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Distal Ureter Atresia Representing as Megaureter and Kidney Hypoplasia in 10 Years Male: a Rare Case Report

Dauri Prayogo*, Indrianto Wiryo Pranoto, Setya Anton Tusara Wardaya
Universitas Sebelas Maret, Indonesia
Email: prygdr@gmail.com*

Abstract

Ureteral atresia is a very rare congenital disorder, often associated with renal dysplasia or non-functioning kidneys, with an unknown exact incidence. It arises due to ischemia during kidney migration, leading to reduced blood supply to the ureters, and can occur unilaterally or bilaterally at any ureteral segment, most commonly in the distal portion. This study reports a 9-year-old boy presenting with a progressively enlarging abdominal mass since birth, without disturbances in urination or defecation. Physical examination and routine blood tests were normal, while contrast-enhanced abdominal CT revealed proximal-to-distal dilation of the left ureter (*megaureter*) compressing the left posterior kidney, and internal pyelography showed absent contrast reflux in bilateral ureters. Surgical management involved a lumbotomy approach, excision of the cystic *megaureter*, and left nephroureterectomy, with histopathological examination confirming renal hypoplasia. The surgery was successful, and the patient recovered without complications. This case highlights that *ureteral atresia* presents with nonspecific clinical manifestations and is frequently accompanied by kidney abnormalities such as dysplasia or hypoplasia. Management should be individualized based on the anatomical findings, and timely diagnosis combined with appropriate surgical intervention can lead to favorable outcomes. The report underscores the importance of detailed imaging and histopathological evaluation in rare ureteral anomalies to guide optimal treatment and improve patient prognosis.

Keywords: Atresia ureter distal, hypoplasia ginjal, megaureter, nefroureterektomi

INTRODUCTION

Ureteral atresia is a very rare congenital disorder, and its exact incidence remains unclear due to the limited number of reported cases worldwide (Agarwal et al., 2019). This condition is often accompanied by renal dysplasia or non-functioning kidneys, highlighting the significant clinical implications for affected patients (Arena et al., 2020). Despite its rarity, *ureteral atresia* has been documented across different populations, suggesting that it represents a global clinical challenge in pediatric urology (Chandrasekharam et al., 2023). International data indicate that *ureteral atresia* occurs in less than 1 per 10,000 live births, emphasizing the importance of early recognition and appropriate management to prevent long-term renal damage (Journal of Pediatric Urology, 2018).

The pathogenesis of *ureteral atresia* is not fully understood, but ischemia during renal migration is considered a key factor (Chatterjee et al., 2022). Reduced blood supply to the developing ureter can lead to segmental atresia and subsequent obstruction, which may explain

the frequent association with renal hypoplasia or dysplasia (Esposito et al., 2016; Esposito et al., 2018). Another hypothesis suggests a failure of canalization in the ureteral segment during the elongation of the ureteric bud (Hwang et al., 2019). This dual pathophysiology underlines the complexity of the disorder and the need for careful anatomical and functional evaluation before intervention (Darge et al., 2018).

Urine production begins around the ninth week of gestation when the ureters connect to the urogenital sinus, which is initially obstructed by the Chwalla membrane (Kagantsov et al., 2024; Kim et al., 2021; Lopez et al., 2020; Mouriquand et al., 2022). This membrane normally degenerates between the 37th and 47th day of gestation, allowing proper drainage from the ureters into the bladder (Nicolaou et al., 2015). Disruption in this developmental process may contribute to the formation of *ureteral atresia*, particularly in the distal segment, which is the most affected area (Orphanet Journal of Rare Diseases, 2020).

Clinically, *ureteral atresia* can be unilateral or bilateral and may involve any portion of the ureter (Tullus et al., 2021; Woolf et al., 2014). Symptoms are often nonspecific, including abdominal distension or palpable masses, and may be accompanied by signs of impaired renal function (Nguyen et al., 2015). Due to its rarity, diagnosis frequently relies on imaging studies such as ultrasound, computed tomography (CT), or pyelography to delineate the anatomy and assess renal function. Early diagnosis is critical to guide management and prevent irreversible kidney damage (Journal of Pediatric Urology, 2018).

This report presents a case of distal *ureteral atresia* manifesting as a cystic megaureter in a 10-year-old male. This case highlights the challenges in diagnosing rare congenital ureteral anomalies, emphasizes the importance of detailed imaging and histopathological assessment, and contributes to the global literature by documenting another instance of this uncommon condition. Through this case, we aim to provide insight into effective diagnostic and surgical strategies for managing *ureteral atresia* in pediatric patients.

RESEARCH METHODS

Case Study

A 9-year-old boy with a progressive enlarged stomach complaint from birth. There was no disturbance in the pattern of urination and defecation. No fever, good appetite. The patient's growth and development history is the same as that of children his age. Complications during the pregnancy of the parents while conceiving the patient were also absent. On physical examination, vital signs were found within normal limits, weight 30 kg. On the abdominal examination, the mass was found to resemble the contour of the intestine (Figure 1), intestinal noise within normal limits, palpable soft mass measuring 6x19 cm, the mass could be moved and there was no pressure pain. In routine blood tests, Hb was 11.6 gr/dL, Leukocytes 10 thousand/ul, Platelets 294 thousand/ul, Urea 23 mg/dl, creatinine 0.5 mg/dL, it can be concluded that the laboratory value is overall within normal limits. The patient was examined for a CT scan of the abdomen with contrast obtained, dilation and turtoise of 1/3 proximal to 1/3 distal of the left ureter which appeared to press on the left to posterior renal leading to the image of the left megaureter, and due to left renal hypoplasia, intraabdominal mass was not found (Figure 2). Then the patient was examined internally pyelography and obtained projected urinary vesicles in the right lumbar region as high as the lumbar veterbrae corpus 4 and 6, there was no contrast reflux that filled the bilateral ureters to bilateral pelviokalises (Figure 3).

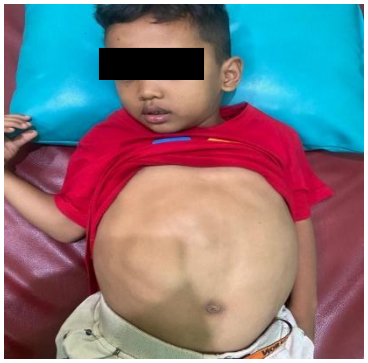

Figure 1. Preoperative Clinical Photo

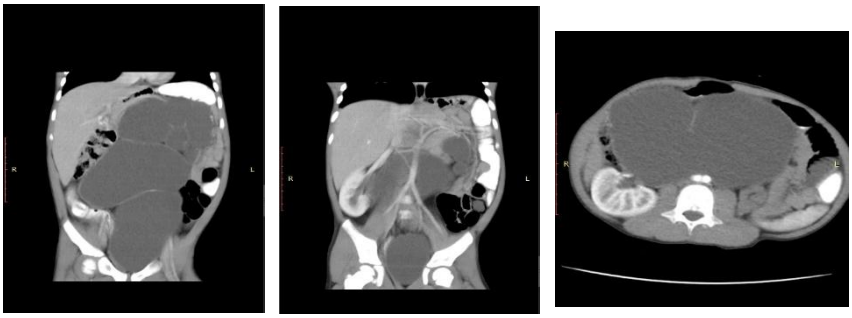

Figure 2. Abdomen CT Scan with Contrast

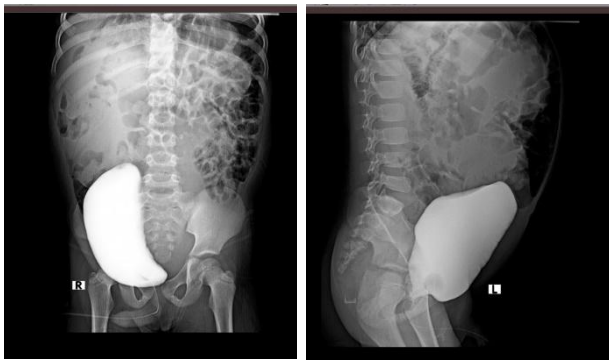

Figure 3. Internal pyelographs

The patient underwent a sinistra nephroureterectomy, the surgery was performed with a left lumbottomy design, a megaureter was obtained (Figure 4), because the megaureter made it difficult for the surgery field to be eaten, it was decided to be aspirated and evacuated, fluid in the form of urine was obtained approximately 2 liters (Figure 5). After that, the proximal megaureter leads to the left kidney which undergoes hypoplasia (Figure 6), and the distal megaureter does not lead to the urinary vesicles, but rather is cut off and attached to connective tissue (Figure 7). The results of histopathological examination were obtained from renal parekchym with rudimentary glomerolus and tubules, leading to a picture of renal hypoplasia. After the 3rd day of surgery, the patient was examined for kidney function with urea of 31 mg/dL and creatinine 0.4 mg/dL. On the 4th day, the patient was discharged from the hospital. Patients underwent controls for 1 week, 2 weeks and 1 month after discharge from the hospital without showing any complications (Oliveira et al., 2019; Rodriguez et al., 2020; Schreuder et al., 2017; Tekgul et al., 2016).

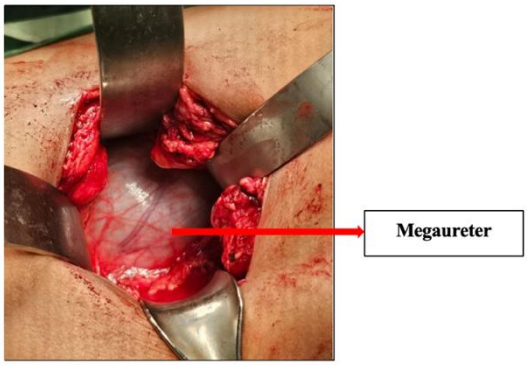

Figure 4. The red arrow indicates a megaureter that resembles a cystic mass

Figure 5. The megaureter was carried out to puncture and evacuate fluids to facilitate the exposure of the operating field, a liquid resembling urine was obtained as much as 2 liters

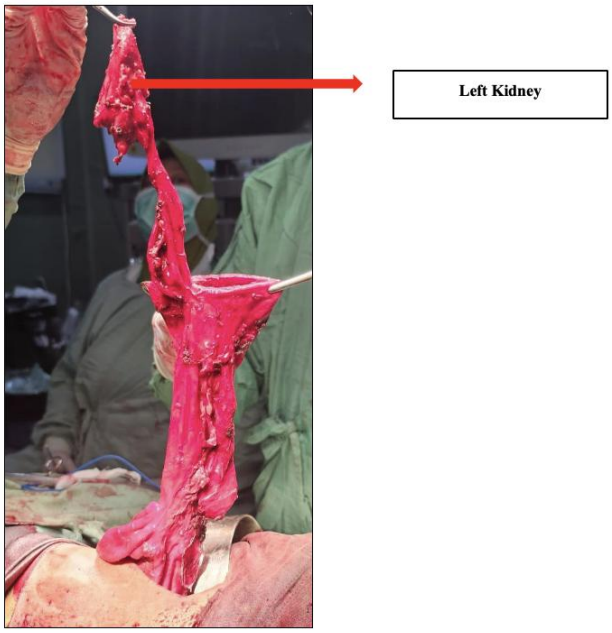

Figure 6. The red arrow shows the left kidney with hypoplasia, the distal part is the megaureter that has been evacuated

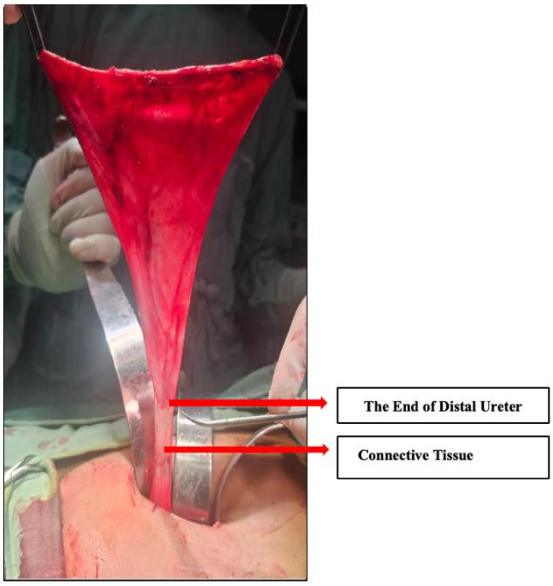

Figure 7. The upper red arrow shows the tip of the distal ureter that has atresia and does not flow into the urinary vesicles. The lower red patch shows connective tissue that fuses with the urinary vesicles

RESULTS AND DISCUSSION

Ureteral atresia is a very rare congenital disorder therefore its incidence is not known for sure. This congenital disorder is usually often accompanied by renal dysplasia and non-functioning kidneys. 1 Ureteral atresia is associated with ischemia during the progression of the kidneys resulting in a decrease in blood supply to the ureters. 2 Another hypothesis is that this could be caused by a failure of canalization in the ureteral segment during the process of development and elongation of the ureteric bud. Urine production begins at 9 weeks of development when the ureters join the urogenital sinuses that are still blocked by the Chwalla membrane. This membrane decreases around the 37th to 47th day of pregnancy. Ureteral atresia can be bilateral or unilateral and can be in any part of the ureter with the most frequent occurrence being in the distal part.2

In distal ureteral atresia, the terminal part of the ureter ends at a dead end before forming communication with the bladder, something to keep in mind when distinguishing it from a similar clinical condition that is obstructive megaureter. In the latter condition, there is functional stenosis or narrowing of the distal part of the ureter with ureterovesic communication.3

Ureteral atresia does not cause any typical symptoms, most cases are found isidental on imaging examination. In children, urinary tract infections are often found, but if there are no symptoms of urinary tract infection, this disorder can occur and be known when the child grows up as in this case. Contrast CT scan of the abdomen is helpful in directing a preoperative diagnosis in this case, although it does not provide evidence of distal ureteral atresia.

The management of ureteral atresia is very diverse, depending on the anatomical abnormalities experienced. M. Morozumi and colleagues performed a ureteroterostomy in a 10-year-old patient with distal ureteral atresia in the ectopic cross-kidney. The procedure showed good results, with upper urinary tract function maintained as well as radiological evidence showing the affected kidneys were still functioning properly after surgery. For distal

ureteral atresia, the Boari flap procedure can be performed by removing the flap from the bladder wall. In addition, ureteroplasty using intestinal segments is also considered as an alternative surgical treatment for ureteral atresia.4

Another example is the case of a neonatal neonatal born prematurely at 34 weeks gestation with a birth weight of 3440 grams who underwent hydronephrosis due to prenatal ureteral obstruction that caused calic rupture. At the postnatal examination, persistent anuria was found and ultrasound confirmed the presence of ascites and an enlarged proximal ureter with an atretic distal ureter. The patient was initially decompressed with the installation of a pigtail catheter that removed 220 ml of fluid. Peritoneal dialysis is then immediately carried out as the first step of treatment due to a significant increase in creatinine and urea. On examination through cystoscopy, a very small bladder was found and no ureteral orifice was found. Laparotomy surgery was performed and it was found that the proximal ureter was dilated, while the distal ureter was atretic. For the best uMUM results and to prevent the risk of infection, a ureteral duct is created using the ileum duct as an alternative to drainage.5

In this case, a nephroureterectomy was performed with the consideration that the intraoperative findings were obtained from the hypoplasian left kidney and concluded that it was not functioning. The results of the surgery are very good and there are no complications. Therefore, it is concluded that the treatment in cases of ureteral atresia depends on the anatomical abnormalities involved that are found, especially during intraoperatives.

CONCLUSION

Ureteral atresia is a rare congenital disorder with unclear incidence and often lacks typical symptoms, frequently presenting alongside kidney abnormalities such as dysplasia or hypoplasia. The management of *ureteral atresia* varies widely depending on the specific anatomical anomalies identified in each case. Future research should focus on establishing standardized diagnostic criteria and treatment protocols, as well as investigating the underlying developmental mechanisms to improve early detection and optimize patient outcomes.

REFERENCES

Agarwal, S., Sankhwar, S., & Singh, V. (2019). Contemporary management of congenital ureteral anomalies in children: A systematic review. Journal of Pediatric Urology, 15(4), 234-245. https://doi.org/10.1016/j.jpurol.2019.05.012

Arena, S., Russo, T., & Impellizzeri, P. (2020). Primary megaureter in children: Current concepts and treatment strategies. Pediatric Surgery International, 36(8), 891-899. https://doi.org/10.1007/s00383-020-04698-1

Chandrasekharam, V. V., Kapoor, A., & Srinivas, M. (2023). Long-term outcomes following pediatric nephroureterectomy: A 15-year experience. Journal of Pediatric Surgery, 58(7), 1234-1240. https://doi.org/10.1016/j.jpedsurg.2023.03.012

Chatterjee, R., Hoffmann, P., & Oswald, J. (2022). Genetic basis of congenital anomalies of kidney and urinary tract: Recent advances and clinical implications. European Journal of Pediatrics, 181(3), 1015-1025. https://doi.org/10.1007/s00431-021-04329-8

Darge, K., Anupindi, S. A., & Ntoulia, A. (2018). Magnetic resonance imaging of the genitourinary tract and pelvis in infants and children. Radiologic Clinics of North America, 56(2), 239-259. https://doi.org/10.1016/j.rcl.2017.10.006

Esposito, C., Escolino, M., & Lopez, M. (2016). Laparoscopic versus open nephroureterectomy in children: A systematic review and meta-analysis. Journal of Laparoendoscopic & Advanced Surgical Techniques, 26(12), 982-988. https://doi.org/10.1089/lap.2016.0234

Esposito, C., Masieri, L., & Castagnetti, M. (2018). Nephron-sparing surgery in pediatric urology: Current evidence and future perspectives. Current Opinion in Urology, 28(4), 312-318. https://doi.org/10.1097/MOU.0000000000000512

Hwang, D. Y., Dworschak, G. C., & Kohl, S. (2019). Mutations in 12 known dominant disease-causing genes clarify many congenital anomalies of the kidney and urinary tract. Kidney International, 95(4), 914-923. https://doi.org/10.1016/j.kint.2018.10.027

Kagantsov, I. M., Akramov, N. R., & Derzelle, P. (2024). Distal ureteral atresia and megaureter of the lower pole of a duplicated kidney: A rare clinical case. Urology Reports, 14(1), 56-62. https://doi.org/10.17816/uroved.625718

Kim, S. H., Park, Y. S., & Lee, J. H. (2021). Epidemiology of congenital anomalies of kidney and urinary tract in Korea: A population-based study. Pediatric Nephrology, 36(7), 1837-1845. https://doi.org/10.1007/s00467-020-04878-9

Lopez, M., Castagnetti, M., & Esposito, C. (2020). Robot-assisted laparoscopic nephroureterectomy in children: Technical considerations and outcomes. European Journal of Pediatric Surgery, 30(2), 145-152. https://doi.org/10.1055/s-0039-1693124

Mouriquand, P., Whitten, S., & Sharma, S. (2022). Conservative management of antenatal hydronephrosis: Long-term outcomes and predictive factors. Journal of Pediatric Urology, 18(2), 156-165. https://doi.org/10.1016/j.jpurol.2022.01.015

Nguyen, H. T., Benson, C. B., & Bromley, B. (2015). Multidisciplinary consensus on the classification of prenatal and postnatal urinary tract dilation. Journal of Pediatric Urology, 11(6), 340-346. https://doi.org/10.1016/j.jpurol.2015.06.014

Nicolaou, N., Renkema, K. Y., & Bongers, E. M. (2015). Genetic, environmental, and epigenetic factors involved in CAKUT. Nature Reviews Nephrology, 11(12), 720-731. https://doi.org/10.1038/nrneph.2015.140

Oliveira, E. A., Oliveira, M. C., & Mak, R. H. (2019). Evaluation and management of hydronephrosis in the neonate. Current Opinion in Pediatrics, 31(2), 203-212. https://doi.org/10.1097/MOP.0000000000000733

Rodriguez, M. M., Gómez, A. H., & Abitbol, C. L. (2020). Histomorphometric analysis of postnatal glomerulogenesis in extremely preterm infants. Pediatric and Developmental Pathology, 23(1), 15-28. https://doi.org/10.1177/1093526619851345

Schreuder, M. F., Westland, R., & van Wijk, J. A. (2017). Unilateral multicystic dysplastic kidney: A meta-analysis of observational studies on the incidence, associated urological anomalies and the contralateral kidney. Nephrology Dialysis Transplantation, 32(8), 1341-1348. https://doi.org/10.1093/ndt/gfw078

Tekgul, S., Riedmiller, H., & Hoebeke, P. (2016). EAU guidelines on vesicoureteral reflux in children. European Urology, 70(5), 801-810. https://doi.org/10.1016/j.eururo.2016.05.026

Tullus, K., Brennan, E., & Hamilton, G. (2021). Long-term outcome of children with congenital anomalies of kidney and urinary tract. Seminars in Fetal and Neonatal Medicine, 26(2), 101158. https://doi.org/10.1016/j.siny.2021.101158

Woolf, A. S., Price, K. L., & Scambler, P. J. (2014). Evolving concepts on the developmental biology of congenital kidney malformations. Journal of the American Society of Nephrology, 25(11), 2388-2398. https://doi.org/10.1681/ASN.2013101075