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Duplex Kidney with a Segmental Solitary Cystic Dysplasia and Ureteric Atresia: A Rare Case

Dupleks Böbrekte Segmenter Soliter Kistik Displazi ve Üreter Atrezisi: Nadir Bir Olgu

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ABSTRACT

Dysplastic kidney cysts are developmental cysts of the kidney. They are mostly multi-cystic, and usually involve the whole kidney. In addition, the segmental and solitary form is rare. A one-year-old boy with an antenatal ultrasound diagnosis of a left kidney cyst was followed-up. Postnatal ultrasound examination revealed a duplex left kidney with a cystic dilatation in the upper pole and a normal appearing parenchyma in the lower pole. The cystic lesion (10×10 cm) with an upper polar location that did not show any regression was excised laparoscopically. The ureter communicating with the cystic component of the kidney was not seen on the left side during laparoscopy. Histopathologic diagnosis was a cystic renal dysplasia. We present a very rare case of a duplex kidney with a solitary dysplastic cyst and ureteric atresia and discuss the diagnostic and therapeutic process.

Keywords: Duplex kidney, cystic dysplasia, ureteral atresia, children

ÖZ

Displastik böbrek kistleri böbreğin gelişimsel kistlerinden olup çoğunlukla multi-kistik ve genellikle tüm böbreği tutan patolojilerdir. Segmenter ve soliter formu nadirdir. Bir yaşında erkek olgu antenatal tanımlanmış sol böbrek kisti nedeni ile doğum sonrası takip edilmiş. İncelemelerinde solda dupleks böbrek, alt kutupta normal böbrek dokusu, üst kutbunda soliter kistik genişleme tespit edildi. Takiplerinde gerileme göstermeyen üst kutup yerleşimli kistik yapı (10x10 cm) laparoskopik olarak eksize edildi, üreteri görülmedi. Histopatolojisi kistik renal displazi olarak tanımlandı. Dupleks böbrek anomalisi ve aynı tarafta displazik kistik böbrek yapısı bulunan, üreteral atrezili olgu, çok nadir görülmesi ve tanı-tedavi süreçlerinin irdelenmesi için sunuldu.

Anahtar Kelimeler: Dupleks böbrek, kistik displazi, üreter atrezisi, çocuk

Introduction

Multi-cystic dysplastic kidney (MCDK) is the most common form of renal dysplasia that is usually described as a cystic kidney enlargement during routine antenatal ultrasonographic examination. Dysplasia rarely occurs in solitarily form. The importance of the differentiation and definitive diagnosis of cystic kidney diseases lies in the fact that surgical treatment is not necessary unless complications, such as malignant transformation or hypertension, develop (1). In this study, we presented the clinical findings, imaging features, diagnosis, and treatment of a segmental solitary cystic dysplastic kidney (SSCDK). Our patient was being followed-up since a cystic kidney enlargement was detected by antenatal ultrasonography.

Case Report

Verbal and written informed consent was obtained from the parents of the patient who participated in this study. Physical examination of the one-year-old boy with a left hypochondriac mass showed a left kidney upper pole cyst on ultrasonography. He had a history of left kidney hydronephrosis on antenatal ultrasonography.

Kidney ultrasonography revealed a smooth-edged upper pole cyst of the left kidney with dimensions of 10×10 cm. Intravenous urography showed a lucent mass displacing the calyces at the left upper pole. Mild calyceal dilatation was seen with a normal draining ureter in the lower pole (Figure 1). Lower pole of the kidney had a normal appearance (Figure 2), and a bifid pelvis was reported. Magnetic resonance urography showed

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a duplex collecting system with a double ureter. There was a normal ureter draining the lower pole of the kidney. There was also a dysgenetic dilated ureter with a blind proximal ending, having a separate bladder opening. The bladder was normal in the voiding cystourethrography, and reflux to the ureter was not seen (Figure 3).

The removal of the lesion by laparoscopic surgery was decided due to the growth of the mass and the compression symptoms. Orifices of the left and right ureters were seen during cystoscopy, which was performed under anaesthesia. There was no left complete double ureteric orifice, which was defined by the magnetic resonance imaging (MRI). The upper pole, which was in solitary cystic structure morphology, was excavated almost totally with the unroofing technique. No separate ureter was communicating with the upper pole. Histopathological diagnosis was a renal dysplasia. The final diagnosis was a left kidney segmental solitary dysplasia of the upper pole with a normal lower pole and duplex collecting system. After the 4th day of the surgery, the left kidney (lower pole), right kidney and bilateral ureters were evaluated and found as normal in the intravenous urography.

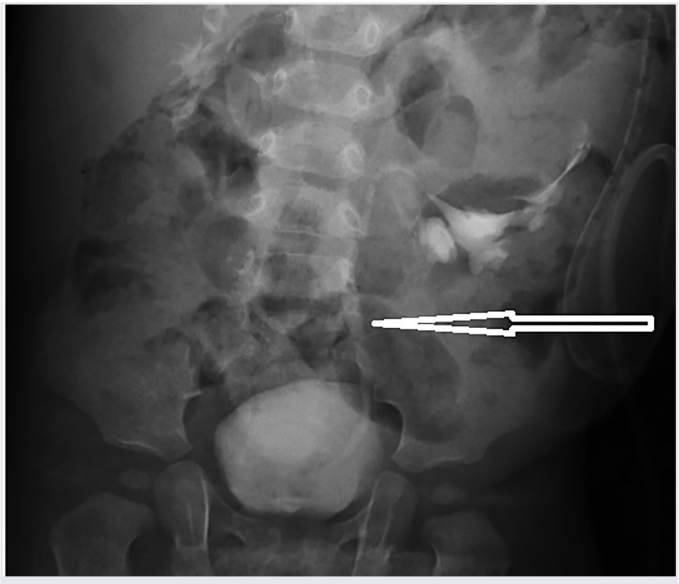

Figure 1. Intravenous urography shows a lucent defect displacing the calyces at the left upper pole. Mild calyceal dilatation is seen with a normal ureter draining the lower pole calyces

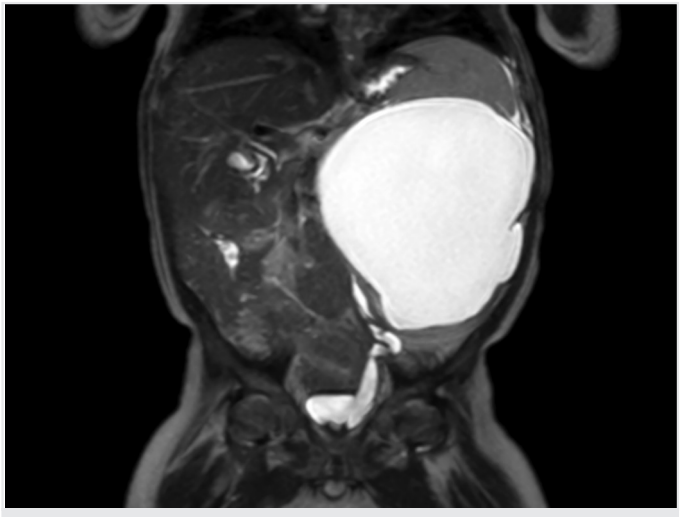

Figure 2. Coronal thick slab T2-weighted magnetic resonance urography shows a left-sided cystic kidney mass in the upper pole. A low lying dilated and tortuous left upper moiety ureter is seen

Discussion

Dysplastic renal development anomalies are most commonly seen in the multi-cystic form. MCDK is the most common cause of abdominal mass in neonates following hydronephrosis. Renal dysplasia, the most common cystic disease in childhood, occurs during the metanephric phase of embryological development. If an obstruction occurs in the period after the embryological development, hydronephrosis instead of dysplasia will occur. The most common one is unilateral renal dysplasia. This form consists of 80%-90% of this anomaly, and occurs as a result of a pelvi-ureteral blockage of the entire kidney. If obstruction occurs at the infundibular level, the upper pole of the kidney can also be seen in a segmental or focal shape (1,2). Based on this information, it can be said that an infundibular blockage occurred at the late embryonic stage; therefore SSCDK was formed in our presented case.

SSCDK is rarer than MCDK, which has an occurrence rate of 1:4300 (3). These cases were first described as duplex kidneys and are difficult to identify because they have atypical clinical presentations. Abdominal mass occurs clinically in childhood, but it is sometimes not noticeable until adulthood. Patients sometimes apply to hospitals with complaints of recurrent urinary tract infections, abdominal pain, haematuria, and developmental delay (4). Since the SSCDK is a developmental anomaly, it may be accompanied by additional anomalies such as ureteropelvic obstruction, ureteral agenesis, dilatation of the collecting system without renal hypoplasia or ureterocoele, vesicoureteral reflux, malrotation, renal agenesis and horseshoe kidney. Less frequently, there are other abnormalities such as ectopic ureters opening in the dysplastic kidney or vesicoureteral reflux (5). The case did not have any findings other than the growing abdominal mass, and it was accompanied by a urethral atresia.

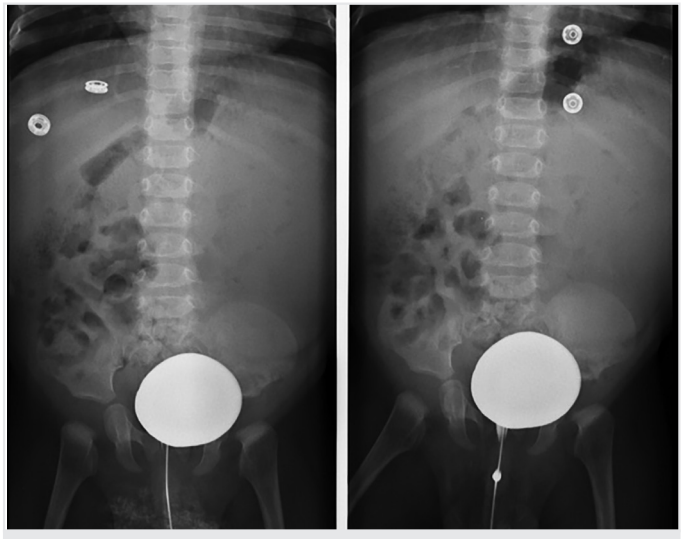

Figure 3. No vesicoureteral reflux is seen on voiding cystourethrogram

SSCDK can give the impression of a mass on abdominal radiography, with a kidney that is increased in size. Cyst wall calcifications are rarely seen in childhood, although they are seen in one third of cases among adults. Cystic structures replacing the normal kidney parenchyma is the ultrasonographic finding (6,7). In computed tomography and MRI, MCDK is a kidney mass composed of a large number of small cystic masses separated by non-associative septal defects with no contrast involvement (8-10). Due to the fact that the presented case is in the solitary form, there are not enough distinctive imaging findings in these cases and therefore, it was identified as hydronephrosis in the duplex kidney. The identification of the complete double-collector system was not confirmed by both cystoscopy and laparoscopy.

Histopathologic examination is used to distinguish various aetiologies among small kidneys, because this distinction is important for disease prognosis and genetic counselling. The diagnosis of renal dysplasia is not difficult; however, the diagnosis is sometimes confused with other conditions including polycystic kidney disease, foetal kidney, renal hypoplasia and renal atrophy. Histopathologically, some structures unique to the foetal stage such as metaplastic cartilage residues, hypoplasia, primitive mesenchymal tissues and canals are seen. Their number and size depends on whether they are hypoplastic or dysplastic. The absence of metaplastic cartilage tissue does not rule out renal dysplasia (4). In the presented case, the presence of primitive tubules and of cystic structures containing thin mesenchymal tissue confirmed the diagnosis of SSCDK.

As a result, SSCDK may occur with different clinical and radiological findings. For this reason, SSCDK should be included in the differential diagnosis of cystic kidney and cystic kidney masses. SSCDK are difficult to diagnose because they do not have pathognomonic imaging features, if there is no collector system duplication. In typical cases, SSCDK occurs at the upper pole. In symptomatic patients with the absence of involution, surgical treatment may be used instead of non-operative treatment, if there is hypertension.

Ethics

Informed Consent: Verbal and written informed consent was obtained from the parents of the patient who participated in this study.

Peer-review: Externally peer-reviewed.

Authorship Contributions: Surgical and Medical Practices- Ü.B., M.S., T.T. ; Concept- Ü.B., T.T., A.K. ; Design- Ü.B., T.T., A.K.P. ; Data Collection or Processing- M.S., A.K.P. ; Analysis or Interpretation- T.T., A.K.P. ; Literature Search- M.S., A.K. ; Writing- Ü.B., T.T., A.K.

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