Unusual presentation of extensive spontaneous coronary dissection: Case report and literature review

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Case Report

Abstract

BACKGROUND: The etiology of spontaneous dissection of coronary artery (SDCA) is not well understood yet. Different studies have linked this entity to pregnancy, physical stress, collagen diseases and vasculitis. In general, patients do not exhibit the classic risk factors for coronary artery disease, which mandates the suspicion of this condition, especially in young adults with acute coronary syndrome.

CASE REPORT: In this article, we report the case of a 63-year old male patient, asymptomatic, who came for periodic evaluation and after evaluation by exercise and myocardial scintigraphy had high suspicion for severe coronary artery disease and underwent coronary angiography, which showed spontaneous dissection of the left and right branches of the coronary arteries.

CONCLUSION: The choice of therapeutic strategies (clinics, percutaneous or surgical) remains uncertain and should be individualized by the features and form of presentation of the SDCA.

Keywords: Dissection, Coronary Angiography, Coronary, Computed Tomography Angiography (CTA)

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Introduction

The spontaneous dissection of coronary artery (SDCA) is defined as a non-traumatic and non-iatrogenic separation of the coronary artery walls, creating a false lumen.[1] This separation may occur between the intima and media, or between media and adventitia, with formation of intramural hematomas (IMH) which compresses the arterial lumen, decreasing the anterograde blood flow and causing myocardial ischemia and/or subsequent heart attack. The SDCA is a rare event, with an estimated incidence of 0.04% to 0.2% of coronary angiographies.[1,2]

Sudden death is the clinical presentation in 50% of cases. It is estimated that 75% of affected patients are female, 40-years old average. It is rare cause of acute coronary syndrome (ACS) and sudden death may be associated with several predisposing factors, such as: fibromuscular dysplasia (FMD), pregnancy, connective tissue diseases (systemic lupus erythematosus, Crohn's disease, polyarteritis nodosa sarcoidosis), Marfan syndrome, Ehlers-Danlos, cystic medial necrosis, hormone replacement therapy, cocaine use, severe hypertension, smoking, strenuous exercise and vasospasm.[2,3] The first case reports and case series about SDCA were formed through post-mortem diagnosis. The current wide availability of coronary angiography allowed the early diagnosis of SDCA.[4]

Case Report

Patient AVT, 63-years old, smoker, with hypertension and dyslipidemia performed ergometric test for cardiovascular risk stratification. There was no previous description of chest pain episodes. The ergometric stress test in Ellestad protocol stopped at 6:04 minutes because of limiting physical fatigue. There was a change of the test due to observation of blood pressure plateau in the effort, being interrogated an inotropic deficit. The same was repeated with performing myocardial scintigraphy with sestamibi injection. There was a description of images with sharp and persistent hypoperfusion predominantly affecting the entire cardiac apex, apical anteroseptal region, and septum associated with ischemic component. As a result, it was decided to carry out evaluation by coronary angiography. The

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right coronary artery had good caliber, tortuous dissection image compromising proximal, middle and distal third involving the origin of the posterior and downward ventricular branches (Figure 1). Anterior descending artery (aDA) with atheromatous plaque and image dissected after origin of the first septal branch, involving the origin of the first two diagonal branches (Figure 1).

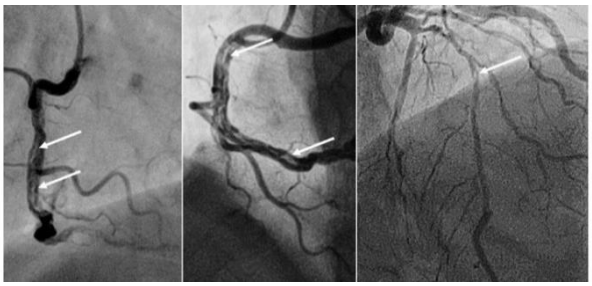

Figure 1. The tortuous dissection image compromising proximal, middle and distal third (I) involving the origin of the posterior and downward ventricular branches. Anterior descending artery with atheromatous plaque and image dissected after origin of the first septal branch, involving the origin of the first two diagonal branches (III)

Patient remained asymptomatic from diagnosis and so we opted for expectant management and prescription of the following medications: losartan, atenolol and aspirin. Patient underwent angiography of coronary arteries about two months after the diagnosis of SDCA that only showed multivessel atherosclerotic disease, with significant reduction of the lumen in aDA and to a lesser extent in the lumen of the right coronary artery (RCA) (Figure 2).

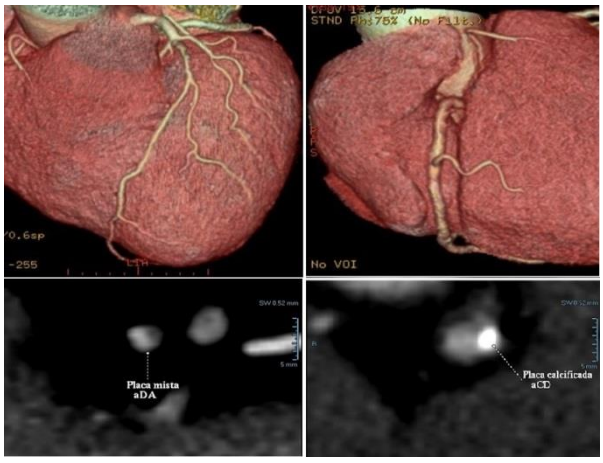

Figure 2. The anterior descending artery (aDA) displays mixed noncalcified plaques with 80% obstruction in the middle third (I/III). The right coronary artery (aCD) has a thick wall plate and calcified source and reduced lumen of about 50% at the distal end (II/IV)

Discussion

The occurrence of SDCA is substantially higher in young patients, and its incidence, etiology and pathophysiology remain unclear.[1,5] However, some authors propose mechanisms to explain the pathogenesis of SDCA.

The first involves a tear in the inner layer of the vessel wall, resulting in blood entry inside the endovascular space to the inner layer, creating a false light full of blood in the vessel.[5-7] The second mechanism of formation may be due to rupture of vasa vasorum, creating an intramural hematoma.[5,8]

Hormonal changes especially those resulting from estrogen levels during pregnancy alter the conformation of normal elastic fibers, alter collagen synthesis and hinder the formation of mucopolysaccharide content, causing the middle layer weakens and increases the risk of creating false lumen and thrombosis.[1,5,8]

Estrogen is believed to be involved by creating a hypercoagulable state. Eosinophilic infiltration in the arterial adventitia has been observed in autopsies of women in the peripartum. It is believed that these eosinophilic granules may cause collapse of the medial adventitial layer and increased lytic substances, predisposing the dissection of the artery.[2,5,8]

The proposed mechanism for our patient finds grounding in previous publications, as in patients with significant risk factors for artery diseases, the SDCA can be precipitated by stressors such as strenuous exercise or emotional stress, which can trigger the event.[9]

Vigorous exercise, especially in isometric form, can increase cardio-circulatory stress and shear forces against the wall of the coronary artery.[7,9] Some authors attribute an intense relationship between the SDCA and FMD. This disease is a non-inflammatory disorder, non-atherosclerotic arterial vasculature leading to arterial stenosis, occlusion, aneurysm formation or dissection. Some case series have a ratio of up to 72% between appearance of SDCA and FMD.[9,10,11] However, our patient had no history of FMD. The clinical presentation of the patients with SDCA is variable. In recent retrospective studies, chest pain was the most common presenting symptom.[2,12,13] Ventricular arrhythmia occurred in 8%-14% of patients.[14] In a recent series of cases, all cases of SDCA had troponin elevation, with 26% presenting with ST-segment elevation, and 3.6% with ventricular arrhythmia.[9,12] Three angiographic patterns are described for SDCA: (i) type 1 (evident

defects of the arterial wall): this is pathognomonic angiographic appearance of SDCA, contrast shows arterial wall defects and multiple radiolucent lumens; (II) type 2 (diffuse stenosis with varying severity): this angiographic entity is not well appreciated and is often misdiagnosed. The SDCA commonly involves the middle and distal segments and can be so great that reaches the distal tip; (III) type 3 (atherosclerosis simulation): this is the most difficult appearance of atherosclerosis to differentiate and less likely to be diagnosed. Some features that favor the angiographic diagnosis of SDCA are: (i) absence of atherosclerotic changes in other coronary arteries, (ii) long lesions (11-20 mm); (iii) blurred stenosis; and (iv) the linear stenosis. The SDCA type I is easily diagnosed with coronary angiography, though type II with shorter lengths, and type III indistinguishable from atheromatous plaques require intravascular ultrasound for correct diagnosis, in order to provide better accuracy for evaluation of these lesions. The natural history of SDCA seems to imply spontaneous healing in most cases. The performance of a new angiography in selected patients in previous studies showed a variable cure, but a full resolution at 40 month follow-up.[15] In another series of cases, 100% of 79 patients treated conservatively had spontaneous healing in angiography performed in ≥ 4 weeks after their SDCA event.[5,8,14]

Coronary computed tomographic angiography (CTA) with multiple detectors were used for patient follow-up with SDCA and found the presence of IMH within 3 days after the event and images approaching the full resolution, with only slight thickening of the wall, at 10 days post-event. Despite improvements, the CTA still does not substitute coronary angiography in the diagnosis of SDCA, but serves as a good follow-up examination and evaluation of healing.[15,16] The standard medical therapy does not yet have a specific guideline and treatment is usually the same as ACS. For pharmacological treatment, the use of aspirin in all patients presenting with ACS and SDCA, as well as its maintenance in subsequent processing is recommended. However the majority of the authors differ in the real benefit of clopidogrel in untreated patients with stent. Some argue that a dual antithrombotic therapy would reduce the IMH and therefore could be beneficial to the patient, however there are cases in series to compare different groups in this specific situation.[17,18]

The role of the new P2Y12 antagonists (prasugrel and ticagrelor) to SDCA is not clear. The role of the GPIIb/IIIa inhibitors in acute management SDCA was also not evaluated, but because of its greater potency, increased bleeding risk and potential risk of prolongation of the dissection, they are not routinely used for SDCA.[18] The role of anticoagulation to SDCA is controversial as the risk of dissection extension overlaps the potential benefit of resolving thrombus and improved permeability of the true lumen. Heparins are routinely used in patient with ACS presentation, though some authors recommend their suspension from the diagnosis of coronary dissection regarding the risk of IMH extension.[19,20] Beta-blockers reduce blood pressure shearing forces and are presumably beneficial in reducing stress in the coronary artery wall, similar to the effect seen in the aortic dissection. Therefore, its use is always recommended, unless contraindications are present.[21]

The use of nitroglycerin can be useful in relieving the symptoms of ischemia superimposed in the vasospasm SDCA presentation, but are not used in the long term. Inhibitors of angiotensin converting enzyme are administered routinely in the same indications of ACS.[8,19] The use of statins for non-atherosclerotic SDCA has not been studied, and the recommendation of the principal authors is to administer statins to these patients only with pre-existing dyslipidemia.[19]

In recently published work, the choice of therapeutic strategies (clinics, percutaneous or surgical) confirmed the uncertain nature of the ideal approach to the SDCA. The predominance of conservative strategy (56%) was based probably on clinical stability, single-vessel feature and inaccurate location of the dissection flap. In most cases, the initial site of intimal rupture is difficult to locate. In case of persistence of symptoms and identification of the entrance hole dissection, percutaneous treatment may be indicated. Urgent surgical treatment seems to be the most suitable for dissections with coronary occlusion, involving multi-vessel or left main coronary artery, and in the presence of cardiogenic shock.[22,23]

Acknowledgments

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Conflict of Interests

Authors have no conflict of interests.

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