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Case report

Localized Aortic Dissection in a Patient with Polycystic Kidney Disease: A Case Report

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Abstract

Aortic dissection (AD) is the most frequent life-threatening aortic disorder. It is commonly associated with hypertension; however, aortic dissection occasionally represents a complication of more complex syndromes. In this article we aim to report. A 40-year-old male patient, with a known case of ADPKD and a strong family history of ADPKD. He presented to the emergency department with prolonged sharp retrosternal chest pain radiating to the back and uncontrolled hypertension. Computed tomography angiography showed a localized dissection flap at the aortic root and multiple cysts in the right kidney. AD is a life-threatening condition and should be suspected in patients presenting with acute chest pain with history of ADPKD.

Keywords: ***Aortic Dissection, Polycystic Kidney Disease, Case Report***

Introduction

Aortic dissection (AD) is the most frequent life-threatening aortic disorder [1]. It is commonly associated with hypertension; however, aortic dissection occasionally represents a complication of more complex syndromes [2]. AD can be a complication of extremely rare genetic disorders like autosomal dominant polycystic kidney disease (ADPKD) [3,4]. ADPKD is associated with mutations in the PKD1 or PKD2 gene, which encode the proteins polycystin1/polycystin2, which expressed in the endothelial and smooth muscle cells of the vascular system [5,6]. Clinically, ADPKD is characterized with progressive development of renal cysts, end-stage renal failure and increases the risk of aneurysm formation [7,8,9].

Case report

A 40-year-old male patient, with a known case of ADPKD and a strong family history of ADPKD. He presented to the emergency department with prolonged sharp retrosternal chest pain radiating to the back and uncontrolled hypertension. On examination was remarkable for blood pressure of 190/110 mmHg in both arms. Electrocardiogram showed left ventricle hypertrophy. Echocardiography revealed a suspicion of aortic root flap suggesting aortic dissection, Figure 1. Computed tomography angiography showed a localized dissection flap at the aortic root and multiple cysts in the right kidney Figure 2. His laboratory results were remarkable for whit blood count of 20, blood creatinine of 1.39 mg/dl, blood urea of 7 mmol/l. The patient underwent an emergency Bentall procedure with mechanical valve replacement. post-operatively he developed right leg acute ischemia which was successfully treated with surgical embolectomy. Eighteen days after the surgery the patient was discharged in stable condition on therapeutic warfarin and antihypertensive medications.

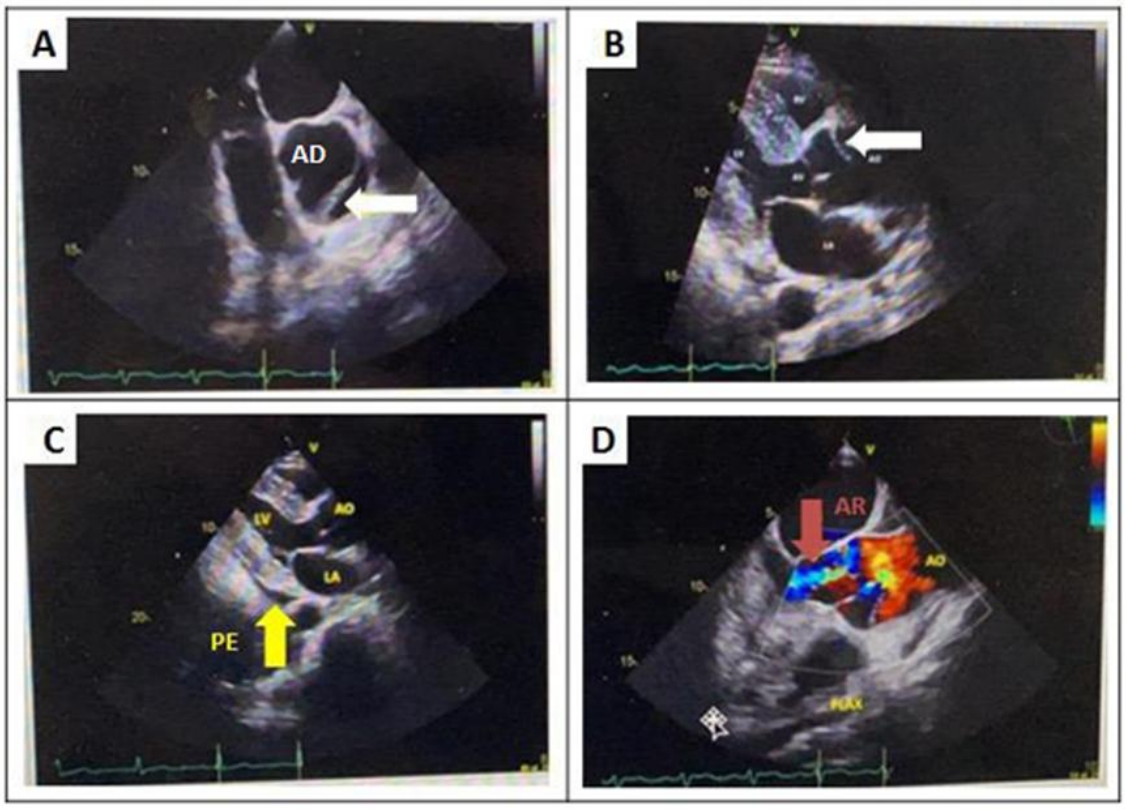

Figure 1: A-D echocardiographic views revealed a suspicion of a flap at the aortic root (white arrow), mild pericardial effusion (yellow arrow), and echocardiographic aortic regurgitation (red arrow).

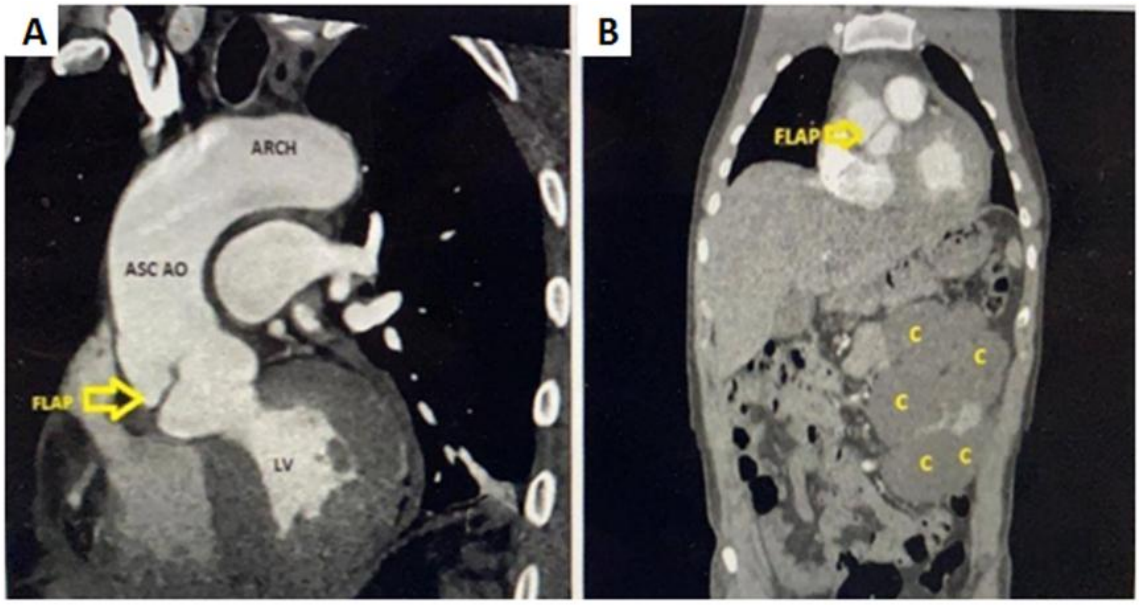

Figure 2: Computed tomography angiography. A: Coronal and B: Sagittal views show a localized dissection flap at the aortic root (open yellow arrow) and multiple cysts in the right kidney (letter C).

Discussion

ADPKD is genetic systemic disorder, affected patients develop cysts in the kidneys and other visceral organs. Aortic dissection and intracranial hemorrhage in well established complications [3]. The most commonly responsible mutations are PKD1 and PKD2 which result in abnormal production of polycystin-1 and polycystin-2 proteins. It is highly expressed in the smooth muscle cell of the blood vessel [10,11]; Therefore, arterial dissections, vascular aneurysms are commonly reported in ADPKD [12]. Rahman et al. [13] reported 363 patients with ADPKD to defined the major causes of death, and he found that, cardiovascular complications account for approximately 46.6% of deaths. AD was responsible only for 2.3% of the overall mortality. The clinical presentation of AD varies between isolated localized AD to diffuse AD accompany with myocardial infarction, renal failure and stroke. Consequently, AD should be always suspected in the the presence of chest pain in patients with ADPKD. CTA is the most important tool and should be considered for AD diagnosis [14].

Conclusion

AD is a life-threatening condition and should be suspected in patients presenting with acute chest pain with history of ADPKD.

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