ABSTRACT
We report a case of a 46-year-old woman with hypertension and autosomal dominant polycystic kidney disease who presented with chest pain and was found to have spontaneous coronary artery dissection (SCAD) on diagnostic catheterization. We review the pathogenesis, management and prognosis of SCAD. We conclude that in patients with polycystic kidney disease who present with angina pectoris and positive cardiac biomarkers, coronary artery dissection should be considered.

KEYWORDS: Polycystic kidney disease, extra renal, spontaneous coronary dissection

INTRODUCTION
It has been suggested that spontaneous coronary artery dissection (SCAD) could be an extra renal manifestation of autosomal dominant polycystic kidney disease (ADPKD).

Some of the known cardiac manifestations of ADPKD include mitral valve prolapse, left ventricular hypertrophy, as well as aneurysms of the aorta, coronary and intracranial arteries.

Case report
A 46-year-old woman with ADPKD and poorly controlled hypertension presented to the emergency department with sharp, substernal chest pain with associated dyspnea and diaphoresis. The pain started abruptly at rest and was described as moderate intensity, radiating to her shoulders and neck. She reported headache, dizziness and anxiety in the setting of recent psychosocial stressors. The patient was noncompliant with her anti-hypertensive medications, which included Losartan 100mg daily and Metoprolol 100mg twice a day. She denied tobacco, alcohol or recreational drugs use. Her family history was significant for polycystic kidney disease in a first-degree relative but otherwise negative for significant cardiovascular disease.

Her initial blood pressure was 162/87 in the left arm and 171/87 in the right arm with normal heart and respiratory rates. The entirety of the physical exam was unremarkable. The initial Troponin I was measured at 0.09ng/mL (reference <0.04 ng/mL) and peaked at 0.82ng/mL. Serial electrocardiograms (EKGs) revealed normal sinus rhythm with no ischemic changes. A chest X-ray was unremarkable. A transthoracic echocardiogram demonstrated mild hypokinesis of the mid to distal inferolateral wall with preserved left ventricular systolic function. A diagnostic cardiac catheterization revealed dissection of the mid ramus intermedius artery branch (Figure 1) with a thrombolysis in myocardial infarction (TIMI) grade 3 flow. There was 70% stenosis seen immediately proximal to the origin of the dissection flap.

Figure 1. Arrow points towards the site of dissection in the ramus intermedius artery.
No other angiographic evidence of coronary artery disease was identified. At the time of coronary angiography, the patient had been angina free for 24 hours and there was no evidence of left ventricular compromise on echocardiography. After initial anticoagulation, the patient was ultimately managed conservatively with aspirin, a high-dose statin, an angiotensin receptor blocker and a beta blocker to optimize her cardiac risk factor profile. Percutaneous intervention was not performed given the stability of her symptoms and hemodynamics.

**DISCUSSION**

There are only four other reported cases of SCAD in autosomal polycystic kidney disease. In ADPKD, the genes PKD1 and 2 are mutated, compromising the expression of Polycystin 1, a glycoprotein which is responsible for maintaining the structural integrity of the arterial wall. The altered glycoprotein likely is the culprit of arterial dissections in patients with polycystic kidney disease. Familial clustering of aortic artery dissections in polycystic kidney disease suggests a causal association with the altered polycystin. Hypertension, which is a common finding in ADPKD is a known risk factor for SCAD. However, in our patient it was unlikely to be the trigger since our patient did not present with a hypertensive emergency. Previously, segmental arterial mediolysis was demonstrated at pathology in a nonhypertensive ADPKD patient. This supports the hypothesis that there is an inherent deficiency that promotes vascular abnormality in ADPKD patients.

The preferred course of treatment of SCAD is highly variable and can range from optimal medical treatment to invasive therapies. The “conservative approach” which is comprised of medical management, with revascularization reserved for patients with ongoing symptoms or evidence of recurrent ischemia, has been associated with an excellent prognosis. Tweet et al retrospectively evaluated 87 SCAD patients and found a high rate of peri-procedural complications in the 39 patients who underwent PCI. Even a quarter of the interventions that were deemed ultimately successful was complicated by propagation of the dissection flap or the development of an intramural hematoma and thus required additional stent placement. Given these findings, caution should be taken in choosing an invasive approach when patients present with a spontaneous coronary dissection. Nine months after her initial presentation, our patient continues to be asymptomatic.

In summary, we are proposing that spontaneous coronary dissection should be in the differential diagnosis in ADPKD patients who present with chest pain or syncpe.

**References**


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