Spontaneous Coronary Artery Dissection in a Woman With Polycystic Kidney Disease

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Autosomal dominant polycystic kidney disease (ADPKD), characterized by renal cyst formation, is known to cause such vascular abnormalities as arterial dilatation and dissection. However, spontaneous coronary artery dissection (SCAD) is observed only rarely in patients with ADPKD. We report a patient with ADPKD who developed SCAD and presented with acute myocardial infarction. Her coronary angiography showed a long spiral dissection of the left anterior descending coronary artery. She underwent successful coronary angioplasty with insertion of 3 drug-eluting stents. To the best of our knowledge, this is the first reported case of percutaneous coronary intervention for coronary dissection in a patient with ADPKD. The pathophysiological characteristics of vascular complications in patients with ADPKD are discussed. Polycystins are strongly expressed in human adult vascular smooth muscle cells, and the vascular abnormalities in patients with ADPKD may be related to altered expression of polycystins. Because early recognition and prompt efforts at mechanical reperfusion, if indicated, are crucial for successful management of SCAD, it would be worthwhile to consider SCAD in the differential diagnoses of acute coronary syndrome in patients with ADPKD.


INDEX WORDS: Spontaneous coronary artery dissection; polycystic kidney disease; percutaneous transluminal coronary angioplasty.

Autosomal dominant polycystic kidney disease (ADPKD) is the most common monogenic disorder characterized by the formation of cysts in both kidneys, leading progressively to end-stage renal disease in more than 50% of patients.1 It is considered a systemic disease with the development of cysts in various other organs, such as the liver, pancreas, seminal vesicles, and arachnoid membrane.2 Vascular abnormalities, including intracranial aneurysms and dolichoectasias, aortic aneurysms, and dissections of the aorta and cervicocephalic arteries, are the other important extrarenal manifestations of this disease.3-7 We report a case of spontaneous coronary artery dissection (SCAD) in a patient with ADPKD, resulting in acute myocardial infarction. We believe this to be the first reported case of percutaneous coronary intervention for coronary dissection in a patient with ADPKD.

CASE REPORT

The patient was a 43-year-old woman who had been given a diagnosis of ADPKD 7 years earlier after a screening ultrasound scan. Her father and sister were known to have ADPKD. She was asymptomatic, with normal kidney function and no history of hematuria. She had mild hypertension that was well controlled with perindopril, 5 mg once daily. The patient's creatinine level was 0.8 mg/dL (69 μmol/L), and estimated glomerular filtration rate was 86 mL/min (1.43 mL/s).

The patient experienced central chest pain of moderate intensity while stopping at a gas station and was taken to a local hospital, where an electrocardiogram showed inverted T waves in the anterolateral leads (Fig 1). Peak troponin I level was 4.2 ng/mL (4.2 μg/L; normal, <0.06 ng/mL), and peak creatine kinase level was 269 U/L (normal, <200 U/L).

A diagnosis of non-ST elevation myocardial infarction was made, and she was administered aspirin, clopidogrel, metoprolol, and subcutaneous enoxaparin. She experienced a second episode of chest pain while in the hospital without new electrocardiographic changes. She was then transferred to our hospital for coronary angiography.

The patient was a nonsmoker with no family history of coronary artery disease and had normal serum cholesterol levels. She had no clinical features of connective tissue disorder, vasculitis, or recreational drug use before her symptoms. However, she was overweight (body mass index, 29.9 kg/m²) and had joined an exercise program in a gymnasium 3 months earlier. She had attended the gymnasium 1 day before her presentation.

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The patient’s left ventriculogram showed an akinetic apex with an ejection fraction of 64%. Coronary angiography (Fig 2) showed a long spiral dissection of the left anterior descending coronary artery involving the proximal, mid, and distal segments with slow flow. The left circumflex coronary artery and right coronary artery were normal. The patient underwent successful coronary angioplasty with insertion of 3 sirolimus-eluting Cypher stents (Johnson & Johnson, Miami Lakes, FL) covering the left anterior descending coronary artery dissection (2.75 × 33, 3 × 33, and 3.5 × 18 mm). At the end of the procedure, there was evidence of persistent dissection in the very distal parts of the left anterior descending coronary artery, with normal flow. She was advised to take clopidogrel for 12 months in addition to treatment with aspirin, metoprolol, perindopril, and atorvastatin. Treadmill exercise tests (Bruce protocol) performed 3 and 7 months after presentation were negative for inducible ischemia. Seven months after the procedure, the patient remained asymptomatic.

Figure 1. Electrocardiogram shows deeply inverted T waves in the anterolateral leads.

Figure 2. Coronary angiogram shows (A) a long spiral dissection (white arrows) of the left anterior descending coronary artery and (B) after successful angioplasty with stenting.
DISCUSSION

This is an unusual case of a young woman with mild hypertension as her only cardiac risk factor presenting with acute myocardial infarction secondary to SCAD. The option of coronary artery bypass grafting (CABG) was considered after coronary angiography, but it was believed that if a guidewire could be negotiated into the true lumen, angioplasty and stenting would be successful and less invasive. Potential disadvantages of percutaneous coronary intervention include the risk of restenosis and stent thrombosis. With sirolimus-eluting stents, the risk of in-segment restenosis was reported as 6.4% at 6 to 8 months, and the risk of 4-year cumulative stent thrombosis was 1.2%. Because our patient had 3 overlapping stents inserted, her risk is likely to be higher than average. Seven months after the procedure, the percutaneous coronary intervention strategy appears to have been successful, and the patient remains asymptomatic.

SCAD is a rare cause of myocardial ischemia, with about 350 cases reported in the literature. More than 70% of cases occurred in women, with one-third during the peripartum period. The most frequently affected vessel was the left anterior descending coronary artery (75%), followed by the right coronary artery (20%), and left circumflex coronary artery (4%). Left main coronary artery involvement was seen in only 1% of cases.

SCAD produces a hematoma in the media of the arterial wall, creating a false lumen that can expand further, compressing the true lumen and causing myocardial ischemia or infarction. Patients with SCAD usually present with acute coronary syndrome or sudden cardiac death.

Various conditions associated with SCAD include underlying atherosclerosis, pregnancy, oral contraceptive use, intense physical activity, blunt chest trauma, connective tissue disorders, Marfan’s syndrome, vasculitis, cocaine use, and increased homocysteine levels. In our patient, it is possible that heavy exertion in the gymnasium 1 day before presentation might have contributed to spontaneous arterial dissection.

SCAD is rarely observed in patients with ADPKD. To the best of our knowledge, there has been only 1 case reported in the literature in the past, and ours is the first case of percutaneous coronary intervention for SCAD in a patient with ADPKD.

The prevalence of ADPKD varies from 1 in 1,000 in the US population to 1 in 4,000 in Japan. ADPKD is a genetically heterogeneous disorder caused by mutation in the \( PKD1 \) gene (chromosome region 16p13.3) in 85% of patients and the \( PKD2 \) gene (4q21) in about 15%. Polycystin 1 and polycystin 2 are the protein products of the \( PKD1 \) and \( PKD2 \) genes, respectively. Polycystin 1 is believed to be a receptor for an unknown ligand, whereas polycystin 2 functions as a nonselective cation channel. Polycystins are found on the single primary cilium present on the surface of most mammalian cells. Other locations where polycystin 1 is observed include focal adhesions, desmosomes, and adherens junctions.

Polycystins are strongly expressed in human adult vascular smooth muscle cells. The tunica media, the middle layer of the arterial wall, consists of multiple concentric sheets of elastic lamina with adjoining smooth muscle cell layers. Specialized electron-dense areas known as dense plaques found on the smooth muscle cell membrane and interlamellar elastic fibers provide mechanical coupling between the various layers of the tunica media. Polycystins, located in the dense plaques, probably have an important role in maintaining the integrity of this myoelastic structure of the arterial wall. The observation that mouse embryos homozygous for either the Pkd1 or Pkd2 null mutation die in utero with diffuse vascular rupture and hemorrhage supports the role of polycystins in maintaining vascular integrity. Histological evaluation of the aortic wall in patients with ADPKD with aortic root dilatation has shown evidence of cystic degeneration and collagen disruption. Griffin et al reported altered expression of polycystin in arterial smooth muscle cells and myofibroblasts in specimens of thoracic aortic dissection, cerebral aneurysms, and dolichoectatic arteries from patients with ADPKD. Thus, altered expression of polycystins could be directly responsible for the various vascular complications in patients with ADPKD.

Management of SCAD depends on the clinical presentation, extent of dissection, and area of ischemic myocardium at risk. Early revascularization is essential in patients with refractory
ischemia. CABG is the preferred option in patients presenting with left main coronary artery dissection or multivessel involvement.\textsuperscript{10,13} Successful percutaneous coronary intervention with stenting has also been reported in patients with left main coronary artery dissection.\textsuperscript{9}

In a series of 7 cases of SCAD reported by Roig et al.,\textsuperscript{13} 3 patients were treated successfully with percutaneous coronary intervention, 2 underwent CABG, and 1 patient required cardiac transplantation. Successful medical management with antiplatelets, nitrates, and \(\beta\)-blockers has also been documented in stable patients with SCAD.\textsuperscript{9} Hendiri et al.\textsuperscript{19} reported 6 patients with SCAD, 4 of whom were successfully managed with medications, whereas 1 patient each required CABG and percutaneous coronary intervention. Although successful thrombolysis has been described in patients with SCAD,\textsuperscript{9} its use is controversial because of the potential risk of enlarging hematoma and worsening dissection.\textsuperscript{9,20}

It would be prudent to consider such vascular abnormalities as aortic dissection and SCAD in a patient with ADPKD who presents with chest pain or acute coronary syndrome. Early recognition of SCAD with coronary angiography and, if appropriate, prompt treatment with percutaneous coronary intervention or CABG to reestablish coronary perfusion are crucial for successful management of these patients.

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