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GLOMERULOCYSTIC KIDNEY
A CASE REPORT

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This paper presents a rare type of renal cystic disease involving the Bowman's capsule: the glomerulocystic disease associated with multiple malformation. Etiology or pathogenesis of glomerulocystic kidney remained unclear.

INTRODUCTION

Glomerulocystic kidney is characterised by dilatation of Bowman's capsule without intra- or extrarenal obstruction /4, 5, 7-10/. In the literature about 20 cases could be found. The pathogenesis is not known.

CASE REPORT

A 2950 g weight male infant was born with Apgar 6-7 from the fourth pregnancy of his mother. He was a product of a 41 - 42 weeks' gestation. His mother did not take medicine during her pregnancy which was symptomless. After his birth the boy was cyanotic. His breathing was feeble, superficial. He was resuscitated several times. The lumbal liquor was massively bloody. The infant died in the 21st hour.
At autopsy diffuse subarachnoideal hemorrhage was observed with massive cerebral edema. We found multiple malformations, palatoschisis, clubfeet, coarctation of the aorta, abdominal visceral transposition, ectasia of the Botallo's duct and accessory spleen. Ren arcuatus was observed with double pyelon and ureter. One of them was megaloureter. The urinary bladder and urethra were normal. Sectioning of kidney demonstrated multiple small cysts, 1-4 mm in diameter, diffusely scattered throughout the renal cortex. There were no cysts in other organs. Cystic dilatation of Bowman's capsule was the most striking abnormality (Fig. 1). The cysts were lined by flatt-

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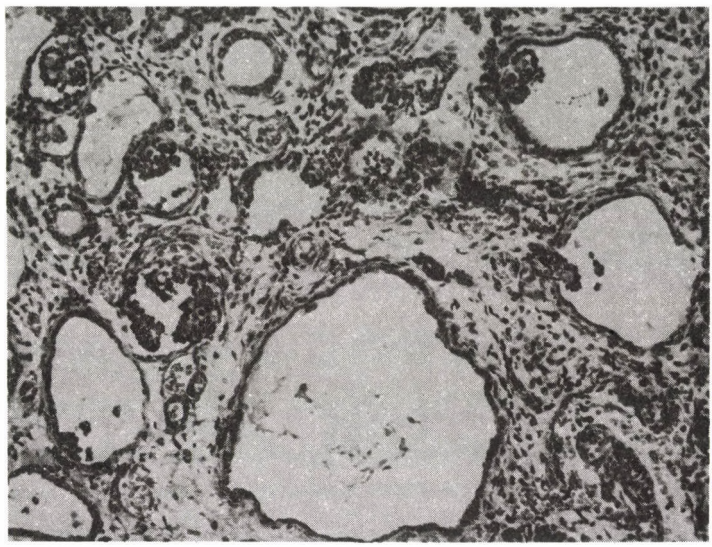

Fig. 1 Multiple glomerular cysts with compressed glomerular tufts (Periodic acid-Schiff stain, x 150)

ened or cuboidal primitive epithelium. The glumerular tufts manifested regressive and atrophic changes (Fig. 2). The Bowman's spaces were filled by PAS-positive proteinaceous material. Among the cystic glomeruli atrophic, sclerotic and primitive glomeruli could be found. The lumen of the proximal tubuli was focal and mild. The vessels were normal. Histological diagnosis: Glomerulocystic kidney.

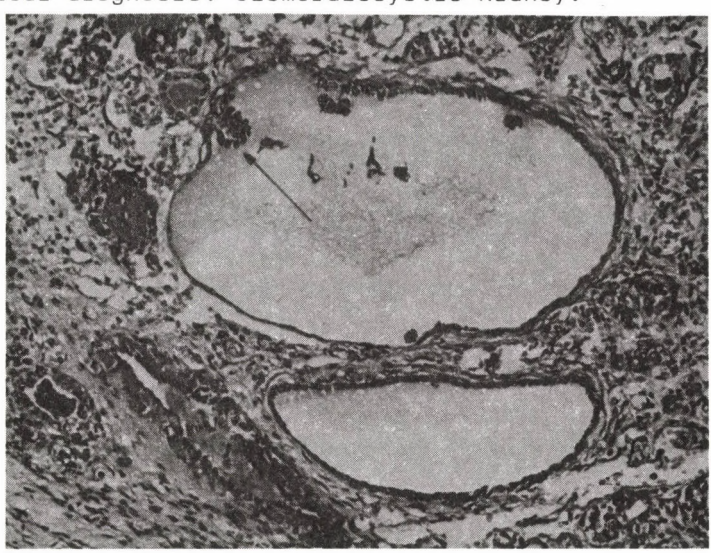

Fig. 2 Two glomeruli with dilated urinary spaces and collapsed glomerular tufts (Periodic acid-Schiff stain, x 200)

DISCUSSION

Glomerulocystic kidney is a rare, distinctive entity. Roos /7/ described this lesion in 1941, but Taxy and Filmer /9/ analysed it morphologically. They proposed the term "glomerulocystic kidney" for this disease. This bilateral renal lesion is characterised by dilatation of the glomerular Bowman's capsule, diffusely in the renal cortex. Tubular cysts are never associated with cystic glomeruli. The glomerular tufts are always atrophic. The Bowman's capsule is lined by flattened epithelium or podocytes /7/. Scattered sclerotic and atrophic glomeruli could also be seen /5/ as it was in our case too. Authors demonstrated interstitial fibrosis, calcification and inflammation /3, 5/. Glomerulocystic kidney occurs mostly in infants, but was observed in childhood, too /10/. The clinical symptoms are hematuria, cylindruria, azotemia and albuminuria. Symptomless cases were also described. In our case urinalysis was not performed, but we could not observe renal functional deterioration. Etiology and pathogenesis of this disease are unknown. Theoretically the etiologic role of chromosomal alterations /1/, renal medullary inflammation with tubular obstruction /4/, drugs (phenacetine, corticosteroid), toxins, viral infection /5/ may come up. Other organ malformations were observed in patients with glomerulocystic kidney, and so was in our case, too /1/.

The glomerulocystic kidney can relatively easily be distinguished from the infantile type polycystic kidney, which is an autosomal recessive hereditary disease. In the familial dysplastic cystic kidney /1, 3/ dysplastic tissue /cartilage etc./ can be found. The multicystic kidney is usually unilateral and associated with ureter obstruction and/or atresia.

This article is the first in Hungary to describe a case of glumerulocystic kidney.

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