Renal Hypoplastic Dysplasia

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✔ Renal dysplasia is a disorganized development of the kidney because anomalous differentiation of the metanephrosis. Abnormalities of the collecting system are common. They include obstruction of the ureteropelvic junction, ureteral atresia and urethral obstruction.
The case was a six months old female child with ultrasonografically detected right hydronephrosis and oligohydroamniosis at the 20 th week of gestation. She underwent an operation for bilateral hydroureteronephrosis at 1 month of age. There was no serious and related illness in family history and self-history. She underwent right nephrectomy at 6 months of age. The specimen included an atrophic nephrectomy material with lobulations and a dilated ureter. When sectioned, it was observed that the pelvicaliceal areas were dilated and the cortex-medulla border was obscured.
On microscopic examination, the cortex was thin, contained rare small glomerules, primitive ducts and mononuclear cell infiltration were seen. A focus of cartilage was observed at the cortex-medulla junction. The case was diagnosed as renal hypoplastic dysplasia. During 4 months of follow-up, no complication was reported.
This case is presented considering the uncommon occurence of renal dysplasia and its differential diagnosis from other congenital cystic renal malformations.
Key words: *Renal dysplasia, cystic renal malformation, urethral obstruction*

✔ **Renal Hipoplastik Displazi**
Renal displazi, metanofreoz sırasında diferansiasyon anomalisine bağlı olarak böbrek gelişimindeki bir bozukluktur. Toplayıcı sistem bozuklukları sıktır. Bu anomaliler; üreteropelvik bileşkenin obstrüksiyonu, üreteral atrezi ve üretral obstrüksiyondur.
Olgu; 20. gebelik haftasında ultrasonografi ile sağ hidronefroz ve oligohidramnioz belirlenen 6 aylık kız çocuğudur. Hasta bir aylık iken bilateral hidroüreteronefroz için opere edildi. Hastanın özgeçmişi ve soygeçmişinde hastalığı ile ilişkili ciddi bir rahatsızlığı belirlenmedi. 6 aylık iken sağ nefrektomi uygulandı. Nefrektomi materyali, dilate üreter ve lobülasyonlar içeren atrofik bir böbrekten oluşmaktaydı. Kesitinde pelvikalisiyel alanların dilate olduğu ve korteks-medulla sınırının seçilemediği gözlendi.
Mikroskobik incelemede, korteks oldukça ince gözlendi ve medullada az sayıda küçük glomerüller, primitif duktuslar ve mononükleer iltihabi hücre infiltrasyonu izlendi. Korteks-medulla bileşkesinde kartilaj odağı belirlendi. Olgu; renal hipoplastik displazi olarak tanı aldı. 4 aylık takibi sonrasında olguda komplikasyon belirlenmedi.
Bu olgu, renal displazinin nadir görülmesi ve diğer konjenital kistik renal malformasyonlardan ayırımı açısından sunulmaktadır.
Anahtar kelimeler: *Renal displazi, kistik renal malformasyon, üretral obsruksiyon*

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INTRODUCTION

Dysplastic kidneys are abnormally differentiated, as shown by abnormal structural organization with abnormally developed metanephric elements. The features that can be regarded as clearly dysplastic are metaplastic cartilage, primitive ducts, and lobar disorganization. Most cases are associated with ureteropelvic obstruction, ureteral agenesis or atresia, and other anomalies of the lower urinary tract. The disorder is the most common form of cystic renal disease in children and the most common cause of abdominal masses in newborns. Most dysplastic kidneys arise sporadically, but few are familial or occur in syndromes of multiple malformations. Dysplasia can be unilateral or bilateral and is usually cystic[1–3].

The clinical presentation largely depends on the extent of the dysplastic involvement and the degree of associated urinary obstruction[3]. In gross appearance, the kidney is usually enlarged, extremely irregular, and multicystic. The cysts vary in size from microscopic structures to spaces of several centimeters in diameter[2]. On histological examination, they are lined by flattened epithelium. Although normal nephrons are present, many have immature ducts. The characteristic histological feature is the presence of islands of undifferentiated mesenchyme, often with cartilage[2]. Abnormalities of the collecting system are common and they include obstruction of the uretero-pelvic junction, ureteral atresia, and urethral obstruction[3]. These renal abnormalities bear a strong relationship to other urinary tract malformations, including ureteral atresia and urethral valves, suggesting that urinary obstruction or urinary reflux during metanephric development leads to renal dysplasia[1]. Malformations of other organs, especially of the heart, can occur in conjunction with renal dysplasia[2].

CASE REPORT

The case was a six months old female child. Bilateral mild hydroureteronephrosis and oligohydramniosis were detected with prenatal ultrasonography at the 20 th week of gestation. After birth, the patient evaluated with appropriate diagnostic procedures and bilateral hydroureteronephrosis, right non-functioning kidney, bladder outlet obstructing left ureterocele with double collecting system were determined. Thereafter, she managed with suppressive antibiotic treatment and close follow-up. At 1 month of age, due to clinical worsening she underwent an operation (Open ureterocelectomy and bilateral ureteroneocysistectomy). On the postoperative sixth month, right nephrectomy was executed due to resistant urinary infection episodes with no functional improvement. There were no serious and related illnesses in her family history or self-history.

The specimen included an atrophic nephrectomy material with lobulations and a dilated ureter. When sectioned, it was observed that the pelvicaliceal areas were dilated and the cortex-medulla border was obscured.

On microscopic examination, the cortex was thin and in the medulla a few small glomerules, primitive ducts, mononuclear cell infiltration were seen. In some areas lymphoid follicules and arteriolar hyaline change was noted. A focus of cartilage was observed at the cortex-medulla junction (Figure: 1–5). The case was diagnosed as renal hypoplastic dysplasia. During 4 months of follow-up no complication was reported.

Figure 1. Dysplastic kidneys cortex and pelvicaliceal area (H&E, x4).

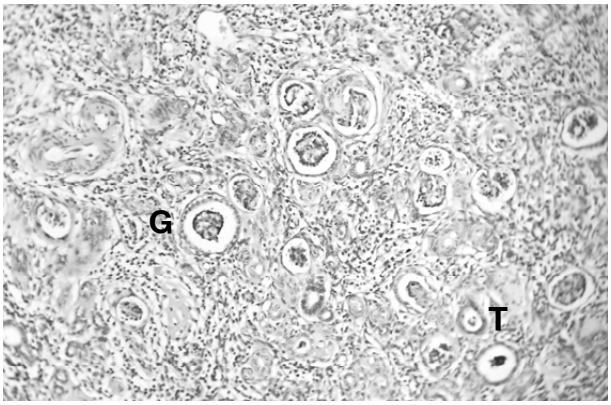

Figure 2. Small glomerules (G), primitive ducts (T) (H&E, x20).

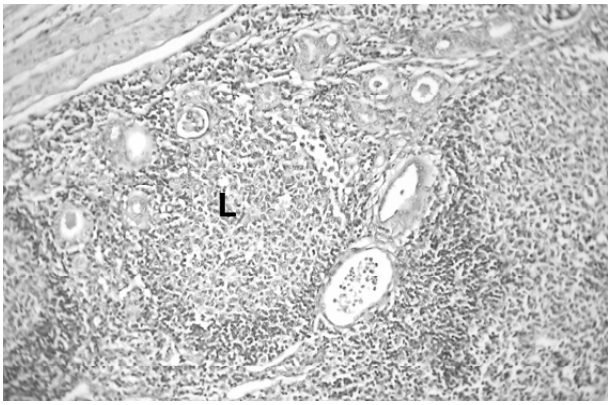

Figure 3. Chronic pyelonephritis, lymphoid follicule (L) (H&E, x20).

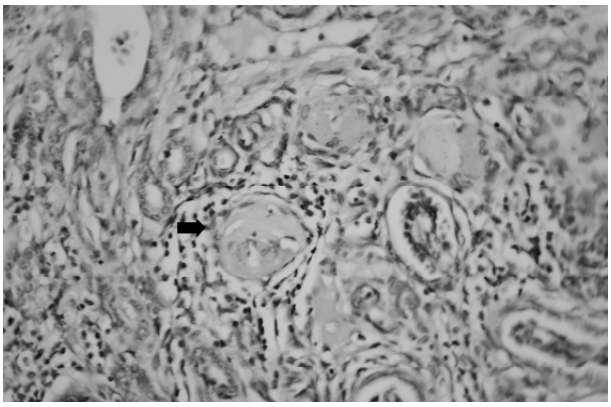

Figure 4. Hyaline arteriolosclerosis (Arrow) (H&E, x40).

This case is presented considering the uncommon occurrence of renal dysplasia and its differential diagnosis from other congenital cystic renal malformations.

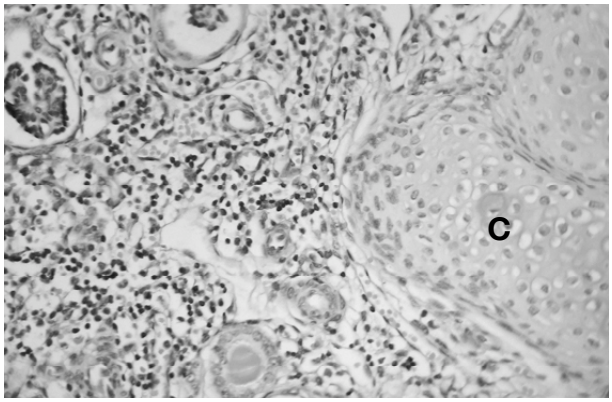

Figure 5. Dysplastic cartilage (C) in kidney parenchyma (H&E, x40).

DISCUSSION

Human renal dysplasia is a collection of disorders in which kidneys begin to form but then fail to differentiate into normal nephrons and collecting ducts. Dysplasia is the principal cause of childhood end-stage renal failure. A retrospective analysis of pediatric autopsies in the past 18 years was done with the aim of studying the histomorphology of renal dysplasia. Renal dysplasia comprised 150 (3.66%) of the 4,099 pediatric autopsies from 20 weeks of gestation to 1 year of life. Primitive ducts with the fibromuscular collar, and renal dysplasia, was seen in all cases. Lobar disorganization and cysts were seen in all cases except for the 7 cases of hypodysplasia[4]. Incidence of unilateral multicystic disease of kidney (MCDK) is reported to be one in 4300 live births, and the combined incidence of unilateral and bilateral MCDK is one in 3600 live births. Bilateral MCDK occurs in about 20% of prenatally diagnosed cases of MCDK. The left kidney is involved in 55% of cases, and the right kidney is involved in 45%. Renal dysplasia and hypoplasia are the two most frequently seen urinary malformations associated with MCDK[5]. Two main theories have been considered in its pathogenesis: A primary failure of ureteric bud activity and a disruption produced by fetal urinary flow impairment[6]. Macroscopically a large reniform mass of cysts of various sizes

obscures any renal parenchyma that may be present. In focal and segmental dysplasia, only part of the kidney is involved by the dysplasia and cyst formation[3]. This sporadic disorder is characterized histologically by the persistence in the kidney of abnormal structures; cartilage, undifferentiated mesenchyme, and immature collecting ductules and by abnormal lobar organization[2]. Metaplastic cartilage customarily appears within the cortex as bars and nests of hyaline cartilage. Primitive ducts, which may be cystic, are altered collecting ducts lined with undifferentiated epithelium and surrounded by fibromuscular collars. Incomplete and abnormal corticomedullary relationship and rudimentary medullary development constitute lobar disorganization. The incompletely developed medullary pyramids are deficient in vasa recta and Henle's loops and are associated with incomplete calyceal and forniceal development[1]. Recent theories have suggested the role of extracellular matrix proteins in the genesis of renal dysplasia. During normal nephrogenesis, collagen type I, III, and fibronectins are lost and laminin and syndecan appear once proper induction has occurred. Any deviation from the normal pattern is said to lead to dysplasia[7].

Experimental models of ureteral or urethral obstruction in fetal animals resulted in renal dysplasia and in utero decomprassion of obstruction prevented to the development of this disease. In utero urinary tract obstruction is one of the most important factors in the pathogenesis of renal dysplasia, but its pathogenesis is still unclear. In Shibata et al's study 16 fetal multicystic dysplastic kidneys and three fetal obstructive renal dysplasia were analyzed with light microscopy, immunohistochemistry and scanning electron microscopy. According to Shiba, urinary obstruction in utero may result in fluid stasis at the functioning glomeruli, leading to glomerular cysts and giving rise to initial pathology of dysplasia. Renal dysplasia has been showed in several congenital syndromes like Prune-Belly syndrome, megacystis micro-colon intestinal hypoperistaltis syndrome, bilateral renal dysplasia with Potter's facies[8–11]. In our case, there was no accompanying syndrome.

When unilateral, the dysplasia is discovered by the appearance of a flank mass that leads to surgical exploration and nephrectomy. The function of the opposite kidney is normal, and such patients have an excellent prognosis after surgical removal of the affected kidney. In bilateral renal dysplasia, renal failure may ultimately result[2].

In summary, renal dysplasia is a development abnormality accompanied by heterogeneous disorders. In experimental studies was indicated that the most important reason of dysplasia is urinary tract obstruction in uterine life. Our case shown to have bilateral mild hydroureteronephrosis and oligohydramniosis with prenatal ultrasonografically at the 20 th week of gestation. She had bilateral hydroureteronephrosis, right nonfunctioning kidney, bladder outlet obstructing left ureterocel with double collecting system. In all fetal obstructive uropathy should be researched for maldevelopments of urogenital system, family history, and its nature.

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