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Coronary Artery Ectasia- a review

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ABSTRACT

Coronary artery ectasia (CAE) is an extreme type of expansive vascular remodelling (i.e. excessive expansive remodelling) as a result of atherosclerotic plaque formation; atherosclerosis being the most prevalent cause. Despite being identified more than 50 years ago, the clinical relevance of CAE is not well-defined, and its therapy is still a matter of discussion. In this review paper, we have highlighted the aetiology, pathophysiology, and currently recommanded treatment of coronary artery ectasia based on all accessible literature from PubMed and other international journals.

Keywords: *Coronary artery ectasia, aneurismal coronary artery disease*

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INTRODUCTION

Coronary artery ectasia (CAE) is a rare form of coronary artery disease, atherosclerosis being the most common cause. Coronary angiography shows that 3-8% of patients have CAE, either alone or in combination with stenotic lesions.[1] Regardless of stenotic lesions, the presence of ectatic segments

leads to slow blood flow and can increase exercise-induced angina and myocardial infarction. Anti-platelet medications are the mainstay of therapy for isolated CAE, which has a better prognosis.

Most people with the condition have no symptoms. Males are 4 times more likely to develop coronary artery ectasia than females, and persons with risk factors for heart disease, such as smoking, are also more likely to develop this condition.[2,3] Although it is more frequent in those with atherosclerosis and coronary artery disease, the condition can also occur on its own.

AETIOLOGY AND EPIDEMIOLOGY

When a portion of a coronary artery swells at least 1.5 times the neighboring normal artery, this is called as coronary artery ectasia (CAE) or aneurysmal coronary artery disease. CAE occurs in 3%-8% of angiographic studies and 0.22%-1.4% of autopsy data.[2,4] It may be limited to a specific area in the coronary artery or can spread across the entire artery. In CAE, one or more of the coronary arteries may be affected.

More than 50% of all cases are caused by atherosclerosis, whereas 20%-30% are thought to be congenital. In most of these people, ectasia appears along with coronary artery disease (CAD). However, only around 10% to 20% of CAE have been linked to inflammatory or connective tissue disorders.[2] Some people with connective tissue illnesses with an exacerbated inflammatory response, such as Marfan syndrome and Kawasaki disease, develop coronary artery ectasia.[5] Connective tissue illnesses including scleroderma, Ehlers-Danlos syndrome, various forms of ANCA-related vasculitis, and syphilitic aortitis are also linked to coronary dilatation.[1]

In a study of 8812 patients in Turkey, CAE was discovered in 122 (1.38%) individuals, of whom 72 (59%) had concurrent severe CAD, which was defined as > 70% diameter stenosis of major coronary arteries or 40% stenosis of the left main stem. Twenty-nine patients (23.7%) had isolated CAE, while twenty-one patients (17.2%) had non-significant CAD. In individuals with CAE, smoking, hypertension, and dyslipidemia were more prevalent.[6]

The prevalence of CAE varies between 1.2–6% in different literature.[7,8,9,12] Coexisting stenotic CAD was found in 90.8% of the CASS (coronary artery surgery study) registry[7] and 84.7% in a series of patients studied by Demopoulos *et al*.[8] In the study of Pinar *et al*,[10] CAE was found to be associated with the classic coronary risk factors like male sex and smoking except for diabetes, a pathology that was less frequent than usual. CAE was found more in younger patients. Demopoulos *et al*,[8] however, found no specific predisposing factor. The prevalence of systemic hypertension was higher in some studies.[7] Sudhir *et al* found an increased prevalence of ectasia in familial hypercholesterolaemia and showed a strong inverse association with HDL cholesterol, but ectasia was not related to age, hypertension, smoking or ethnicity.[11] Age seems to have no additional influence according to most investigators.[7-11] But in all studies, CAE was found commonly in males. Dyslipidaemia, hypertension and smoking were significant predisposing factors.

In some studies, the RCA was reported to be the most commonly involved vessel, whereas others reported the LAD to be the main involved vessel.[7-11] Significant CAD is associated with a higher incidence of major adverse coronary events in individuals with CAE than in persons without CAD. In individuals with CAE without CAD, recurrent bouts of chest discomfort without concomitant dynamic ECG alterations or enzyme elevations were prevalent. Patients with CAE are at increased risk for angina, but the risk of myocardial infarction and cardiac related mortality is mostly determined by the presence of obstructive CAD.[6]

Angiographic follow up of CAE patients in one study revealed, coronary ectasia was more frequent in males (88%). The frequency of involvement was: the right coronary (47%), the left circumflex (30%), the left anterior descending (21%) and the left main arteries (2%). Proximal segments were most frequently involved (48%). Diffuse involvement was found in 29% of patients. The severity of ectasia progressed in 14% and 2 new ectatic segments appeared over the follow-up period of two to eight years. During that period, 2 patients had myocardial infarction, 1 of them due to a total occlusion of an ectatic segment. There were no deaths. In conclusion, coronary ectasia has a relatively benign course.[9]

CAE should be distinguished from coronary aneurysms after coronary procedures. On occasion, massive coronary plaques with ulcers might be misdiagnosed angiographically as coronary aneurysms. Intravascular ultrasonography can uncover the underlying reason (IVUS). There is no association between ectasy at the level of the coronary arteries and ectasy in other arteries of the peripheral vascular system, although they may coexist in some instances.

Atherosclerosis in the coronary arteries may be a contributing factor in the development of later cardiac events.[13]

PATHOPHYSIOLOGY

The condition known as coronary artery ectasia is distinguished by a rise in the wall stress of the vessel as well as a weakening of the arterial wall, both of which lead to a gradual dilatation and remodeling of the vessel. It is believed that inflammation, which may have been brought on by illness, chemicals, or physical stress on the vessel, is the primary factor responsible for the artery's persistent dilation.[14,15] The inflammatory reaction leads to an increased production of matrix metalloproteinases, cysteine proteinases, and serine proteinases, which causes the artery to partially break down and weakens it as a result.[15,16] Platelets get activated when there is an inflammatory reaction, which raises the likelihood of blood clots forming. The turbulent blood flow of the larger artery, which can activate platelets and lead to the formation of blood clots, is another factor that contributes to the increased risk of developing blood clots. 17 In coronary artery ectasia, inflammation contributes to elevated levels of oxidative stress, while at the same time antioxidant activity is reduced. This imbalance has the potential to cause harm to the cells, which will further weaken the vessels.[18] The activation of the inflammatory response results in a large rise in the production of proinflammatory cytokines such as C reactive protein, interleukin-6, tumor

necrosis factor alpha, and cell adhesion molecules, all of which can be utilized as diagnostic markers.[19,20]

The presence of aneurismal segments produces sluggish or turbulent blood flow, with an increased incidence of typical exercise-induced angina pectoris and myocardial infarction, regardless of the severity of coexisting stenotic lesions. Patients with pure ectasia (15% of the total population with CAE) have a more benign course, but 39% still present signs of previous myocardial infarction.[2]

This process of "arterial remodelling" is fundamental to the pathophysiology of CAD. The in vivo experience with IVUS has confirmed that both arterial expansion and shrinkage can be a manifestation of coronary atherosclerosis. Positive remodelling (arterial expansion) is frequently associated with unstable coronary syndromes, whereas negative remodelling (arterial shrinkage) is associated with stable coronary syndromes.

DIAGNOSIS

Coronary angiography is the most common diagnostic method for identifying coronary artery ectasia. Angiography frequently reveals CAE in undiagnosed IHD patients with symptoms.

IVUS is a great method for measuring luminal diameter and identifying artery wall alterations. The majority of CAE patients have a considerable atheromatous load, with plaque regions uniformly distributed across proximal and distal reference segments, as well as within the aneurysmal segment. As a result of the greater vessel size, the percentage of stenosis is much lower within the CAE, demonstrating the challenge of evaluating stenosis severity when it manifests within an ectatic section. True aneurysms produced by plaque rupture can be distinguished by IVUS from fake aneurysms. Angiographic findings of empty plaque cavities mistaken for CAE are clinically relevant because fake aneurysms can cause acute coronary syndromes.

CAE is characterized by slow blood flow filling and delayed washout, both of which are directly proportional to the degree of ectasia. Coronary blood flow decreases with increasing ectasia size.[28] Angiographic signs of turbulent and stagnant flow include delayed antegrade dye filling, a segmental backflow phenomenon, and local deposition of dye in the dilated coronary segment.

Akyurek et al. measured blood flow velocity and coronary flow reserve using a Doppler wire in patients with isolated CAE and healthy controls. The CAE group tended to have a lower resting blood flow velocity than the control group.[21]

Due to the necessity of frequent angiograms, monitoring ectatic veins is challenging. The great majority of the proximal and middle segments of the left main, left anterior descending, and right coronary arteries may be seen with three-dimensional, non-contrast-enhanced, free-breathing coronary magnetic resonance angiography (MRA). Coronary MRA has previously demonstrated clinical use for the evaluation of anomalous coronary artery disease, and in certain instances it is superior to X-ray coronary angiography in defining the course of an anomalous arterial.

Coronary MRA is equivalent to quantitative coronary angiography, with the added benefit of being noninvasive. In contrast to CT,

MRA does not need exposure to ionizing radiation or the injection of a contrast agent. When combined with coronary flow data, coronary MRA may provide further useful information on the risk of thrombotic blockage of the aneurysmal arteries. In addition, the MRA, which is non-invasive and non-radioactive, may be conveniently utilized for the efficient monitoring of these patients.

Coronary artery computed tomography (CACT) has been utilized in ectatic vessel assessment. Contrast attenuation measures using CT coronary angiography are well correlated with flow abnormalities measured with conventional X-ray coronary angiography.[22] However, CACT cannot be suggested as a technique of choice for the follow up of patients because of its high radiation dose. Further improvements in terms of radiation dose are awaited with interest in the near future.

THERAPEUTIC APPROACH

The medical care of CAE patients is not yet well-defined. Based on the considerable flow disruptions inside the ectatic segments, persistent anticoagulation has been indicated as the primary treatment in previous investigations. However, this therapy has not been prospectively evaluated and cannot be advised without more research. In sporadic cases of acute thrombotic occlusions, heparin infusion and fibrinolysis have been utilized effectively for re-canalization, occasionally demonstrating the lack of flow-limiting stenoses.[23] When CAE coexists with CAD, the prognosis and therapy are identical to those of CAD alone. In isolated CAE, the prognosis is more favorable, and anti-platelet medications are the basis of treatment. The mechanism behind CAE must be clarified utilizing more clinical, histological, and pathophysiological research in order to improve patient care. In fact, every patient with CAE should be routinely assessed for pathological alterations in other vascular areas, in both the arterial and venous systems, that may emerge as a result of the illness.[24]

Obstructive coronary lesions co-exist in the majority of patients with CAE and the observed incidence of myocardial infarction, even in patients with isolated coronary ectasia, suggested the generalized administration of aspirin in these patients. The role of combined antiplatelet therapy, with the addition of adenosine diphosphate inhibitors, has not yet been evaluated in prospective randomized studies. However, Yasar et al have recently reported that patients with isolated CAE have elevated plasma levels of P-selectin, beta-thromboglobulin and platelet factor 4 compared with control participants who have angiographically normal coronary arteries, suggesting increased platelet activation in patients with CAE.[24] Medications with vasodilating properties against coronary spasm have also been proposed. It is of interest that nitrates, by causing further coronary epicardial dilation, have been shown to exacerbate myocardial ischemia and are discouraged in patients with isolated CAE. At present, there are no vasoactive medications that have already been tested and can be widely recommended to patients with CAE. As CAE represents a form of atherosclerotic heart disease, intense risk factor modification for primary and secondary prevention is obviously necessary.

CAE is more frequent among patients with familial hypercholesterolemia than in a control group, suggesting a link between abnormal lipoprotein metabolism and aneurysmal coronary artery disease.[11] For patients with coexisting obstructive lesions and symptoms or signs of significant ischemia despite medical therapy, percutaneous and/or surgical coronary revascularization can safely and effectively restore normal myocardial perfusion.[25] Special attention should be paid to the need for adequate stent expansion and wall stabilization in these vessels. The implantation of covered versus bare metal stents offers a superior acute angiographic result, excluding the ectatic segment, but the long term benefit has not been adequately proven.[26]

Significant coronary artery disease accompanied by ectatic coronary segments has been treated by coronary artery bypass grafting. Several surgical treatments, including proximal and distal ligation and aneurysm removal, have been developed in response to the presence of thrombus inside the CAE and the question of the need to remove big aneurysms.[27]

CONCLUSION

CAE is a less prevalent type of coronary artery disease that tends to impact male patients more than female patients and affects people with diabetes less frequently. The most common cause is atherosclerosis, although it can also occur in conjunction with diseases of the connective tissue or in a congenital form. The diagnostic procedures and therapy for this condition are quite similar to those used for atherosclerotic stenotic coronary artery disease. However, it is interesting to note that nitrates are not recommended for patients with CAE and that antiplatelet medications are the primary therapy. In patients with CAE, recurrent ischemic chest pain is a symptom; nevertheless, the risk of myocardial infarction and mortality is lower, and the prognosis is better than in patients with stenotic CAD.

REFERENCES

1. *Sophie Mavrogeni. Coronary Artery Ectasia: From Diagnosis to Treatment. Hellenic J Cardiol 2010; 51: 158-163*
2. *Hartnell, G.G., Parnell, B.M. and Pridie, R.B. Coronary artery ectasia: its prevalence and clinical significance in 4993 patients. Br Heart J 1985; 54: 392-405*
3. *Li, J., Nie, S., Qian, X., Zeng, H., and Zhang, C. (2009) Chronic inflammatory status in patients with coronary artery ectasia. Cytokine2009; 46: 61-64*
4. *Markis JE, Joffe CD, Cohn PF, Feen DJ, Herman MV, Gorlin R. Clinical significance of coronary arterial ectasia. Am J Cardiol. 1976; 37: 217-222.*
5. *Sayn, T., Doven, O., Berkalp, B., Akyurek, O., Gulec, S. and Oral, D. Exercise-induced myocardial ischemia in patients with coronary artery ectasia without obstructive coronary artery disease. Inter J Cardio, 2001.78,143-149*
6. *Y Gunes, B Boztosun, A Yildiz et al. Clinical profile and outcome of coronary artery ectasia. Heart. 2006 Aug; 92(8): 1159–1160.*
7. *Swaye P S, Fisher L D, Litwin P et al. Aneurysmal coronary artery disease. Circulation, 1983;67:134–138.*
8. *Demopoulos V P, Olympios C D, Fakiolas C N et al. The natural history of aneurysmal coronary artery disease. Heart 1997;78:136–141.*
9. *Ilia R, Kafri C, Carmel S et al. Angiographic follow-up of coronary*

artery ectasia. Cardiology1995;86(5):388-90.

10. *Pinar B E, Lopez P R, Lozano M I et al. Coronary ectasia: prevalence, and clinical and angiographic characteristics. Rev Esp Cardiol 2003;56:473–479.*
11. *Sudhir K, Ports T, Amidon T et al. Increased prevalence of coronary ectasia in heterozygous familial hypercholesterolemia. Circulation, 1995; 91:1375–1380.*
12. *Devabhaktuni S, Mercedes A, Diep J et al. Coronary Artery Ectasia-A Review of Current Literature. Curr Cardiol Rev. 2016;12(4):318-323.*
13. *Endoh S, Andoh H, Sonoyama K et al. Clinical features of coronary artery ectasia. J Cardiol, 2004;43(2):45-52.*
14. *Antoniadia, A.P., Chatzizisis, Y.S., and Giamoglou, G.P. Pathogenic mechanisms of coronary ectasia. Int J Cardio,2008; 130: 335-343*
15. *Theodoros, A., Korovesis, S., Giazitzoglou, E., et al. Clinical and angiographic characteristics of patients with coronary artery ectasia. Int J Cardio, 2013; 167: 1536-1541*
16. *Varol, E., Uysal, B, A., Dogan, A., Ozaydin, M. and Erodogan, D. Mean platelet volume as a prognostic value in patients with coronary artery ectasia. Clin Appl Thromb-Hema,2012; 18(4): 387-392*
17. *Sezen, Y., Bas, M., Polat, M., Yildiz, A., et al. The relationship between oxidative stress and coronary artery ectasia. Cardio J, 2010; 17(5): 488-494*
18. *Li, J., Nie, S., Qian, X., Zeng, H., and Zhang, C. Chronic inflammatory status in patients with coronary artery ectasia. Cytokine, 2009; 46: 61-64*
19. *Aydn, M., Texn, I.O., Dogan, S.M., et al. The levels of tumor necrosis factor-alpha and interleukin-6 in patients with isolated coronary artery ectasia. Mediat Inflamm. 2009; Available at: http://dx.doi.org/10.1155/2009/106145*
20. *Akyurek O, Berkalp B, Sayin T, et al. Altered coronary flow properties in diffuse coronary artery ectasia. Am Heart J. 2003; 145: 66-72.*
21. *Zeina A-R, Sharif D, Blinder J, Rosenschein U, Barmeir E. Noninvasive assessment of coronary artery ectasia using multidetector computed tomography. Coron Artery Dis. 2007; 18: 175-180.*
22. *Lau GT-M, Brieger D, Freedman SB. Infarction due to right coronary artery thrombus. Clin Cardiol. 2004; 27: 96.*
23. *Yetkin E, Waltenberger J. Novel insights into an old controversy: is coronary artery ectasia a variant of coronary atherosclerosis? Clin Res Cardiol. 2007; 96: 331-339.*
24. *Yasar AS, Erbay AR, Ayaz S, et al. Increased platelet activity in patients with isolated coronary artery ectasia. Coron Artery Dis. 2007; 18: 451-454.*
25. *Ochiai M, Yamaguchi T, Taguchi J, et al. Angioplasty of stenoses adjacent to aneurysmal coronary artery disease. Jpn Heart J. 1990; 31: 749-757.*
26. *Stefanadis C, Toutouzas K, Tsiamis E, et al. New stent design for autologous venous graft-covered stent preparation: first human application for sealing of a coronary aneurysm. Catheter Cardiovasc Interv. 2002; 55: 222-227.*
27. *Harandi S, Johnston SB, Wood RE, Roberts WC. Operative therapy of coronary arterial aneurysm. Am J Cardiol. 1999; 83: 1290-1293.*
28. *Hasahya T, Kai M, Bangwei W, et al. Among Ectasia Patients with Coexisting Coronary Artery Disease, TIMI Frame Count Correlates with Ectasia Size and Markis Type IV Is the Commonest. Cardiology Research and Practice, Volume 2015. Available at : http://dx.doi.org/10.1155/2015/282170*