AUTOSOMAL DOMINANT POLYCYSTIC KIDNEY DISEASE AND LIVER INVOLVEMENT IN AN END-STAGE KIDNEY DISEASE PATIENT

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ABSTRACT

None

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Introduction

Autosomal Dominant Polycystic Kidney Disease (ADPKD) is the most common life-threatening hereditary disorder characterized by cyst formation and enlargement in the kidney and other organs. Hypertension is one of the first signs of renal disease development and is related to progressive kidney enlargement and loss of renal function[1].

The major extrarenal manifestation of ADPKD is Polycystic Liver Disease (PLD). Often these cysts are incidental findings and clinically insignificant, remaining asymptomatic until adulthood.

The symptoms become more frequent with age and increase as a result of increased life expectancy, especially in patients with ADPKD due to dialysis and transplantation. The symptoms related to PLD may result from mass effects or complications related to the cysts[2].

Case report

Female, 81 years old, followed up in Internal Medicine consultation due to Autosomal Dominant Polycystic Kidney Disease. Under renal replacement therapy by hemodialysis (performed by a Central Venous Catheter placed in the Left Internal Jugular Vein). Comorbidities include arterial hypertension, dyslipidemia and osteoporosis. Hepatic polycystosis was described 10 years ago in an imaging exam (Computerized Tomography) performed in the context of hospitalization for renal cyst infection. More recently, hospitalization for CVC infection and verified frank progression of renal and hepatic involvement of Autosomal Dominant Polycystic Kidney Disease, abdominopelvic computed tomography demonstrated exuberant hepatic and renal polycystosis (figures 1, 2 and 3).

Conclusion

A minority of ADPKD patients develop massive cystic involvement of the liver, although most are women, similar to this case[3]. The risk to develop severe cysts of the liver is independent of the ADPKD genotype but is related to the severity of renal disease[1]. We emphasize that despite the great liver involvement and advanced age, the patient has normal liver parameters and no symptoms associated with liver involvement. We highlight the exuberance of kidney and liver lesions in the context of Autosomal Dominant Polycystic Kidney Disease.

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Conflict of interest

There are no conflicts of interest to declare by any of the authors of this study.

References


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Figure 1 – Abdominopelvic CT scan – transversal view. Figure 2 - Abdominopelvic CT scan – sagittal view. Figure 3 - Abdominopelvic CT scan – coronal view.