CASE REPORT

Polycystic Kidney Disease with Coronary Aneurysm and Acute Coronary Syndrome

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Abstract

Autosomal dominant polycystic kidney disease (ADPKD) is a common disorder occurring in approximately 1 out of every 400 to 1,000 live births. An extremely rare association between ADPKD and coronary aneurysms has been determined. No connection between this association and acute coronary syndromes has been identified; however, a few reports have appeared in the literature. We present and demonstrate the very rare association of ADPKD and coronary aneurysm, and briefly review this unusual relationship.

Key words: polycystic kidney, coronary aneurysm, acute coronary syndrome

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Introduction

Autosomal dominant polycystic kidney disease (ADPKD) affects 4 to 6 million people worldwide, and is the reason for dialysis in up to 7 to 10 percent of patients. Marked cystic enlargement of both kidneys is a characteristic of polycystic kidney disease. Affected persons present with hypertension, hematuria, polyuria and flank pain and are prone to recurrent urinary tract infections and renal stones (1, 2). The most prominent extra-renal manifestations include hepatic cysts, gastrointestinal tract diverticula, hypertension, and cardiac valve abnormalities, suggesting the role of ADPKD as an underlying disorder in the connective tissue matrix formation (2, 3). Involvement of the vascular walls may cause arterial aneurysms, most frequently including intracranial berry aneurysms that act as the leading cause of death in these patients. Aneurysms may also be seen through the aorta and very rarely in the coronary arteries (4, 5). In this case, we presented and demonstrated the very rare association of ADPKD and a coronary aneurysm, and briefly reviewed this unusual relationship.

Case Report

A 56-year-old man, who was regularly followed up at a nephrology department because of his transplanted kidney, as a result of end-stage renal disease from of polycystic kidney disease, was referred to our cardiology clinic for evaluation of progressive exertional angina pectoris. He had hypertension, dyslipidemia, and mild renal dysfunction (urea: 56 mg/dL and creatinine: 1.4 mg/dL). He was not a smoker. His family history was positive for coronary artery disease and renal diseases; however, there was no clear evidence of ADPKD history. Electrocardiography on admission showed subacute inferior myocardial infarction. A transthoracic echocardiography showed mild left ventricular hypertrophy and mild mitral regurgitation. Coronary angiography showed a long lesion causing 100% obstruction, from the proximal circumflex artery to its middle portion with late antegrade flow and subsequent lesions causing 60%, 50% and 95% occlusions on the right coronary artery (RCA) (Fig. 1). Also, it revealed an aneurysm in the proximal part of the first diagonal branch of the left anterior descending artery. We performed percutaneous coronary intervention to the RCA lesions. The patient was discharged with medical therapy including dual antiplatelet therapy (DAT) (clopidogrel and aspirin).

Discussion

Aneurismal dilatation of the coronary arteries may be
Congenital or acquired; it occurs in 1.5% of patients studied by autopsy or coronary angiography (6). Coronary aneurysms are most commonly associated with atherosclerosis in the adult population. In the United States, Kawasaki disease is the most common cause in children (7). Destruction of the coronary intima and media may be responsible for the abnormal dilatations (8). Other causes include Takayasu’s arteritis, cerebrotendinous xanthomatosis, and neurofibromatosis (9). The incidence of coronary aneurysm in patients with ADPKD is still unknown. Hadimeri et al found coronary aneurysms in only 4 out of 30 (13%) patients with ADPKD on coronary angiography, excluding minor ecstasies in 5 patients (3). Swan et al found coronary aneurysms in 5 (15%) out of 32 patients by coronary angiography (5). The incidence of atherosclerotic coronary ecstasies or aneurysms in patients undergoing coronary angiography, excluding minor ecstasies and autopsy ranges from 0.2 to 4.9% (10). Therefore, the coronary aneurysm rate in ADPKD patients is higher than that of the general population with suspected coronary artery disease (11). Additionally, Magadle et al suggested that the association of ADPKD and coronary aneurysms in members of the same family supported the hypothesis that local inflammation of the coronary vessels, which leads to the development of coronary aneurysms, is a part of the generalized defect of the extracellular collagen tissue in ADPKD patients, much like a similar disorder in collagen metabolism noted for the Ehler-Danlos (type I and type III) and Marfan Syndromes (11, 12). Hadimeri et al found more vessels with significant stenosis than in the control group (3). Magadle et al detected a high prevalence of coronary aneurysm in a family with ADPKD (11). A few case studies have reported the association between coronary aneurysms and myocardial infarction, suggesting that this association could be caused by thrombus formation within the aneurysm with or without embolisation to the distal vessels (13, 14).

In conclusion, the patient had no clinical, laboratory, or imaginary findings of vasculitis, cerebrotendinous xanthomatosis, or neurofibromatosis. In addition we know that atherosclerosis is the main cause of coronary aneurysms. The plaque mass through the destruction of the coronary intima and media, and the altered post-obstructional flow patterns are believed to be responsible for the atherosclerotic aneurysms. However, in the present case the aneurysm was located on the mid-portion of the first diagonal branch of the left anterior ascending artery, which seems not to have an atherosclerotic plaque, indicating ADPKD as the primary cause of the aneurysm. The second point is that the correlation between ADPKD and aneurysms is not clear yet, and needs further investigation. Therefore, we have not had precise evidence to make a clear-cut recommendation for such cases. However, coronary CT could be used as a non-invasive evaluation method for such patients in order to make a risk analysis. In addition, DAT or anticoagulants might be the treatment of choice according to the patient’s physician, and further interventional therapies might be evaluated according to aneurysm type. In conclusion, this case represents a very rare association of polycystic kidney disease with coronary aneurysm and acute coronary syndrome.

References

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Figure 1. Arrow shows the aneurysmal dilatation in the non-atherosclerotic first diagonal branch of the LAD.

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