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Review Article

Coronary artery ectasia

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ABSTRACT

Coronary artery ectasia (CAE) is defined as localized or diffuse dilatation of coronary artery lumen exceeding the largest diameter of an adjacent normal vessel more than 1.5 fold. The incidence of CAE is reported as 0.3–4.9% of patients undergoing coronary angiography. The rate of recognition may increase with the use of new non-invasive imaging methods like computed tomography (CT) and magnetic resonance (MR) coronary angiography. Atherosclerosis is considered as the main etiologic factor responsible for more than 50% of cases in adults while Kawasaki disease is the most common cause in children or young adults. Coronary ectasia is thought to be a result of exaggerated expansive remodeling, which is eventuated as a result of enzymatic degradation of the extracellular matrix and thinning of the vessel media. Patients with CAE without significant coronary narrowing may present with angina pectoris, positive stress tests or acute coronary syndromes. Ectatic vessel may be an origin of thrombus formation with distal embolization, vasospasm or vessel rupture. The prognosis of CAE depends directly on the severity of the concomitant coronary artery disease. Antiplatelet drugs underlie the therapy. Other management strategies in CAE involve both the prevention of thromboembolic complications and percutaneous or surgical revascularization.

Contents

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1. Introduction

Coronary artery ectasia (CAE) is a well-recognized but relatively uncommon finding encountered during diagnostic coronary angiography [1–3]. It is commonly defined as inappropriate dilation of the coronary arteries exceeding the largest diameter of an adjacent normal vessel more than 1.5 fold [1,4] (Fig. 1). The term 'ectasia' refers to diffuse dilation of a coronary artery whilst focal dilation is called as 'coronary aneurysm' [5]. Even though several mechanisms are suggested the pathophysiology of CAE is still underrecognized. Similarly no consensus exists about the natural history and management of this condition because of the relative scarcity of data. CAE represents not only an anatomical variant but also a clinical constellation of coronary artery disease (CAD) like association with myocardial ischemia and acute coronary syndromes. The purpose of this review is to update and summarize the clinical features of CAE.

2. Epidemiology and classification

Incidence of CAE detected by means of coronary arteriogram has been found to vary between 0.3% and 4.9% [1,3]. In the largest series from the CASS registry, Swaye et al. found CAE in 4.9% of coronary angiograms and the incidence is higher in men than in women (2.2% vs. 0.5% respectively) [3] and postmortem incidence is given 1.4%. Advent of new non-invasive technologies such as computed tomography (CT) and magnetic resonance (MR) coronary angiography, may increase the rate of recognition [6]. Zeina et al. found the prevalence of CAE 8% by coronary CT angiography [7]. Markis suggested a classification of CAE according to the number and diffuseness of involved coronary vessels (Table 1) [7,8]. According to the diameter of the vessel lumen CAE is classified as small (<5 mm), medium (5–8 mm) or giant (>8 mm).

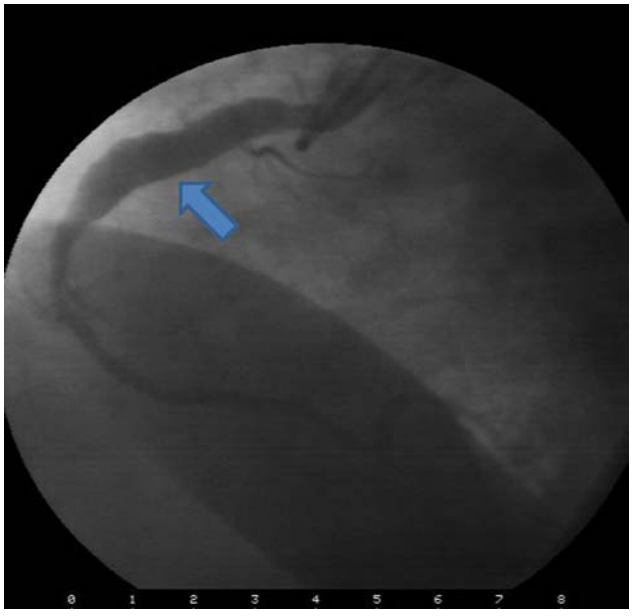

Fig. 1 – Ectasia of right coronary artery (arrow) in a 78 yo female patient.

Table 1 – Markis classification of coronary artery ectasia.

Types of CAE	Definition
Type 1	Diffuse ectasia of two or three vessels
Type 2	Diffuse ectasia in one vessel and localized disease in another
Type 3	Diffuse ectasia in one vessel only
Type 4	Localized or segmental involvement

Table 2 – Etiology of coronary artery ectasia.

- Atherosclerosis
- Coronary artery revascularisation procedures (balloon angioplasty, stent implantation, laser angioplasty, atherectomy, brachytherapy)
- Vasculitides (Kawasaki disease, polyarteritis nodosa, syphilis, Takayasu disease, Wegener granulomatosis, Giant cell arteritis, Churg Strauss Syndrome)
- Congenital malformations
- Chest traumas
- Connective tissue disorders (rheumatoid arthritis, Systemic lupus erythematosus, scleroderma, ankylosing spondylitis, Behçet's disease, Psoriasis)
- Collagenopathies (Marfan's syndrome, Ehlers-Danlos syndrome, hereditary hemorrhagic telangiectasia)
- Primary hyperaldosteronism

3. Etiology and pathophysiology

Atherosclerosis is considered as the main etiologic factor responsible for more than 50% of cases in adults [1,3,4,7,9] while Kawasaki disease is the most common cause in children or young adults [6–8]. Etiology of CAE is demonstrated in Table 2 [10,11]. There are marked histopathological similarities between ectasia and atherosclerosis. Arterial lumen may be narrowed, preserved or dilated with progression of atherosclerosis. The exact mechanism of luminal dilation in some atherosclerotic vessels is unclear while atherosclerosis predominantly causes narrowing of the vessel lumen. Certain plaques, as a result of a phenomenon so called 'arterial remodeling', do not reduce luminal size, presumably because of expansion of the media and external elastic membrane [12]. This finding also may be operative in the case of ectasia or aneurysm of other vessels. Observations with the use of intravascular ultrasound demonstrated that arterial remodeling may be bidirectional according to the expansion or shrinkage of external elastic membrane (i.e. positive and negative remodeling respectively) [12]. Positive (or expansive) remodeling is principally a compensatory mechanism to preserve luminal size during the progression of atherosclerosis. CAE is thought to be a result of exaggerated expansive remodeling in which both external elastic membrane and luminal size increase [11,13]. Enzymatic degradation of the extracellular matrix by matrix-metalloproteinases and other lytic enzymes and thinning of the tunica media associated with severe chronic inflammation is suggested as the key pathogenetic mechanism of the exaggerated expansive remodeling. The severity of the changes in the media correlates positively with the diameter