Coronary Artery Ectasia in a Patient with Polycystic Kidney Disease

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A B S T R A C T
A 44 years old male patient with acute coronary syndrome and polycystic kidney disease is described. Coronary angiography showed ectatic coronary arteries. Polycystic kidney disease is the most common inherited kidney disorder which is commonly associated with extra renal manifestations including cardiovascular conditions. Involvement of vessels may lead to arterial aneurysms which most commonly present as intracranial aneurysms. The rare association between coronary aneurysms and polycystic kidney disease has also been noted.

Introduction
Autosomal dominant polycystic kidney disease (ADPKD) had been associated with cardiovascular abnormalities. We describe a 44 years old man with ADPKD who presented with chest pain and diagnosed with marked coronary ectasia.

Care Report
A 44 years old man presented to our emergency department with typical resting retrosternal chest pain associated with sweating and nausea of one-hour duration. He reported several episodes of ejectional chest pain during the previous week for which he had not sought medical attention. Vital sign on presentation included a blood pressure of 130/85, heart rate of 88 and respiratory rate of 18. He was afebrile. Heart and lung examination was not remarkable. His past medical history included hypertension and polycystic kidney disease (PKD) diagnosed during a screening kidney sonography for a positive family history a few years earlier. Several members of his family including his brother and aunt were also affected and had been on chronic dialysis. His previous creatinine levels were normal. Drug history included enalapril 10 mg twice a day. His electrocardiogram was within normal limits and first set of cardiac enzymes were normal. He was admitted and was put on anti ischemic medications. Echocardiography showed no regional wall motion abnormality and mild mitral valve insufficiency. His symptoms did not recur. Repeat routine lab tests, electrocardiogram and repeat cardiac enzymes were normal. We decided to proceed with exercise stress test which turned out to be positive with ST segment depression of 2 mm in inferior leads. The patient was scheduled for coronary angiography. Coronary angiography revealed ecstatic coronary arteries without any significant obstruction but with delayed washout (Figure 1).

Discussion
Autosomal dominant polycystic kidney disease (ADPKD) is the most common inherited disorder of kidney with a frequency of in 1 of 400-1,0001 and accounts for 7-10% of end-stage renal disease (ESRD) patients on dialysis.2 Along with the advancement of renal replacement techniques for patients with ESRD, cardiovascular complications have emerged as a leading cause of mortality in affected individuals.3 Hypertension occurs in 50-70% of patients with ADPKD and usually begins before any remarkable reduction in kidney function tests.4 A greater prevalence of intracranial aneurysms has also been shown in these patients.5
Cardiac valvular abnormalities have been reported with higher frequency in ADPKD patients. In a study of 228 patients with ADPKD, Timio et al reported mitral valve prolapse in 25%, mitral insufficiency in 30%, tricuspid valve prolapse in 5% and aortic regurgitation in 19% of patients. In a recent study on echocardiographic findings of renal transplant patients with ADPKD valvular abnormalities were found to be infrequent. Screening echocardiography is not routinely recommended for these unless a murmur is noticed on physical examination. The most common extra renal manifestation of the condition is hepatic cysts. Involvement of the vascular walls may lead to arterial aneurysms, which most commonly manifests as intracranial berry aneurysms, yet aneurysms may also be seen in the aorta and seldom in the coronary arteries. Though there is no guideline for evaluation of coronary arteries in ADPKD, coronary CT angiography has been suggested by some authors as a non-invasive evaluation method for risk assessment. Several reports of spontaneous coronary artery dissection in ADPKD patients have also been described. The most commonly used angiographic definition of coronary artery ectasia, is the dilation of the ectatic segment more than 1.5 times larger than an adjacent healthy reference segment. It is reported in 1-5% of coronary angiographies. Krueger et al have documented angina, positive exercise test and pacing-induced ischemia in patients with coronary artery ectasia in the absence of significant coronary obstruction, a phenomenon that they have termed as “dilated coronaropathy”. Besides, acute coronary syndromes have been reported to occur in absence of stenosis. In one study a history of myocardial infarction in the corresponding myocardial territory in 38.7% of patients with coronary ectasia was present. Yet cardiac event rate appears to be low.

The most frequent etiology is coronary atherosclerosis, accounting for more than one half of the cases, but some other conditions may also lead to coronary ectasia. Coronary ectasia has been reported in association with connective tissue disorders such as scleroderma, Ehlers–Danlos syndrome, and polyarteritis nodosa, and also with bacterial infections and the Kawasaki disease. Congenital ectasia has also been noted in a small percentage of patients. The incidence of coronary ectasia in ADPKD is not precisely known. In a study by Swan et al. 5 out of 32 (15%) patients with ESRD due to ADPKD had coronary aneurysms. In another study 4 of 30 (13%) patients had coronary aneurysms. Though the definite rate is not known but it is remarkably higher than the general population.

**References**