Case Report
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A Rare Heart Valve Involvement in Adult Polycystic Kidney Disease: A Case Report

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Abstract

In adult polycystic kidney patients, heart involvement is frequently observed in the progressing process of the disease, as well as the pathologies created in various other organs. Among others, left ventricular hypertrophy and mitral valve prolapse are generally observed. A 42-year-old female patient, receiving dialysis due to chronic renal failure resulting from autosomal dominant polycystic kidney disease, was admitted to the cardiology outpatient clinic with complaints of high blood pressure, dyspnea and abdominal pain. During clinical examinations, a cyst was detected in the tricuspid valve on echocardiography. In this case study, we aimed to present a patient with a cyst on the tricuspid valve, which has not been previously encountered in adult polycystic kidney patients.

Keywords: adult polycystic kidney disease, left ventricular hypertrophy, tricuspid valve, giant hepatic cyst, dyspnea, heart failure

Introduction

Autosomal dominant polycystic kidney disease is a disease characterized by the development of widespread cysts in the kidneys[1,2]. Cysts usually develop due to a genetic anomaly associated with epithelial cell differentiation and extracellular matrix dysfunction[3]. Apart from kidney involvement, liver, pancreas, lung and cardiovascular involvements are also present. Cardiovascular disorders are the main complication contributing to both morbidity and mortality[4]. In this case study, we aimed to present a patient with a cyst on the tricuspid valve, which has not been previously encountered in adult polycystic kidney patients.

Case Reports

A 42-year-old female patient, receiving dialysis due to chronic renal failure resulting from autosomal dominant polycystic kidney disease, was admitted to the cardiology outpatient clinic with complaints of high blood pressure, dyspnea and abdominal pain. In contrast-enhanced computed tomography of the whole abdomen, both kidney sizes were observed to be larger than normal and extend to the pelvis level. Multiple cortical and parenchymal cysts were detected on the right and the left kidneys the largest ones were of 62mm and 82mm in diameter, respectively. (see Figure 1)

At this level, densities of millimetric stones were detected between the cysts. There were many cysts in the pancreatic body and tail, and intestinal segments and colon were compressed due to the significant increases in the size of both kidneys. (see Figure 2) A 3cm diameter cyst was observed in the left ovary. When evaluated together with the other findings, it was interpreted as an autosomal dominant type 2 polycystic kidney disease.

In the echocardiography of the patient with shortness of breath, the cystic structure, which was not encountered in any other cases in the related literature, was observed under the tricuspid valve (see Figure 3,4). Although the aortotubular junction was slightly enlarged, left ventricular hypertrophy was observed (Figure 5). Despite the attempts to control the patient's blood pressure by regulating her medical treatment, she passed away two months later due to intracranial hemorrhage.

Discussion

Cardiovascular problems are among the main causes of death in patients with Autosomal dominant polycystic kidney disease (ADPKD)[4]. Hypertension is one of the early symptoms[5]. Left ventricular hypertrophy is the most

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common cardiac finding. Intracranial and extracranial aneurysms and heart valve defects are other potential problems. Our patient had hypertension and left ventricular hypertrophy[6]. The cause of our patient's death was bleeding from an intracranial aneurysm.

Histological analysis of heart valve tissue obtained from lesions obtained from numerous published case series and case reports suggests that it is caused by myxomatous degeneration associated with collagen loss and degeneration[3]. As a matter of fact, in our patient, cystic formation was observed with the degeneration of the tricuspid valve. This histological structural disorder is expected to be multisystemic and involve organs such as lung, liver, pancreas, ovary, kidney, and heart. In line with the literature, our patient had other organ involvements other than kidney.

ADPKD is a systemic disease associated with a variety of extrarenal manifestations, including aortic root enlargement and cardiac valve abnormalities, mostly mitral valve prolapses[7]. In a combined retrospective and prospective study of 11 autosomal dominant polycystic patients, multiple cardiac and aortic lesions were found. Aortic root width was detected in 7 patients with aortic valve insufficiency. Two of them required valve replacement due to severe insufficiency. 3 patients had mitral valve insufficiency and two of them had chordae tendinea rupture[8].

Another study examined echocardiography findings to evaluate the prevalence of cardiac anomalies with 163 patients diagnosed with ADPKD, 130 unaffected family members, and 100 control subjects. The prevalence of mitral valve prolapse was observed as 26%, 14%, and 2% in these three groups, respectively. In addition, tricuspid valve prolapses, along with valvular insufficiencies, were also detected in ADPKD patients[9]. To the knowledge of the researchers, the cystic structure detected in the tricuspid valve in our patient with ADPKD was not previous reported in any cases in the related literature.

Conclusion

The overall cardiac involvement of ADPKD supports the hypothesis that the disease contains a defect in the extracellular matrix, and that cardiac abnormalities are caused by this defect. Therefore, it can be concluded that the cyst development in the tricuspid valve might be suspected in patients with ADPKD.

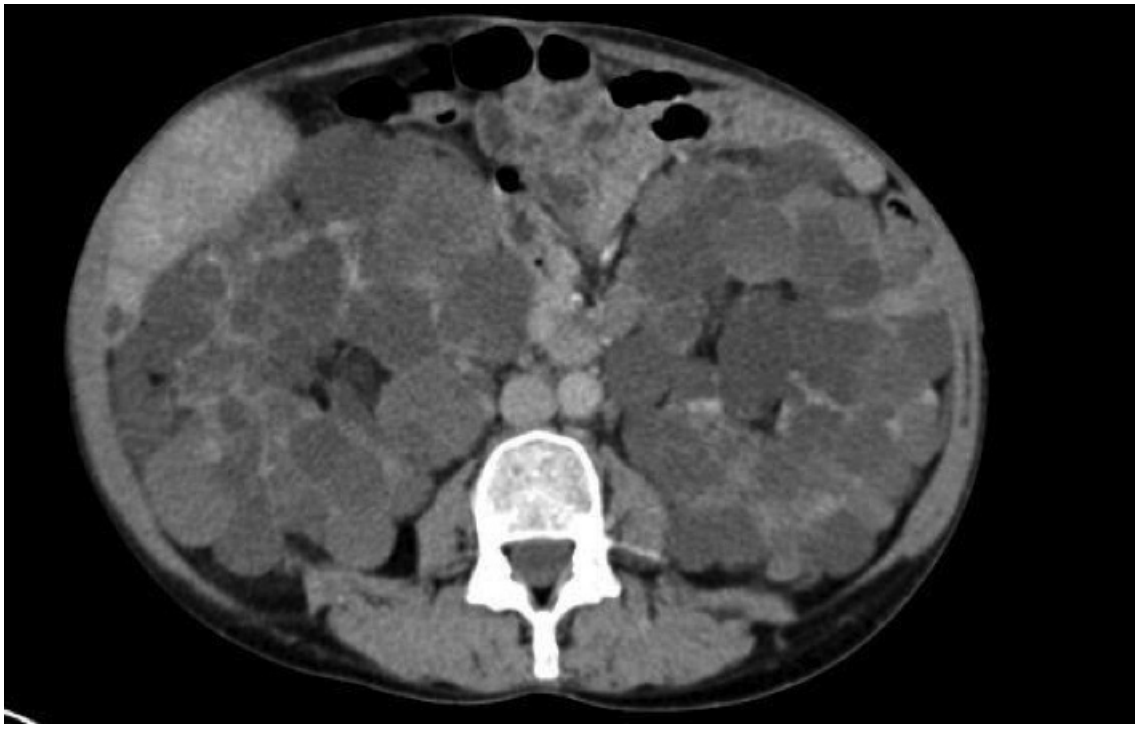

Figure 1: Bilateral Renal Polycystic

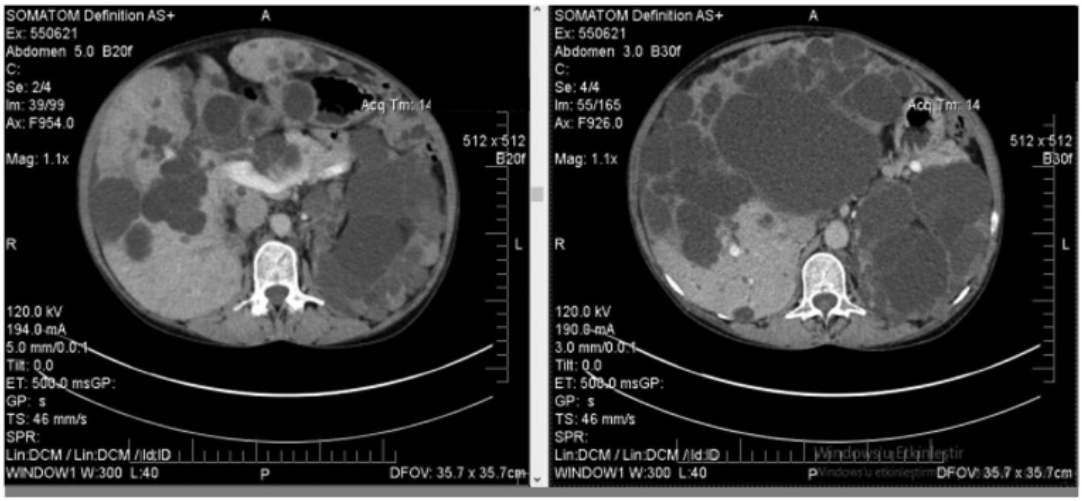

Figure 2: CT image of ADPK disease

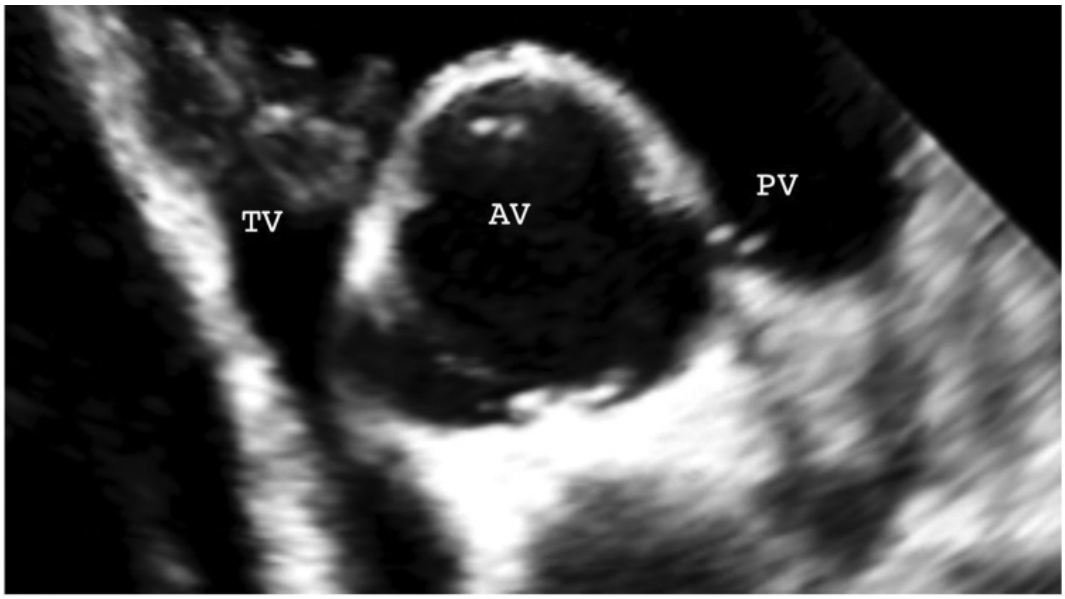

Figure 3: Short axis echocardiography image: involvement of tricuspid valve. (TV is Tricuspid Valve, AV Aorta Valve and PV Pulmonary Valve)

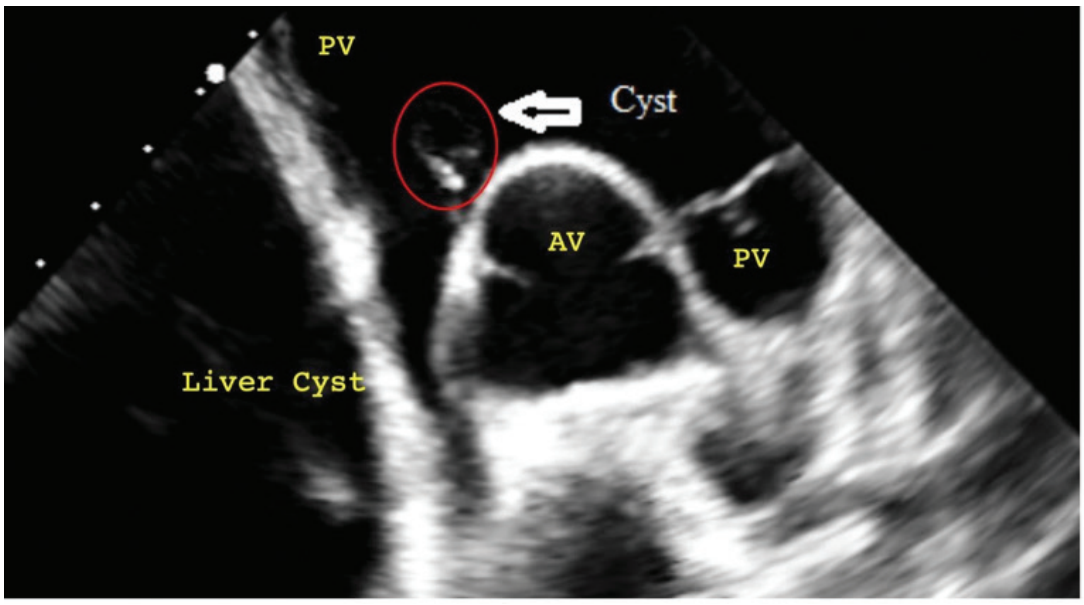

Figure 4: Tricuspid valve opening image (PV is Pulmonary Valve, AV Aorta Valve and PV Pulmonary Valve)

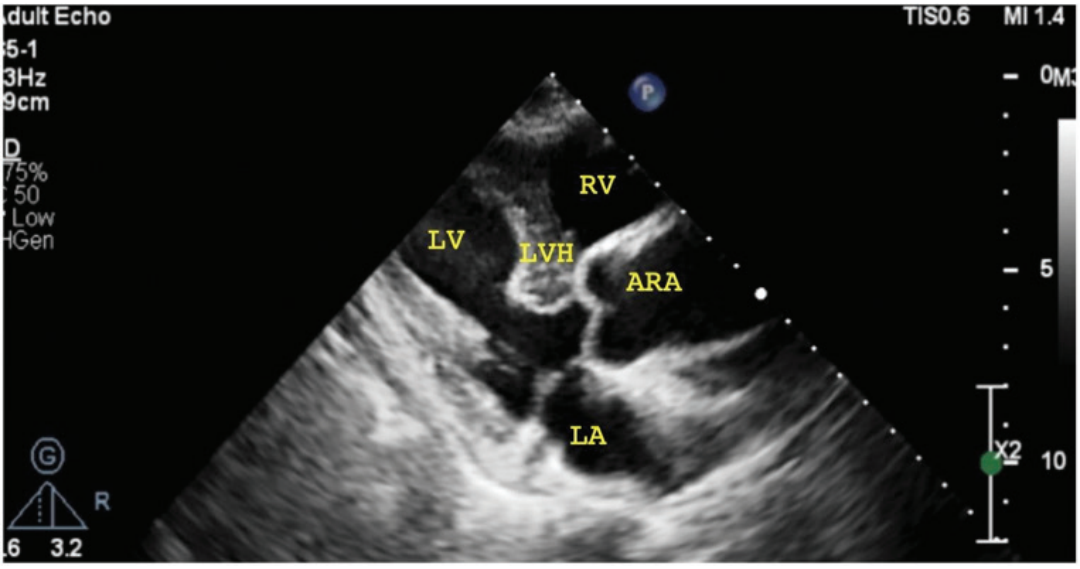

Figure 5: LVH and annular aortic dilatation image
(LV is Left Ventricle, LVH Left Ventricle Hypertrophy, LA Left Atrium, RV Right Ventricle and ARA Aorta Root Annular

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