected side, exacerbated during voiding. The subtlety of these symptoms frequently results in delayed presentation, by which time the affected kidney may have already suffered significant functional deterioration or complications such as infected hydronephrosis or pyonephrosis.The insidious nature of intrasphincteric ectopic ureter necessitates a heightened index of suspicion, particularly in young females who report voiding-aggravated flank pain without incontinence. Such clinical vigilance is imperative for timely diagnosis and intervention.[2]

This case report delineates the diagnostic odyssey and therapeutic management of a 28-year-old continent female presenting with flank pain and fever. She was subsequently diagnosed with an intrasphincteric urethral ectopic ureter resulting in an obstructed and refluxing megaureter, compounded by a duplex moiety with the lower moiety ureter being ectopic. The complexity of this case underscores the diagnostic conundrums and therapeutic challenges posed by intrasphincteric ectopic ureter, emphasizing the necessity for prompt, judicious clinical and radiological evaluation to mitigate progressive renal impairment and avert potential complications.

Case Report

A 28-year-old female presented with acute onset of severe left flank pain accompanied by fever and chills persisting for 10 days. Her medical history revealed intermittent episodes of dull, aching pain in the left flank over the past 7 years, for which she had not sought medical attention. She reported being continent, with her flank pain exacerbating during voiding. Physical examination disclosed marked tenderness in the left flank. Laboratory investigations revealed a total leukocyte count of 17,000/mm^3 and a serum creatinine level of 1.0 mg/dL.

A computed tomography urography was performed, revealing a left duplex moiety with upper moiety infected hydronephrosis. Consequently, she underwent percutaneous nephrostomy placement in the upper moiety, which drained frank pus, and she was commenced on broad-spectrum antibiotics. Following the resolution of the infectious episode, a left nephrostogram was conducted, revealing a grossly dilated and tortuous upper moiety ureter. The contrast study delineated the ureter up to the distal segment, with a complete cut-off below the level of the pubic symphysis (Figure 1). Subsequently, a micturating cystourethrogram (MCU) was performed. During the insertion of a 12F urethral catheter, it was inadvertently advanced into the left ectopic ureter instead of the bladder. The filling phase of the MCU showed no reflux (Figure 2), whereas the voiding phase demonstrated reflux originating from the middle third of the urethra, corresponding to the region of the urethral sphincter (Figure 3), into the ectopic ureter. Consequently, a diagnosis of an ectopic ureter resulting in secondary obstructed refluxing megaureter was established. This case is particularly notable because, in the majority of female patients with ectopic ureters, incontinence is prevalent; however, this patient remained continent. Additionally, the concurrent presentation of obstructive and refluxing megaureter within the same ectopic ureter is exceedingly rare in the literature.

Treatment

Following the resolution of the infectious episode, the patient underwent a laparoscopic extravesical non-refluxing reimplantation of the upper moiety ectopic ureter with DJ stenting. Her post-

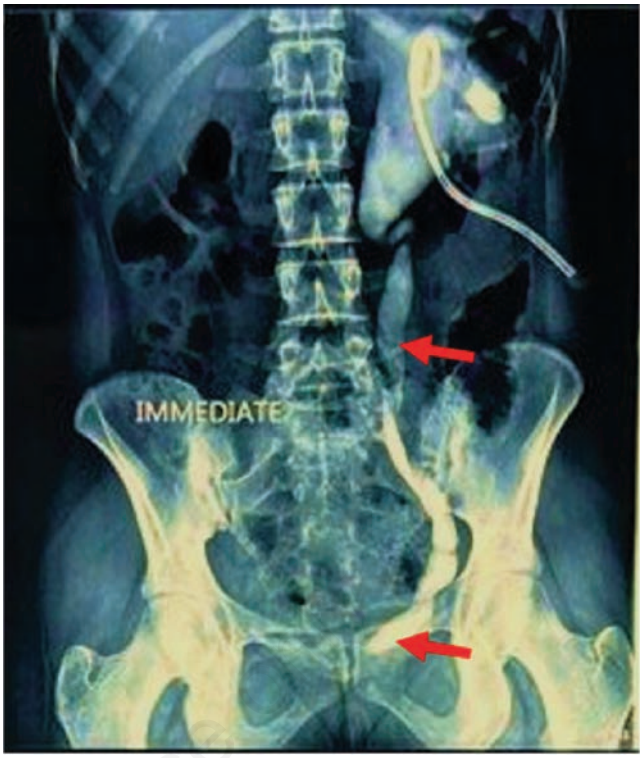

Figure 1. Left nephrostogram: red arrows showing left upper moiety ectopic ureter course with complete cutoff below pubic symphysis.

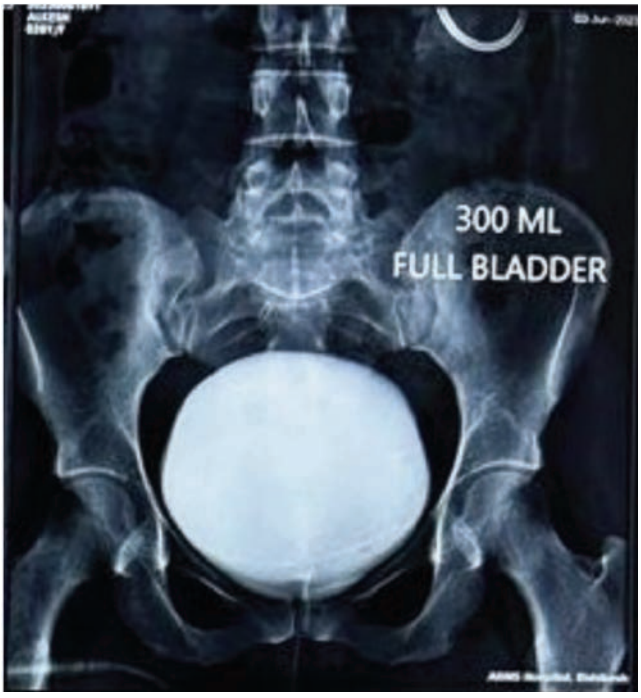

Figure 2. Micturating cystourethrogram showing no reflux in the filling phase.

operative course was uneventful, and she was discharged in stable condition. The DJ stent was subsequently removed after four weeks. Presently, the patient is voiding without experiencing any flank pain or fever. Periodic evaluations of the upper urinary tract have shown no increase in hydronephrosis on the operated duplex moiety side.

Discussion

Ectopic ureter, with an incidence of less than 1% (1 per 1900 population), predominantly affects females more than males.[3] Typically, females with ectopic ureters exhibit urinary incontinence in approximately two-thirds of cases.[1] However, a rare subset of these patients may remain continent, presenting solely with flank pain, which complicates the diagnostic process.[2] In the presented case, the ectopic ureter's intrasphincteric opening, situated below the bladder neck at the urethral sphincter level, preserved the patient's continence. This anatomical anomaly induced repeated brief reflux episodes occurring exclusively during the voiding phases of micturition. Over time, these intermittent refluxes accumulate, leading to a megaureter that is both obstructed and refluxing, as evidenced by the deterioration of the upper moiety's function in our patient. This unique presentation has not been previously documented in the literature. Early identification of such cases in childhood could significantly mitigate the risk of renal function deterioration by adulthood.[4] The insidious nature of the symptoms, coupled with the preservation of continence, necessitates a high index of suspicion for timely diagnosis. Initial manifestations are often subtle, and patients remain continent, further obscuring the clinical picture.[2] In this case, the patient's duplex system with an intrasphincteric ectopic ureter exemplifies the complexity and rarity of such presentations. The dual pathology of obstructive and refluxing megaureter within the same ectopic ureter underscores the necessity for comprehensive clinical and radiological evaluation. Prompt diagnosis and intervention are crucial to prevent progressive renal impairment and complications such as infected hydroureteronephrosis. Clinicians should consider ectopic ureter as a differential diagnosis in young females with persistent flank pain, even in the absence of incontinence, to facilitate early intervention and preserve renal function.[4]

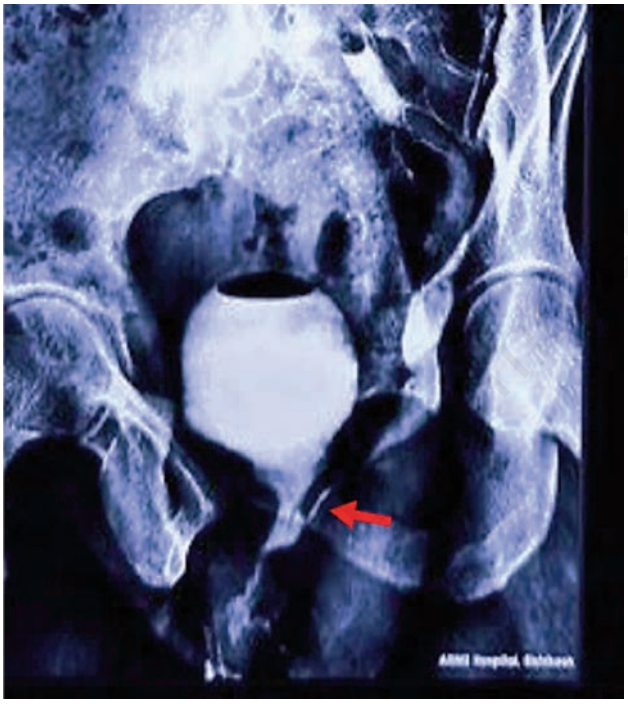

Figure 3. Voiding phase of micturating cystourethrogram: showing (red arrow) reflux into ectopic ureter seen in the region of urethral sphincter.

Conclusions

This case highlights the intricate and uncommon presentation of an intrasphincteric ectopic ureter resulting in a dual pathology of obstructed and refluxing megaureter without incontinence. Such rare anomalies necessitate heightened clinical vigilance, especially in young females with prolonged flank pain. Timely and precise diagnosis, facilitated by comprehensive radiological evaluation, is imperative to avert progressive renal impairment and potential complications. The preservation of continence in these patients further underscores the complexity, mandating early intervention to prevent deterioration of renal function and improve long-term outcomes.

References

1. Wang Q, Wu Z, Zhang F, et al. Gynecological diagnosis and treatment of ectopic ureter insertion into vagina: analysis of five cases and a literature review. J Clin Med 2022;11:6267.
2. el Ghoneimi A, Miranda J, Truong T, Monfort G. Ectopic ureter with complete ureteric duplication: conservative surgical management. J Pediatr Surg 1996;31:467-72.
3. Glassberg KI, Braren V, Duckett JW, et al. Suggested terminology for duplex systems, ectopic ureters and ureteroceles. J Urol 1984;132:1153-4.
4. Singh S, Bhusal NP, Mishra S, et al. Bilateral complete duplication of ureter with ectopic ureter presenting as persistent urinary dribbling with normal voiding pattern in 17-year-old female: case report. Ann Med Surg 2022;84:104824.