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Prikaz slučaja / Case report

MULTICYSTIC DYSPLASTIC KIDNEY – *Case report*
MULTICISTIČNA RENALNA DISPLAZIJA – *Prikaz slučaja*

Matilda Đolai[1,2], Jelena Ilić Sabo[1,2], Sandra Trivunić Dajko[1,3], Tanja Lakić[1,3], B. Krajinović[2], Željka Vrekić[2]

[1] Univerzitet Novi Sad Medicinski fakultet Katedra za histologiju i embriologiju

[2] Klinički centar Vojvodine, Centar za patologiju i histologiju, Novi Sad.

[3] Katedra za patologiju, Medicinski fakultet Univerziteta u Novom Sadu.

Correspondence to:

Prof. dr **Matilda Đolai**
Klinički centar Vojvodine Centar za patologiju i histologiju, Univerzitet Novi Sad Medicinski fakultet, Katedra za histologiju i embriologiju.
Hajduk Veljkova 1-3, 21000 Novi Sad.
mail: matilda.djolai@mf.uns.ac.rs

Key words

Dysplasia, kidney, malformations, fetus.

Ključne reči

Displazija, bubreg, malformacije, fetus

Abstract

Multicystic dysplastic kidney is a congenital anomaly followed by structural disintegration and cystic like changes of kidney shape. This paper presents case of multicystic dysplastic kidney in fetus five months old. By external examination, micrognatia was observed. By internal examination were founded enlargement of lateral cerebral ventricles, macroscopically altered kidneys and hypoplastic bladder. At the place of the left kidney there was a small, rounded, irregular structure, lobulated surface, with diameter 3 mm, while at the place of the right kidney was cyst formation of the same dimension. Both ureters were the corresponding macroscopic properties. Histological analysis of the described structure showed the kidney with abnormal differentiation followed by the primitive glomeruls and tubules, and by cystic dilatation of nephrons. The renal malformation was diagnosed as the multicystic renal dysplasia. Bilateral multicystic dysplastic kidney is a rare anomaly incompatible with life, and its association with malformation of the brain ventricular system is especially rare.

INTRODUCTION

The term renal dysplasia refers to abnormally differentiated kidneys presented as abnormal structural organisation with ill-developed metanephric elements. It may have different presentations that include hypoplasia, aplasia and multicystic dysplasia[1]. Multicystic renal (kidney) dysplasia (MCDK) is the most common antenatally diagnosed cystic renal pathology. It refers to the presence of multiple renal cysts surrounded by dysplastic parenchymal tissue[2].

CASE REPORT

Fetal malformations were found by the ultrasound and clinical examinations. After that Ethics committee approved the termination of pregnancy. The autopsy of a female fetus was performed. The fetus lengthed 23 cm and body massed 240 gr, which corresponded to gestational age of five lunar months. By external examination micrognatia was found. Other external malformations were not presented. The internal examination identified the easily enlarged lateral ventricles of cerebri. In place of the left kidney was found small, irregular, globose fragment. It had finely lobular surface, 3 mm in diameter, which continues to thin exstension that corresponded to ureter. In place of the right kidney was found cystic formation,with diameter of 3 mm, filled with clear content, also continues to thin exstension that corresponded to ureter. Both ureters (macroscopically preserved) were flowed into the bladder, which had reduced diameter and narrow lumen. Histologial analysis of the samples from described rudimentary kidneys showed the abnormal differentiation of renal tissue with the dilatation of primitive glomeruls and tubules, followed by cystic dillatation of nephrons, mostly in the left kidney (Fig. 1), and with rest of preserved parenchyme in right kidney (Fig. 2). On the basis of macroscopic and microscopic analysis of fetus and renal tissue, the final diagnoses were made: *Dysplasia multicystica renum bilateralis, Hypoplasia vesicae urinarie, Micrognatia and Ventriculomegalia cerebri bilateralis*.

DISCUSSION

Congenital abnormalities of kidney and urinary tract include a wide range of possible malformations. In relation to the total number of malformations, a quarter belongs to abnormalities of the urinary tract, with an incidence of 3-6

in 1000 live births. Abnormalities of kidney are the most frequently congenital urinary tract abnormalities. Congenital abnormalities of the urinary tract are a cause of mortality of 1 in 2000 newborns[3,4.5].

Multicystic dysplastic kidney is a variant of renal malformation. Etiology of MCDK (*Multicystic Dysplastic Kidney*) is unknown[6]. In 1955, MCDK was identified as a separate entity. Earlier, it was generally clustered with polycystic kidneys, and by then it was often diagnosed as polycystic kidneys[7].

Multicystic dysplastic kidney, unilateral or bilateral, is present in 30% of renal abnormalities[8], with an incidence of 1 in 4300 live births[9].

MCDK is often identified in association with other anomalies, especially with the anomalies of the urogenital tract. It is usually associated with abnormalities of the contralateral kidney or vesicoureteral reflux[10]. It can also be a part of congenital syndromes: *Meckel-Gruber*, *Zellweger*, orofatial-digital, *Jeunen*, *Von Recklinghausen*, *Turner ect*[11,12].

The outcome of MCDK depends on whether it is unilateral or bilateral. Complete involution rates vary from 19-75% identified cases of unilateral MCDK[10]. Etiology of involution is unknown[10]. Involution occurs in a period of several months to ten years, and therefore conservative treatment of unilateral MCDK[10,11] is recommended. Bilateral MCDK is lethal congenital disorder, considering that both kidneys are afunctional such in presented case[11].

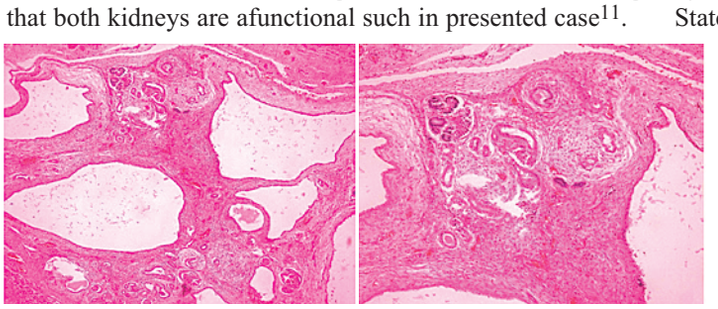

Fig 1. *Left kidney, HE, magnification 5x and 10x.*

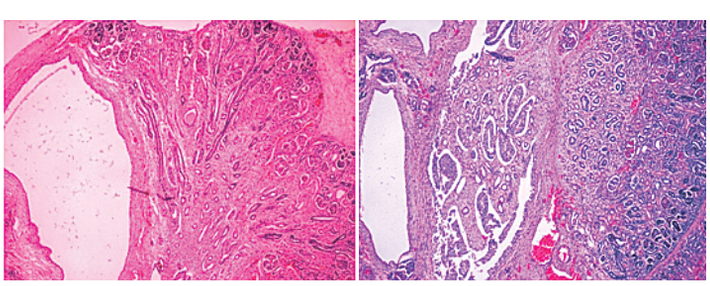

Fig 2. *Right kidney, HE, magnification 5x.*

Grossly in MCDK the kidney can be either small or enlarged, or a normal size . The cut surface shows numerous cysts with different dimension. Histologically, the renal tissue is characterized by structural disorganization. The epithelium is undifferentiated and primitive ducts are surrounded by fibromuscular connective tissue (such as in presented case)[11,13]. There are tree different forms of MCDK: classic, hydronephrotic and solid cystic displasia. The most common form is classic and is always associated with an atretic ureter. The hydronephrotic form presents with a dilated renal pelvis surrounded by cysts, while the solid cystic dysplasia is composed of extremely small cysts with nonfunctional stroma[14].

Renal dysplasia is a form of abnormal renal parenchymal development resulting from anomalous differentiation of metanephric tissue during embryogenesis[1] probably caused by the mutation of the 6th chromosome or in association with autosomal dominant hereditary disorders[15] or mutation in SIX 1 and PAX 2 genes[2].

The association of MCDK and hypoplasia of the bladder is not common. However, the study conducted in 2010, on the territory of the Republic of Serbia registers mentioned association of malformations[16]. Micrognatia is malformation usually present in association with *Poter* syndrome (same in presented case). Association of MCDK with *Poter* syndrome is often described in the literature, which is seen by the results of studies performed in India, the United States and Serbia[15,17]. In this case there is an association of MCDK with ventriculomegaly of cerebri. This is a rare association of malformations previously reported in the literature, also as a case report[16].

It is important to distinguish autosomal recessive polycystic kidney disease and MCDK. The main difference is that in autosomal recessive polycystic kidney disease, the kidneys are enlarged, hyperechogenic, while in MCDK the kidneys could be enlarged, unchanged or reduced size. Histologically, polycystic kidneys are composed of large number of a small cysts, vaguely defined by corticomedullary border[17,18] while in MCDK unconnected cysts are surrounded by immature kidney tissue. The only accurate and definitive diagnoses of these two entities is by histological analysis[18]. Distinguishing between these two malformations is essential, because the risk of recurrence of MCDK in subsequent pregnancy is 3%, while in autosomal recessive polycystic kidney disease is 25%[18].

Bilateral MCDK is incompatible with life. In the case of unilateral MCDK it is significant association with malformations of urinary tract and other organ systems. Therefore, detailed prenatal and postnatal diagnoses in the direction of discovering other malformations is required.

Sažetak

Multicistična renalna displazija je urodjena (kongenitalna) malformacija bubrega koju karakteriše strukturalna dezintegracija bubrežnog parenhima i cističan izgled bubrega. U radu je prikazan slučaj multicistične renalne displazije kod fetusa starosti pet lunarnih meseci. Pregledom ploda od spoljašnjih malformacija nadjena je samo mikrognatija. Unutrašnjim preglednom fetusa nadjena je obostrana dilatacija obe moždane komore, narušena i cistična struktura oba bubrega i hipoplazija mokraćne bešike. Na mestu levog bubega nadjena je mala, iregularna, ovalna struktura, lobulirane površine i prečnika 3mm, dok je na mestu desnog bubrega nadjena mala, policistična formacija, prečnika 3mm. Oba uretera su očuvanih makroskopskih karakteritika. Histološkom analizom tkivnih uzoraka sa mesta bubrega, uočeno je da se nalazi bubrežni parenhim sa primitivnim glomerulima i tubulima i cistična dilatacija nefrona i malformacija je nazvana bilateralna multicitična renalna displazija. Bilateralna multicistična displazija je retka malformacija koja je inkopatibilna sa životom. Prema literaturnim podacima udruženost ove malformacije sa malformacijama ventrikularnog sistema moždanih ventrikula je izuzetno retka.

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