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Distal ureteral atresia, megaureter of a lower pole of a duplicated kidney: the rare clinical case

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ABSTRACT

Ureteral atresia is a rare pathology, the diagnosis of which is difficult, and a unified approach to the method of surgical correction has not been formed. The aim of this study is to demonstrate a rare clinical case of a patient with distal ureteral atresia of a non-functioning lower pole of a duplicated kidney, and also to present the method of surgical treatment. A 12-year-old female patient was diagnosed with a cystic formation of the retroperitoneal space. During the operation, it was determined that this cyst was an atretic, dilated ureter of a lower pole of the duplicated kidney. Lower heminephrureterectomy was chosen as the method of surgical correction. Ureteral atresia is rarely combined with any other anomalies of the upper urinary tract, and the presented clinical case has not previously been described in the available foreign and Russian literature. The methods of surgical correction of ureteral atresia include nephrureterectomy, ureteroureterostomy, intestinal ureteroplasty, and Boari procedure. In the presented case, heminephrureterectomy was chosen because of severe hypoplasia of the lower pole of the duplicated kidney and the almost complete absence of its parenchyma. When examining patients with cystic formations of the retroperitoneal space, it is necessary to remember about the ureteral atresia, which may be accompanied by other anomalies of the upper urinary tract.

Keywords: ureteral atresia; duplicated kigney; heminephrureterectomy; cystic formation.

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Дистальная атрезия мочеточника, мегауретер нижнего сегмента удвоенной почки: редкий клинический случай

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АННОТАЦИЯ

Атрезия мочеточника — редкая патология, диагностика которой затруднительна, а единый подход к способу оперативной коррекции не сформирован. В статье продемонстрирован редкий клинический случай пациента с дистальной атрезией мочеточника нефункционирующего нижнего сегмента удвоенной почки, а также представлена примененная тактика хирургического лечения. Пациентка, 12 лет, с выявленным кистозным образованием забрюшинного пространства. В ходе операции было определено, что данное образование являлось атрезированным, резко расширенным мочеточником нижнего сегмента удвоенной почки. В качестве метода хирургической коррекции была выбрана нижняя геминефруретерэктомия. Атрезия мочеточника редко сочетается с какими-либо другими аномалиями верхних мочевых путей, а представленный клинический случай до этого не был описан в доступной зарубежной и отечественной литературе. Тактика хирургической коррекции атрезии мочеточника включает в себя нефруретерэктомию, уретероуретеростомию, кишечную пластику мочеточника, операцию Боари. В представленном случае выбрана геминефруретерэктомия по причине выраженной гипоплазии нижнего сегмента удвоенной почки, а также практически полного отсутствия его паренхимы. При обследовании пациентов с кистозными образованиями забрюшинного пространства необходимо помнить о возможности наличия атрезии мочеточника, которая может сопровождаться другими аномалиями верхних мочевых путей.

Ключевые слова: атрезия мочеточника; удвоение почки; геминефруретерэктомия; кистозное образование.

Как цитировать

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INTRODUCTION

Ureteral atresia is a rare disorder associated with either inadequate blood supply to the ureter or impaired ureteral canalization during embryonic development. Ureteral atresia is most often associated with renal dysplasia or multicystic kidney. The association with any other urinary anomaly is extremely rare. As a result, there is currently no clear treatment regimen for these patients, and the choice of surgical strategies is individual and varied.

The *aim* is to report a rare case of a patient with distal ureteral atresia of the non-functioning lower duplex kidney. Furthermore, the surgical strategies used are presented.

CLINICAL OBSERVATION

A 12-year-old female patient was examined in the gynecology department for a cystic lesion in the abdominal cavity (presumably a paraovarian cyst), which was found incidentally by ultrasound. Interestingly, the patient did not have any clinical symptoms or active urinary tract infections. Diagnostic laparoscopy revealed no ovarian cysts. A mass protruding into the peritoneal cavity was visualized in the retroperitoneal space. The patient underwent further evaluation. Computed tomography urography revealed a poorly defined retroperitoneal fluid collection without contrast enhancement, located near the right ureter (Fig. 1). The right kidney, however, showed no abnormalities. Cystography showed that the contours of the bladder were smooth and clear, and no diverticula, contrast leakage, or vesicoureteral reflux were found.

The patient was admitted to the operating room with a diagnosis of cystic pelvic lesion. Laparoscopic isolation of the mass with the pelvic part up to 10 × 7 × 5 cm showed that it ended blindly at the level of the round ligament of the uterus (Fig. 2–4). As the cystic mass was isolated more proximally, its tight attachment to the right ureter, stenosis, and extension towards the kidney were noted. The right kidney and its hilum were accessed, and the cystic lesion was found to be an atretic, highly dilated ureter of the lower segment of the duplex right kidney (Fig. 5). Its inferior segment was hypoplastic, measuring up to 12 × 10 × 10 mm, with virtually no parenchyma. Therefore, it was decided to perform an inferior heminephroureterectomy. The final view and schematic presentation of the extracted mass are shown in Figure 6 and Figure 7, respectively. The postoperative course was uneventful. The patient had no active urinary tract infections. Laboratory parameters of renal metabolism were normal, and early postoperative ultrasound showed no abnormalities in the remaining right upper kidney.

DISCUSSION

Ureteral atresia, which is a rare disorder, is most often accompanied by multicystic transformation or dysplasia of the kidney. It is rarely combined with other

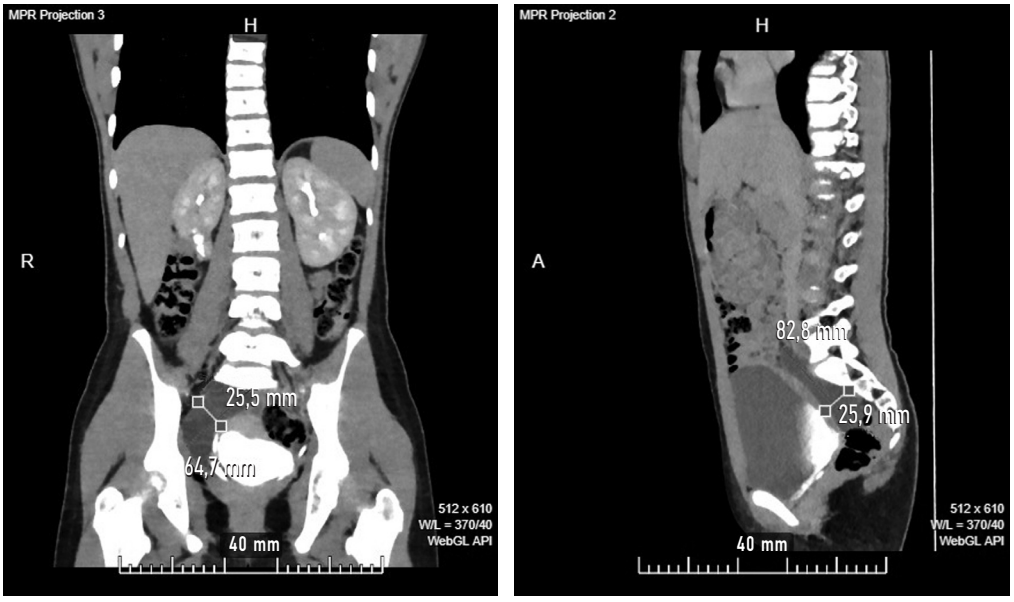

Fig. 1. Computed tomography urography (frontal and sagittal section), delayed phase. A cystic formation is visualized (dimensions are indicated), located along the lower third of the right ureter, which does not accumulate contrast

Рис. 1. Компьютерная томография-урография (фронтальный и сагиттальный срез), отсроченная фаза. Визуализируется кистозное образование (указаны размеры), располагающееся вдоль нижней трети правого мочеточника, ненакапливающее контраст

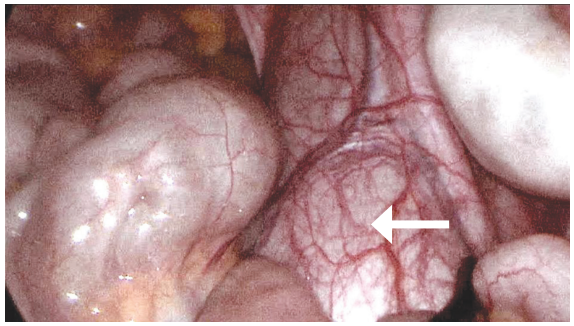

Fig. 2. Intraoperative view of a retroperitoneal formation protruding into the abdominal cavity (indicated by an arrow)
Рис. 2. Интраоперационный вид пролабирующего в брюшную полость забрюшинного образования (указано стрелкой)

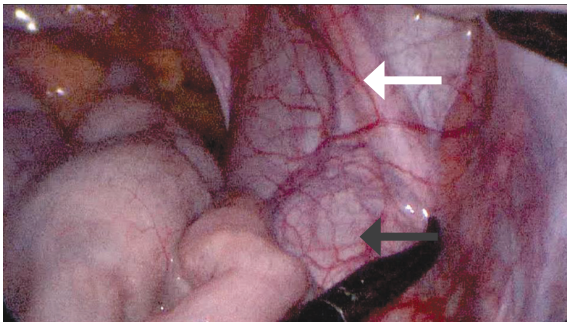

Fig. 3. Type of detected retroperitoneal cystic formation (indicated by a black arrow), along the anterior wall of which the right ureter is visualized (indicated by an arrow with a contour)
Рис. 3. Вид обнаруженного забрюшинного кистозного образования (черная стрелка), по передней стенке которого визуализируется правый мочеточник (белая стрелка)

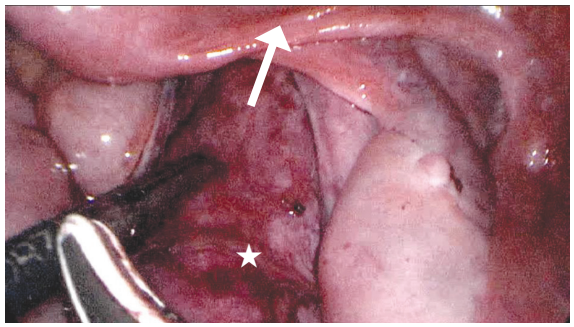

Fig. 4. Intraoperative view of a partially exposed atretic ureter (indicated by an asterisk). The arrow indicates the round ligament of the uterus, at the level of which atresia was visualized
Рис. 4. Интраоперационный вид частично выделенного атрезированного мочеточника (звездочка). Стрелкой указана круглая связка матки, на уровне которой визуализировалась атрезия

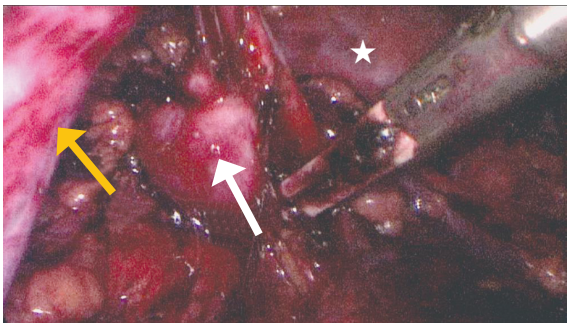

Fig. 5. View of the isolated hypoplastic lower segment of the doubled right kidney (indicated by a white arrow). The upper segment is indicated by an asterisk, the atretic ureter of the lower segment is indicated by a yellow arrow
Рис. 5. Вид выделенного гипоплазированного нижнего сегмента удвоенной правой почки (белая стрелка). Верхний сегмент указан звездочкой, атрезированный мочеточник нижнего сегмента — желтой стрелкой

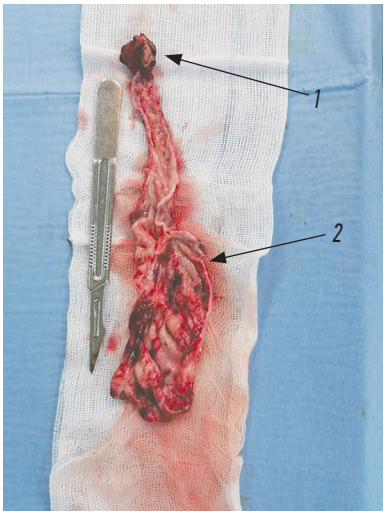

Fig. 6. The final view of the extracted specimen. *1* — sharply hypoplastic lower segment of the double right kidney; *2* — atretic ureter (mucosa)
Рис. 6. Окончательный вид извлеченного препарата.
1 — резко гипоплазированный нижний сегмент удвоенной правой почки; *2* — атрезированный мочеточник (слизистая оболочка)

anomalies of the upper urinary tract, with only individual cases reported. For example, S. Zundel et al. [1] described a clinical case of a newborn girl with aplastic left kidney and distal ureteral atresia in the single right kidney. M. Morozumi et al. [2] reported an even rarer clinical case of distal ureteral atresia in a crossed fused ectopic kidney. This disorder is very difficult to diagnose because it is rare. W. Shuiqing et al. [3] described a clinical case of a 19-month-old girl who first underwent pyeloplasty for hydronephrosis and then underwent antegrade ureteropyelography for acute pyelonephritis after nephrostomy clamping, and distal ureteral atresia was found [3]. There are proximal and distal forms of this condition, with the latter being more common. Clinical symptoms may be absent or pyelonephritis exacerbations may occur. Ultrasound imaging is the first step in the diagnosis to identify a cystic retroperitoneal lesion as an enlarged ureter. Surgical treatments for ureteral atresia include nephroureterectomy for a failed kidney and pyeloureterostomy or ureteroureterostomy for a functioning kidney [4]. For example, M. Morozumi et al. performed ureteroureterostomy in a 10-year-old patient with distal atresia of the ureter of a crossed ectopic kidney [2]. This surgery was considered to have a favorable outcome with adequate postoperative upper urinary tract urodynamics and radiographic evidence of preserved function of the affected kidney. For distal ureteral atresia, the Boari flap procedure can also be performed using a flap taken from the bladder wall [3]. Intestinal ureteroplasty was also considered as an alternative surgical treatment of ureteral atresia [1]. This article presents a rare case of combined distal ureteral atresia and complete duplex kidney. Similar clinical observations were not found in available foreign and Russian literature.

Duplex kidneys may be associated with various obstructive uropathies (obstructive megaureter, ectopic ureteral orifice, ureterocele), but the upper duplex kidney is more prone to such disorders and usually functions worse than the lower segment [5, 6]. This is related to the Meyer–Weigert rule, which describes the orifice of the ureter of the upper duplex kidney as being caudal and medial, and the lower segment as being cranial and lateral. This anatomical relationship is associated with some aspects of the embryonic development of the two ureteric buds and the further rotation of the kidney. There are some rare cases where the Weigert–Meyer rule is not observed. One of these is obstructive megaureter of the lower duplex kidney [7]. However, surgical treatments for obstructive uropathies of the duplex kidney are diverse and include heminephrectomy, ureteroureterostomy, ureteral reimplantation, endoscopic transurethral dissection

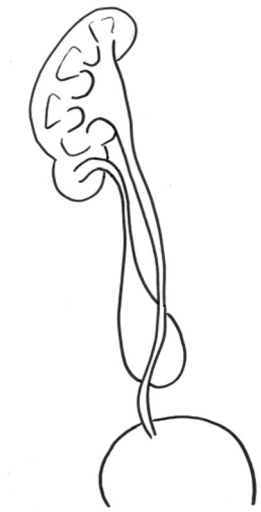

Fig. 7. Schematic representation of the described pathology
Рис. 7. Схематическое изображение описанной патологии

of ureterocele, etc. [6]. In the case presented, the renal duplication was accompanied by distal atresia of the lower ureter, which is characterized by complete loss of its function. Therefore, surgical treatment with heminephrectomy seems to be the most appropriate. Moreover, organ-preserving surgery was unacceptable due to the severe dilatation and structural remodeling of the atretic ureteral wall. In the case presented, the lower segment was involved, which is quite rare. The described observation also shows how difficult it is to establish a correct diagnosis in the preoperative period, including due to the lack of radiographic contrast enhancement in the atretic ureter. Other authors have also noted this factor [1, 3].

CONCLUSION

When examining patients with retroperitoneal cystic lesions, the possibility of ureteral atresia should be considered. Ureteral atresia may be associated with other upper urinary tract anomalies. The interaction between the gynecologist and the urologist is crucial in the management of girls with pelvic cysts.

ADDITIONAL INFORMATION

Authors' contribution. All authors made a substantial contribution to the conception of the study, acquisition, analysis, interpretation of data for the work, drafting and revising the article, final approval of the version to be published and agree to be accountable for all aspects of the study. Personal contribution of each author: I.M. Kagantsov — concept and design of the study, editing the text of the manuscript, performing surgery; E.A. Kondratyeva — concept and design of the study, writing the text of the manuscript, performing surgery, searching and reviewing publications on the research topic; N.A. Kokhreidze — concept and design of the study,

editing the text of the manuscript; S.A. Karavaeva — editing the text of the manuscript; T.M. Pervunina — editing the text of the manuscript.

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Consent for publication. Written consent was obtained from the patient for publication of relevant medical information and all of accompanying images within the manuscript.

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