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Glomerulocystic kidney: an unexpected finding in preimplantation biopsy

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Glomerulocystic kidney disease (GCKD) is a very uncommon entity that encompasses a wide group of kidney diseases characterized by cystic dilation of Bowman's space associating collapse and retraction of glomerular clews. It can appear both in children, usually in the context of congenital disease, and in the adult, more common of acquired etiology. Only a few cases have been published to date. We report a case of special interest due to its finding in a preimplantation biopsy from an asymptomatic donor.

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Introduction

Histopathological evaluation of pre-transplant donor biopsy can provide useful information to accept grafts from expanded criteria donors. We report a case of glomerulocystic kidney disease (GCKD) diagnosed in a deceased donor.

Case Presentation

A 49-year-old man was considered as a kidney donor. He had a history of hypertension, and hypertensive heart disease, obesity, obstructive sleep apnea syndrome and an ischemic stroke 8 years ago.

He was admitted to the intensive care unit in a deep coma situation due to a massive subarachnoid hemorrhage with data of irreversible neuronal damage. In a few hours, brain death was declared and organs and tissue donation were requested to family, who accepted.

At the time of donation, serum creatinine was 1.1 mg/dL without proteinuria. Ultrasound study showed normal size and morphology kidneys with good vascularity and slightly thinned cortical (1 cm). Due to his personal history, a pre-implantation kidney biopsy was requested.

It showed suggestive findings of GCKD, so graft was rejected to the implant.

Kidney macroscopic study showed an increased cortex with an apparent loss of corticomedullary demarcation (Figure 1). Light microscopic study showed atrophy and cystification of more than 50% of glomeruli at different stages of evolution (Figure 2). Interlobar arteries showed mild to moderate atherosclerosis since interstitium showed no fibroinflammatory changes (Figure 3).

Discussion

GCKD is a rare entity, described mainly in children and neonates and characterized by cystic dilation of Bowman's space associating collapse and retraction of glomerular clews.

According to its etiology, five different types are described; 1) associated with polycystic kidney disease (dominant or recessive), 2) familial or associated with inherited diseases (different genes described), 3) syndromic (not obstructive), 4) syndromes with obstructive pathology (with or without dysplasia) and 5) acquired (1-2).

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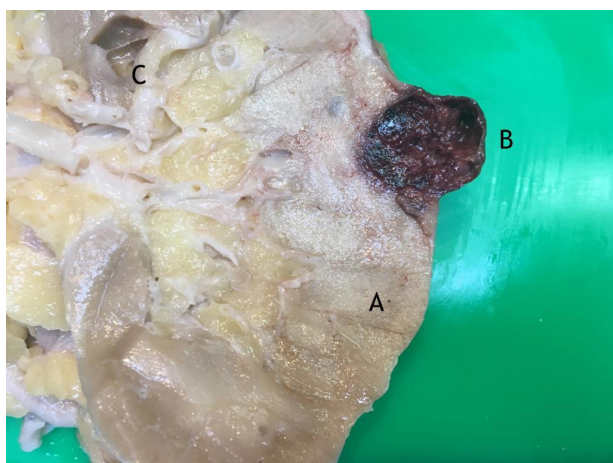


Figure 1. Macroscopic cut; slightly thinned cortical (A), hemorrhage secondary to wedge cut for microscopic study (B), simple cyst (C).

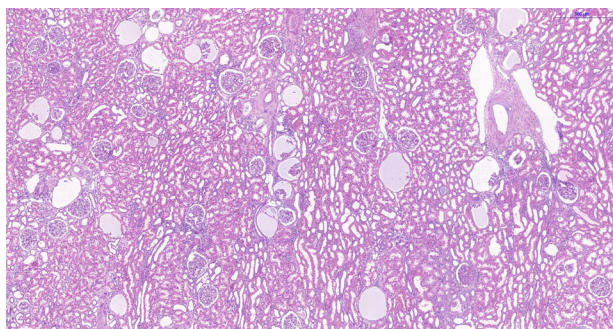


Figure 2. Light microscopy. Hematoxylin and eosin stain section showing glomeruli at different stages of cystification. Interstitium and tubules are without alterations.

Symptoms are nonspecific and very variable. Patients mainly present high blood pressure with difficult control and/or progressive kidney failure. Adults can also have a silent or pauci-symptomatic course. The diagnosis at earlier ages is related to a worse prognosis and a greater risk of progression to end-stage chronic kidney disease (2). In our patient, the absence of another associated pathology oriented in a sporadic or acquired form. In this regard, Lennerz et al propose two subcategories; ischemic and drug-induced. The ischemic form can be unilateral and associates vascular and systemic ischemic changes.

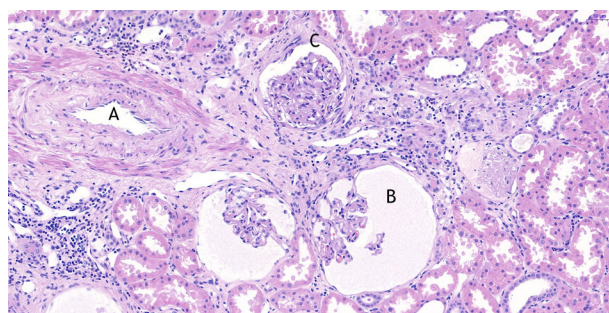


Figure 3. Light microscopy. Hematoxylin and eosin stain section, arteriole with mild-moderate intimal hyperplasia (A) next to cystic glomeruli (B) and a healthy glomerulus (C).

According to the personal history of our patient, this could be the most likely etiology (1).

Authors' contribution

MMR, FV, TG and AM wrote the paper and revisited the case. AC, JP and JMB reviewed samples and reported pathology results. AG was the physician of the patient and completed data and helped in writing draft. All authors read and signed the final paper.

Conflicts of interest

There is no conflict of interest in this paper.

Ethical considerations

Ethical issues including plagiarism, double publication, and redundancy have been completely observed by the authors. The family of the patient gave the consent to publish as a case report.

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