

Case Report

Coronary Artery Ectasia: A Case Report and Review of Literature

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ABSTRACT

Coronary artery ectasia is not an uncommon diagnosis and is well described in the literature in large-scale reports. Two patients presented with chest pain and positive stress ECG test for reversible ischemia. Coronary angiogram

showed coronary artery ectasia with no obstructive coronary artery disease. Only medical treatment was recommended.

KEY WORDS: atherosclerosis, coronary angiography, coronary artery ectasia

INTRODUCTION

Ectasia is characterized by irregular, diffuse, saccular or fusiform dilatation of the coronary arteries, which may be isolated or coexist with concomitant obstructive coronary artery disease^[1]. Coronary artery ectasia with or without concomitant obstruction may occasionally be found in association with angina pectoris^[2]. The frequency of this finding is an infrequent diagnosis and is estimated to be between one and 2.5%^[2].

Case 1:

A 52-year-old male presented with effort-induced retrosternal chest pain and referred to left shoulder, with functional class II angina (Canadian classification). He had a 10-year history of hypertension and 12 years of hypercholesterolemia. He had no history of diabetes mellitus, nor family history of coronary artery disease. His pulse rate was 87/minute and blood pressure was 150/85 mmHg. Electrocardiogram was within normal limits.

Laboratory investigations results were as follows: hemoglobin: 14 g/dl, total cholesterol: 6.7 mmol, triglycerides: 2.4 mmol, uric acid: 387 mg and fasting blood sugar: 5.4 mmol.

Transthoracic echocardiography revealed mild left ventricular hypertrophy, normal left ventricular systolic function, EF=58% and diastolic dysfunction. No segmental wall motion abnormalities were noticed. Pulsed tissue Doppler imaging revealed regional systolic and diastolic dysfunction at the anterior septum and the anterolateral segments.

Treadmill exercise ECG test using Bruce protocol revealed positive stress test for reversible myocardial ischemia as there was a flat ST-segment depression of more than 1 mm in V3, V4, V5 after 0.80 second from J point.

Coronary angiography revealed saccular dilatation of proximal segment of left anterior descending coronary artery up to second septal perforator with luminal diameter = 9 mm and luminal diameter of distal artery = 2.4 mm (Fig. 2). There were no obstructive lesions in coronary arteries but there was a slow flow in the distal coronary artery.

The patient was started on clopidogrel tablets once daily for six months, ASA tablets 150 mg once daily, ACE inhibitor tablets 5 mg once daily and statin tablet once daily 10 mg.

Case 2:

A 54-year-old male presented with effort-induced retrosternal chest pain. There was no history of effort induced shortness of breath, no orthopnea nor paroxymal nocturnal dyspnea. He had a 10-year history of hypertension and a five year history of hypertriglyceridemia. There was no personal or family history of diabetes mellitus. His pulse rate was 92/minute and blood pressure was 130/65 mmHg. ECG was within normal limits.

Transthoracic echocardiography revealed mild left ventricular hypertrophy, normal left ventricular systolic function, EF=62% and diastolic dysfunction. No segmental wall motion abnormalities were

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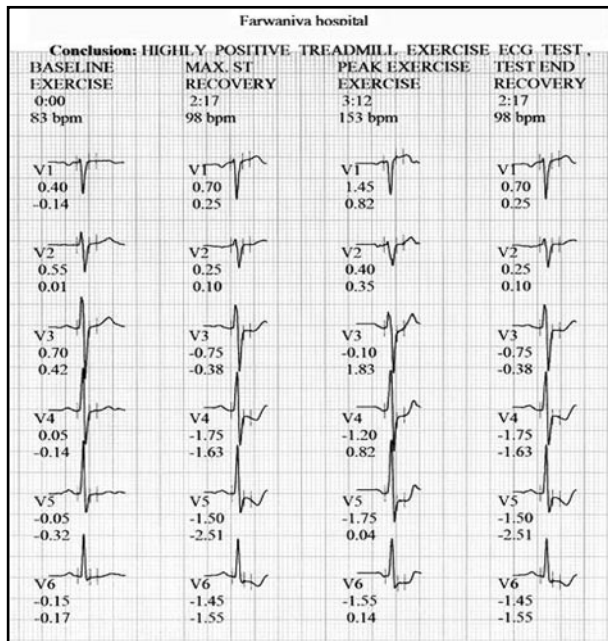


Fig. 1: Treadmill exercise ECG test for case 1, showing positive test for reversible ischemia

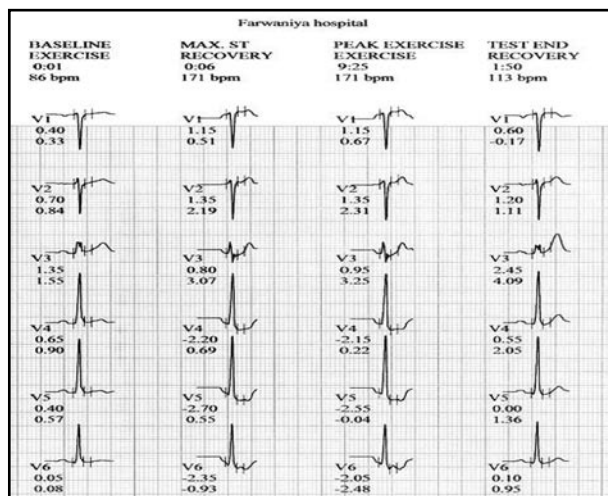


Fig. 3: Treadmill exercise ECG test for case 2, showing positive test for reversible ischemia

noted. Pulsed tissue Doppler imaging revealed regional systolic and diastolic dysfunction at anterior septum and anterolateral segments.

Treadmill exercise ECG test by Bruce protocol revealed positive stress test for myocardial ischemia as there was flat ST-segment depression > 2mm in II, III, aVF, V5, V6 (Fig. 3).

Coronary angiography revealed saccular dilatation of proximal segment of left anterior descending coronary artery up to second septal perforator with luminal diameter = 7 mm and luminal diameter of distal artery = 1.4 mm, saccular dilatation of proximal segment of left circumflex coronary artery up to posterior coronary artery of left ventricle with luminal diameter = 6.8 mm and luminal diameter of distal artery = 1.2 mm. No obstructive lesions in coronary arteries were

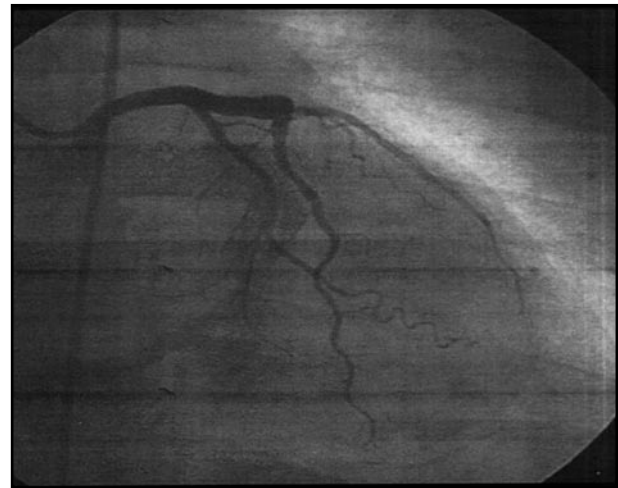


Fig. 2: Coronary angiogram for case 1, showed coronary ectasia at left main, proximal LAD

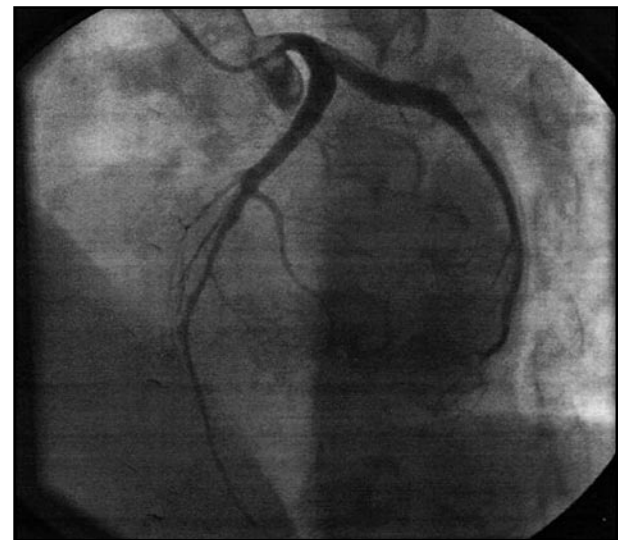


Fig. 4: Coronary angiogram for case 2, showed coronary ectasia at left main, proximal circumflex and proximal LAD

noticed. There was a slow flow in distal coronary artery. The dominant right coronary artery had diffuse dilatation with no coronary collaterals (Fig. 4, 5).

The patient was put on clopidogrel tablets once daily for six months, ASA tablets 150 mg once daily, ACE inhibitor tablets 5 mg once daily, statin tablet once daily 10 mg and metoprolol 50 mg once daily.

DISCUSSION

Since the description by Morgagni of coronary arterial dilatation in a patient with syphilitic aortitis^[3], isolated antemortem cases of ectasia or aneurysms of the coronary arteries have been described as a probable isolated congenital lesion^[4] or in association with congenital heart disease^[5], Ehlers Danlos syndrome^[6], polyarteritis^[7], bacterial infection^[8] and atherosclerosis^[9]. Scott^[10] collected a series of reported cases of aneurysms and he found that the most common type was of congenital



Fig. 5: Coronary angiogram for case 2, showed diffuse coronary ectasia of right coronary artery

origin and that the mycotic-embolic type was next in frequency.

In a consecutive series of 694 autopsies with detailed examination of the coronary arteries, Daoud *et al* in 1963^[11] found in patients over age 16 years, a prevalence rate of 1.4 percent of localized saccular and fusiform atherosclerotic dilatations of the coronary arteries, which he called aneurysms. An aneurysm is a sac that must be distinct from the remainder of the vessel to be appreciated. Markis *et al* in 1976^[12] preferred the word ectasia, since the entire vessel, throughout its course, can be involved and term is descriptive of the anatomic features. The diagnosis and therapeutic implications of this entity with its atherosclerotic origin can then be separated from the diverse group of cineangiographic and postmortem conditions that have previously been collectively termed aneurysms. They described four types of coronary ectasia: diffuse ectasia of two or three vessels was classified as type I, diffuse disease in one vessel and localized disease in another vessel as type II, diffuse ectasia of one vessel only as type III and localized or segmental ectasia as type IV.

Daoud *et al*^[11] described ten cases found at postmortem examination; all patients had severe atherosclerosis. Six patients had a history of hypertension but surprisingly, the cause of death in eight out of the ten was an abdominal aortic aneurysm. Benchimol *et al*^[13] observed only two cases of coronary arterial aneurysm of probable atherosclerotic origin in more than 2000 selective coronary cine angiograms.

Pathologic examination of coronary arteries revealed the typical diffuse hyalinization, lipid deposition, destruction of intima and media, focal calcification and fibrosis, cholesterol crystals, intramural hemorrhage and foreign body giant cell reaction of the atherosclerotic process. Once the process extended to the media, extensive

destruction of the musculoelastic elements was evident, resulting in marked attenuation of the vessel wall. In areas with relatively intact media, the ectasia was absent. The irregular and focal distribution of the process suggests that the ectasia, manifested angiographically as fusiform dilatations, is generated by intraluminal pressures against an elastic vessel wall with decreased stress tolerance^[14].

In 1985, Hartnell *et al*^[2] reported that coronary artery ectasia affects about 2% of the general population, but the aetiology of this coronary enlargement is unknown. One possibility is that there is an imbalance between the beneficial effects of nitric acid (NO) on coronary dilation and the potentially detrimental effects of chronic overstimulation by this endothelium-derived relaxation factor.

Many patients with angina receive chronic glyceryl trinitrate therapy. Yet no one has implicated an increased frequency of ectasia, as one might anticipate since this agent acts *via* NO stimulation. Perhaps the lack of ectasia is related to the mild doses used. Another possibility is that these patients usually have coronary artery disease, and atherosclerosis blunts the ability of the endothelium to produce adequate NO^[15].

The effect of atherosclerosis on NO production was shown by Quyyumi *et al*^[15]. They found that coronary vascular dilation in response to acetylcholine is predominantly caused by increased production of NO and that, despite the absence of angiographic evidence of atherosclerosis, patients with at least one known risk factor for coronary artery disease had reduced resting and stimulated bioavailability of NO from the coronary circulation. A paradoxical vasoconstriction has been seen in atherosclerotic vessels stimulated with acetylcholine. This may be related to the relation between NO-induced vasodilation and endothelin-induced vasoconstriction. When the reduced bioavailable NO is stimulated by acetylcholine, the contrasting action of endothelin becomes dominant and vasoconstriction could result^[16].

Several observations may help us to understand the aetiology of ectasia. England^[17] found an increased frequency or clustering of ectasia in a retrospective review of young men surviving a myocardial infarction in rural Australia. These individuals were farmers who had been exposed to herbicide sprays. Common components of most herbicides include 2, 4-D (dichlorophenoxyacetic acid), 2,4,5-T (trichlorophenoxyacetic acid), or an acetylcholinesterase inhibitor. Extended exposure to these agents might lead to chronically raised concentrations of acetylcholine in the coronary interstitium. A possible mechanism is that 2, 4-D and

2,4,5 -T herbicides (containing acetic acid) increase acetylcholine concentrations through competitive inhibition by directly increasing the end products of acetylcholine breakdown, choline and acetic acid. Herbicides containing acetylcholinesterase inhibitors would directly increase the concentrations of acetylcholine.

Acetylcholine is a potent stimulator of NO^[18]. Therefore, herbicides may be responsible for locally increased NO concentrations. NO stimulates the relaxation of vascular smooth muscle via the guanylate cyclase pathway and release of calcium from the endoplasmic reticulum.

Lam and Ho from Singapore^[19] found the incidence of ectasia was 1.2% and the majority of patients were male in their sixth decade with underlying dyslipidemia or hypertension. They found coronary ectasia was associated with obstructive coronary artery disease in more than 80% of cases.

Akyurek *et al*^[20] from Turkey reported that coronary flow reserve is significantly reduced in patients with diffuse coronary ectasia. Although volumetric coronary blood flow is significantly higher in coronary ectasia, microcirculatory dysfunction that is reflected as depressed coronary flow reserve may be the underlying cause of exercise induced myocardial ischemia. The patients with isolated coronary artery ectasia have raised levels of plasma soluble intracellular adhesion molecule-1, vessel cell adhesion-1 and E-selectin in comparison with patients with obstructive coronary artery disease without coronary ectasia and control group with normal coronary arteries, suggesting the presence of a more severe and extensive chronic inflammation in the coronary circulation in these patients^[21].

Yetkin *et al*^[22] reported that the patients with coronary artery ectasia have an increased prevalence of varicocele compared to those with coronary artery disease and the mechanism underlying coronary artery ectasia might further increase the prevalence of varicocele in susceptible patients. The coronary artery ectasia / aneurysm may lead to exercise induced ischemia, especially in the diffuse form^[23]. Endoh *et al*^[24] reported that coronary ectasia is not benign and must be carefully monitored and coronary atherosclerosis may contribute to the occurrence of subsequent cardiac events.

Akdemir and his colleagues^[25] found that HLA-DR B1*13, DR 16, DQ2 and DQ5 genotypes may be associated with the pathogenesis and increased risk of coronary artery ectasia. The activation of the rennin angiotensin system may lead to an increased inflammatory response in the vessel wall or to an activation of matrix metalloproteinases and an insertion / deletion (ID) polymorphism of ACE has

been associated with development of aneurysm. They also found that an angiotensin converting enzyme DD genotype may be a risk factor for coronary artery ectasia^[26].

Pegel *et al* from Israel in 2002^[27] reported that coronary artery ectasia is associated with increased coronary spasm, dissection and thrombus formation. However, its relative contribution to coronary morbidity remains unclear. The natural evolution of coronary ectasia in the patient who was treated with ligation of aneurysm and distal bypass grafting under cardiopulmonary bypass revealed that a fragile fresh clot was formed within the aneurysm irrespective of coumadin therapy as a standard regimen for the coronary artery ectasia^[28].

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