İstanbul Med J 2019; 20(5): 394–7
DOI: 10.4274/imj.galenos.2019.27928

Prevalence and Clinical Importance of Coronary Artery Ectasia: Tertiary Center Experience

Ektazik Koroner Arter Prevalansı ve Klinik Önemi: Tersiyer Merkez Deneyimi

Ahmet Gürdal, Serhat Sığırcı, Kadriye Orta Kılıçkesmez
İstanbul Şişli Hamidiye Etfal Training and Research Hospital, Clinic of Cardiology, İstanbul, Turkey

ABSTRACT

Introduction: The incidence, prognostic significance, clinical features and risk factors of coronary artery ectasia (CAE) in patients undergoing coronary angiography were investigated.

Methods: A total of 10.320 coronary angiographies performed between June 2015 and August 2018 in our clinic were analyzed retrospectively.CAE, defined as the enlargement of a coronary artery to 1.5 times or more than that of the local or commonly adjacent normal coronary artery segment and classified according to Markis classification. CAE was divided into two groups as mild and severe according to the degree of ectasia.

Results: Of 189 patients (1.8%) with coronary ectasia, 143 were male (76%). One hundred and seven patients (57%) were smokers, 96 (51%) were dyslipidemic, 106 (56%) had hypertension and 43 (22%) had diabetes mellitus. In 92%, the symptoms were chest pain and dyspnea. Of 102 patients (54%) presenting with acute coronary syndrome, 43 (23%) were diagnosed as ST-segment elevation myocardial infarction, while 59 (31%) were diagnosed as non-ST segment elevation myocardial infarction or unstable angina pectoris. The right coronary artery was the most affected vessel from ectasia (54.5%, n=103). Type 3 ectasia was the most common (34.9%, n=66), while type 2 ectasia was the least common (10.6%). Hundred and twenty-nine (68.3%) of the patients had mild ectasia, while 60 (31.7%) had severe ectasia. During the three-year follow-up, a total of 16 (8.4%) patients died, three of which were in-hospital and eight were cardiac-related.

Conclusion: Although it is not common in the community, it is important to detect CAE, which is an important cause of mortality and morbidity, and to follow up the patients closely.

Keywords: Coronary artery ectasia, coronary angiography, coronary artery disease

ÖZ

Amaç: Koroner anjiyografi yapılan hastalardaki koroner arter ektazisi (KAE) sıklığı, prognostik önemi, klinik özellikleri ve risk faktörleri araştırıldı.

Yöntemler: Kliniğimizde Haziran 2015-Ağustos 2018 tarihleri arasında yapılan 10.320 adet koroner anjiyografi retrospektif olarak analiz edildi. KAE, koroner arterin bölgesel ya da yaygın olarak komşu normal koroner arter segmentinin 1,5 katı veya daha fazlası olacak şekilde genişlemesi olarak tanımlandı ve Markis sınıflamasına göre sınıflandırıldı. Ektazik koroner arterler ektazi derecesine göre hafif ve ağır olmak üzere iki gruba ayrıldı.

Bulgular: Yüz seksen dokuz (%1,8) koroner ektaziye sahip hastanın 143'ü erkek (%76) idi. Hastaların 107'si (%57) sigara içerken, 96'sı (%51) dislipidemik, 106'sı (%56) hipertansiyon, 43'ü (%22) diabetes mellitus tanılıydı. Hastaların %92'sinde semptom göğüs ağrısı ve nefes darlığıydı. Akut koroner sendrom ile başvuran 102 (%54) hastanın 43'ü (%23) ST-segment elevasyonlu miyokart enfarktüsü iken, 59'u (%31) ST-segment elevasyonu olmayan miyokart enfarktüsü ya da unstabil angına pektoris tanılıydı. Sağ koroner arter ektaziden en çok etkilenen damardı (%54,5 n=103). En fazla tip 3 ektazi (%34,9, n=66) görülürken, en az tip 2 ektazi (%10,6, n=20) görüldü. Hastaların 129'unda (%68,3) hafif düzeyde ektazi izlenirken, 60'ında (%31,7) ağır düzeyde ektazi saptandı. Üç yıllık takip süresince 3'ü hastane içi, 8'i kardiyak nedenli olmak üzere toplam 16 (%8,4) hastada ölüm gerçekleşti.

Sonuç: Toplumda sık görülmese de önemli bir mortalite ve morbidite nedeni olan KAE hastalığının tespiti ve hastaların yakın takibi önem arz etmektedir.

Anahtar Kelimeler: Koroner arter ektazisi, koroner anjiyografi, koroner arter hastalığı

Address for Correspondence/Yazışma Adresi: Ahmet Gürdal MD, İstanbul Şişli Hamidiye Etfal Training and Research Hospital, Clinic of Cardiology, İstanbul, Turkey
Phone: +90 506 478 14 41 **E-mail:** gurdal27@hotmail.com **ORCID ID:** orcid.org/0000 0002 2168 4937

Cite this article as/Atıf: Gürdal A, Sığırcı S, Orta Kılıçkesmez K. Prevalence and Clinical Importance of Coronary Artery Ectasia: Tertiary Center Experience. İstanbul Med J 2019; 20(5): 394-7.

Received/Geliş Tarihi: 18.05.2019
Accepted/Kabul Tarihi: 03.07.2019

Introduction

Coronary artery ectasia (CAE) is defined as the enlargement of a coronary artery to 1.5 times or more than that of the local or commonly adjacent normal coronary artery segment (1). Enlargements up to 1.5 times of the coronary artery are described as ectasia, whereas enlargements over two-fold are called aneurysms (2). CAE is seen in the population with a frequency ranging between 0.3-4.9% (3). CAE is frequently associated with coronary artery disease (CAD), but can also be isolated. Isolated CAE is rare among patients who underwent coronary angiography with a frequency of 0.1-0.79% (4,5).

CAE may be congenital or acquired. Most of the etiology includes atherosclerosis (50%) (6), congenital syndromes such as Kawasaki syndrome (20-30%) (7), Ehlers-Danlos syndrome (8), Marfan syndrome and Takayasu disease (9), and connective tissue diseases (10-20%) such as inflammatory or scleroderma, polyarteritis nodosa, systemic lupus erythematosus (10). Studies have shown that ectasia leads to slow flow in the coronary arteries, thrombus formation and vasospasm, and perfusion defects in myocardium may be observed due to slow flow and possible microembolies (11,12). CAE is mostly associated with CAD. The most common symptom is chest pain. Although there is no significant stenosis in the coronary arteries, acute coronary syndrome (ACS) may occur. This has been shown to be caused by dissection and thrombus in the ectasia region (13). Recurrent microembolies due to thrombus can cause impaired coronary perfusion, ventricular arrhythmias and sudden cardiac death (5).

In this study, the incidence of CAE, distribution according to coronary arteries, clinical characteristics and risk factors, and prognostic significance of CAE in patients undergoing coronary angiography in our clinic were investigated.

Methods

A total of 10.320 coronary angiographies performed between June 2015 and August 2018 in our clinic were analyzed retrospectively. CAE was defined as dilatation of coronary artery lumen exceeding the largest diameter of an adjacent normal vessel more than 1.5 fold in accordance with the angiographic description of Hartnell et al. (1). Regional or diffuse enlargement without significant coronary artery stenosis was accepted as isolated ectasia. The presence of more than 50% stenosis of the coronary artery was considered to be significant occlusion and CAD. The study was approved by the Ethics Committee of İstanbul Şişli Hamidiye Etfal Training and Research Hospital, (decision no: 2380, date: 30.04.2019). Because of the retrospective nature of the study, informed consent was not obtained from the patients.

CAE was classified according to the classification of Markis et al. (7). According to this classification, diffuse ectasia of two or three vessels was type 1, diffuse ectasia in one vessel and localized disease in another was type 2, diffuse ectasia in one vessel only was type 3, and localized and segmental involvement was type 4. CAE was divided into two groups according to the degree of ectasia. According to the classification of Markis et al. (7), type 1 and type 2 were defined as severe ectasia and type 3 and type 4 as mild ectasia.

Statistical Analysis

SPSS version 20 for Windows (SPSS, Inc. Chicago, IL, USA) was used to evaluate the data. Continuous variables were expressed as mean ± standard deviation, and categorical variables were expressed as percentage.

Results

A total of 10.320 coronary angiography images over 3 years were analyzed and 189 (1.8%) patients with CAE were identified. Of the patients with ectasia, 143 (76%) were male. While 107 (57%) of the patients were smokers, 106 (56%) were diagnosed as hypertension and 43 (22%) were diagnosed as diabetes mellitus (DM). The number of dyslipidemic patients was 96 (51%). Ninety-two percent of the patients presented with chest pain and dyspnea, while the others presented with atypical complaints (Table 1).

Of 102 (54%) patients presenting with ACS, 43 (23%) had ST-segment elevation myocardial infarction, while 59 (31%) had non-ST-segment elevation myocardial infarction or unstable angina pectoris (Table 1). Regarding patients presenting with ACS, 23 patients had CAD without significant stenosis, 35 had single vessel disease, 23 had two-vessel disease, and 21 patients had three-vessel disease. Sixty-eight patients (36%) had isolated ectasia without significant coronary stenosis.

Ectasia was seen in 106 patients (56.1%) in one coronary artery, in 44 (23.3%) in two coronary arteries, in 30 (15.9%) in three coronary arteries and in nine (4.8%) in four vessels including the main coronary artery. The right coronary artery (RCA) was the vessel most affected by ectasia (54.5%, n=103). Of the ectasia, 54% (n=102) were in the circumflex (Cx) artery, 42.9% (n=81) in the left anterior descending (LAD) artery, and 16.9% (n=32) in the left main coronary artery (LMCA) (Table 2).

Table 1. Basal clinical and laboratory characteristics of patients

Clinical features	Ectasia (n=189)
Age (years)	61±12
Male gender	143 (76%)
Smoking	107 (57%)
Hypertension	106 (56%)
Diabetes mellitus	43 (22%)
EF (%)	52±10
Acute coronary syndrome	102(54%)
STEMI	43(23%)
NSTEMI/USAP	59(31%)
Hemoglobin (g/dL)	13.8±2.1
Platelet (10^9/L)	228±59
Total cholesterol (mg/)	193±57
HDL-k (mg/dL)	40±9
LDL-k (mg/dL)	123±54
Triglyceride (mg/dL)	161±96
Creatinine (mg/dL)	1.1±0.9

EF: ejection fraction, STEMI: ST-segment elevation myocardial infarction, NSTEMI: non-ST segment elevation myocardial infarction, USAP: unstable angina pectoris, HDL: high-density lipoprotein, LDL: low-density lipoprotein

Table 2. Characteristics of ectatic coronary arteries in patients

	n	%
Isolated ectasia	68	36
Classification of Markis et al. (7)		
Type 1	40	21.2
Type 2	20	10.6
Type 3	66	34.9
Type 4	63	33.3
Ectasia location		
LMCA (Left main coronary artery)	32	16.9
LAD (Left anterior descending artery)	81	42.9
Cx (Circumflex artery)	102	54
RCA (Right coronary artery)	103	54.5
Degree of ectasia		
Severe (Type 1+Type 2)	60	31.7
Mild (Type 3+Type 4)	129	68.3

LMCA: left main coronary artery, LAD: left anterior descending, Cx: circumflex, RCA: right coronary artery

Regarding Markis et al. (7) classification, the most common ectasia was type 3 (34.9%, n=66), followed by type 4 (33.3%, n=63), type 1 (21.2%, n=40) and type 2 (10.6%, n=20). While 129 patients (68.3%) had mild ectasia, 60 patients (31.7%) had severe ectasia (Table 2). During the three-year follow-up period, a total of 16 patients (8.4%) died, including three in-hospital deaths. Eight out of hospital deaths were cardiac-related deaths.

Discussion

The incidence of CAE detected in coronary angiography performed on suspicion of CAD varies between 0.3-4.9% (3). While this rate was found to be 4.2% in the study conducted by Yilmaz et al. (14), it was found to be 2.8% in the study performed by Sultana et al. (15). The prevalence of CAE was found to be 1.8% in our study. The prevalence of isolated CAE was found to be 1.08% in a series of 3.815 patients by Akyürek et al. (16). In another series, this rate was reported as 3% (9), and Hartnell et al. (1) reported the prevalence of isolated CAE as 17%. In our study, the prevalence of isolated CAE without significant coronary artery stenosis was 36%.

In addition to the studies that reported similar rates of CAE in females and males (17), it was found to be more frequent in males in some studies (1). In our study, 76% of CAE patients were male. In a study, CAD was reported to be 87.1% in patients with ectasia (18). In our study, this rate was 64%. It is known that CAE is most commonly seen in RCA and at least in LMCA (19). In a study based on registry data, the most common involvement was observed in RCA (20). Sultana et al. (15) reported that ectasia was most common in RCA, followed by Cx and LAD, respectively. Similarly, in our patient group, ectasia was most commonly seen in RCA (54.5%), followed by Cx, LAD and LMCA, respectively.

In the study of Demopoulos et al. (17), type 3 ectasia was the most common, while others were found with similar frequency. Yilmaz et al. (14) reported that type 4 ectasia was the most common, whereas type 1 ectasia was the least common. In addition, Markis et al. (7) reported that the most common type was type 1, followed by type 2, 3 and 4, respectively. In our study, the most common type was type 3 with a rate of 34.9%, followed by type 4 (33.3%), type 1 (21.2%) and type 2 (10.6%).

Markis et al. (7) found that hypertension was more common in patients with ectasia and suggested that hypertension could play a role in the pathogenesis of CAE by accelerating the destruction of the media layer. Sultana et al. (15) found hypertension in 55%, DM in 26% and dyslipidemia in 58% of the patients. Similarly, in our study, hypertension was found in 56%, DM in 22%, and dyslipidemia in 51%.

Atherosclerosis plays a major role in the etiology of CAE. Cholesterol crystals, calcification and fibrosis, intima and media destruction, lipid accumulation were detected in pathological examination of CAE and these histological changes were found to be the same as atherosclerotic process. Ectasia occurs as a result of the atherosclerotic process causing widespread destruction of the muscular structure in the media layer and thinning of the vessel wall (1). In our study, obstructive CAD was detected in 64% of the patients and 54% of the patients presented with ACS.

Although the mechanism of ischemia is not clear in patients with isolated CAE, it has been suggested that slow flow in the ectatic vessel, thrombus caused by slow flow and microembolies discharging into the distal coronary bed disrupt perfusion (21). In a study conducted in our country, it was shown that microvascular perfusion is impaired in patients with ectatic coronary disease (22). In another study, it was found that flow velocity in ectatic coronary arteries decreased (23). In our study, 92% of the patients presented with chest pain and dyspnea, while 22 (12%) of the patients who presented with ACS did not have obstructive CAD.

The prognosis of patients with CAE is controversial. While the annual mortality was found to be 15% in the study of Markis et al. (7), Hartnell et al. (1) found annual mortality rate as 4.6% in medical follow-up and 2.4% in surgical groups. In another study, the annual mortality rate was found to be 1.5% in medical follow-up, 2.1% in percutaneous coronary intervention, and 2.9% in coronary bypass surgery (14). In our study, a total of 16 patients (8.4%) died, including three in-hospital deaths, during the three-year follow-up period, while the annual mortality rate was similar to that of many studies.

Conclusion

There is no consensus on the treatment of CAE, which is an important cause of mortality and morbidity, although it is not common in the community. Even if they do not have obstructive CAD, close monitoring of patients with CAE is important. Large-scale prospective studies are needed to determine the choice of treatment and better prognostic evaluation.

Ethics Committee Approval: The study was approved by the Ethics Committee of İstanbul Şişli Hamidiye Etfal Training and Research Hospital, (decision no: 2380, date: 30.04.2019).

Informed Consent: informed consent was not obtained from the patients.

Peer-review: Internally peer-reviewed.

Author Contributions: Surgical and Medical Practices - A.G., S.S., K.O.K.; Concept - A.G.; Design - A.G., S.S.; Data Collection and/or Processing A.G., S.S.; Analysis and/or Interpretation - A.G., K.O.K.; Literature Search - S.S.; Writing Manuscript - A.G.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study received no financial support.

References

1. Hartnell GG, Parnell BM, Pridie RB. Coronary artery ectasia. Its prevalence and clinical significance in 4993 patients. Br Heart J 1985; 54: 392-5.
2. Befeler B, Aranda MJ, Embi A, Mullin FL, El-Sherif N, Lazzara R. Coronary artery aneurysms: Study of the etiology, clinical course and effect on left ventricular function and prognosis. Am J Med 1977; 62: 597-607.
3. Oliveros RA, Falsetti HL, Carroll RJ, Heinle RA, Ryan GF. Atherosclerotic coronary artery aneurysm. Report of five cases and review of literature. Arch Intern Med 1974; 134: 1072-6.
4. Swaye PS, Fisher LD, Litwin P, Vignola PA, Judkins MP, Kemp HG, et al. Aneurysmal coronary artery disease. Circulation 1983; 67: 134-8.
5. Al-Harthi SS, Nouh MS, Arafa M, al-Nozha M. Aneurysmal dilatation of the coronary arteries: diagnostic patterns and clinical significance. Int J Cardiol 1991; 30: 191-4.
6. Swanton RH, Thomas ML, Coltart DJ, Jenkins BS, Webb-Peploe MM, Williams BT. Coronary artery ectasia-a variant of occlusive coronary arteriosclerosis. Br Heart J 1978; 40: 393-400.
7. Markis JE, Joffe CD, Cohn PF, Feen DJ, Herman MV, Gorlin R. Clinical significance of coronary arterial ectasia. Am J Cardiol 1976; 37: 217-22.
8. Dieter RS, Murtaugh T, Black J, Russell DC. Coronary arteriomegaly in a patient with Ehlers-Danlos syndrome and multiple aneurysms-a case report. Angiology 2003; 54: 733-6.
9. Altinbas A, Nazli C, Kinay O, Ergene O, Gedikli O, Ozaydin M, et al. Predictors of exercise induced myocardial ischemia in patients with isolated coronary artery ectasia. Int J Cardiovasc Imaging 2004; 20: 3-17.
10. Chaithiraphan S, Goldberg E, O'Reilly M, Jootar P. Multiple aneurysms of coronary artery in sclerodermal heart disease. Angiology 1973; 24: 86-93.
11. Papadakis MC, Manginas A, Cotileas P, Demopoulos V, Voudris V, Pavlides G, et al. Documentation of slow coronary flow by the TIMI frame count in patients with coronary ectasia. Am J Cardiol 2001; 88: 1030-2.
12. Kruger D, Stierle U, Herrmann G, Simon R, Sheikhzadeh A. Exercise-induced myocardial ischemia in isolated coronary artery ectasias and aneurysms (dilated coronopathy). J Am Coll Cardiol 1999; 34: 1461-70.
13. Mattern AL, Baker WP, McHale JJ, Lee DE. Congenital coronary aneurysms with angina pectoris and myocardial infarction treated with saphenous vein bypass graft. Am J Cardiol. 1972; 30: 906-9.
14. Yilmaz H, Sayar N, Yilmaz M, Tangürek B, Cakmak N, Gürkan U, et al. Coronary artery ectasia: Clinical and angiographical evaluation. Turk Kardiyol Dern Ars. 2008; 36: 530-5.
15. Sultana R, Sultana N, Ishaq M, Samad A. The prevalence and clinical profile of angiographic coronary ectasia. J Pak Med Assoc. 2011; 61: 372-5.
16. Akyürek Ö, Berkalp B, Sayın T, Dinçer İ, Kervancıoğlu C, Oral D. İzole koroner arter ektazisinde azalmış koroner arter rezervi. MN Kardiyoloji Dergisi 2001; 8: 161-7.
17. Demopoulos VP, Olympios CD, Fakiolas CN, Pissimissis EG, Economides NM, Adamopoulou E, et al. The natural history of aneurysmal coronary artery disease. Heart 1997; 78: 136-41.
18. Giannoglou GD, Antoniadis AP, Chatzizisis YS, Damvopoulou E, Parcharidis GE, Louridas GE. Prevalence of ectasia in human coronary arteries in patients in northern Greece referred for coronary angiography. Am J Cardiol 2006; 98: 314-8.
19. Syed M, Lesch M. Coronary artery aneurysm: A review. Prog Cardiovasc Dis 1997; 40: 77-84.
20. Swaye PS, Fisher LD, Litwin P, Vignola PA, Judkins MP, Kemp HG, et al. Aneurysmal coronary artery disease. Circulation 1983; 67: 134-8.
21. Falsetti HL, Carrol RJ. Coronary artery aneurysm. A review of the literature with a report of 11 new cases. Chest 1976; 69: 630-6.
22. Gulec S, Atmaca Y, Kilickap M, Akyurek O, Aras O, Oral D. Angiographic assessment of myocardial perfusion in patients with isolated coronary artery ectasia. Am J Cardiol 2003; 91: 996-9.
23. Hamaoka K, Onouchi Z, Kamiya Y, Sakata K. Evaluation of coronary flow velocity dynamics and flow reserve in patients with Kawasaki disease by means of a Doppler guide wire. J Am Coll Cardiol 1998; 31: 833-40.