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Review

Spontaneous coronary artery dissection

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ABSTRACT

Spontaneous coronary artery dissection (SCAD) is a relatively rare and unexplored type of coronary disease. Although atherosclerosis, hormonal changes during pregnancy and connective tissue disorders might represent a sufficiently convincing explanation for some patients with SCAD, the many remaining cases display only a weak relationship with these causes. While on one side the clinical heterogeneity of SCAD masks a full understanding of their underlying pathophysiologic process, on the other side paucity of data and misleading presentations hamper the quick diagnosis and optimal management of this condition. A definite diagnosis of SCAD can be significantly facilitated by endovascular imaging techniques. In fact, intravascular ultrasound (IVUS) and optical coherence tomography (OCT) overcome the limitations of coronary angiography providing detailed endovascular morphologic information. In contrast, optimal treatment strategies for SCAD still represent a burning controversial question. Herein, we review the published data examining possible causes and investigating the best therapy for SCAD in different clinical scenarios.

1. Introduction

Spontaneous coronary artery dissection (SCAD) is a vessel wall lesion characterized by an intramural hematoma (ie., false lumen) flattening the lumen (ie., true lumen) through the shift of the inner media against the opposite wall, which by definition requires the absence of an iatrogenic, non-coronary or traumatic etiology [1,2]. According to the underlying pathophysiology, SCAD has also been named "primary" coronary artery dissection, in contrast to the definitely more common "secondary" coronary artery dissection, which in contrast is explained by known factors, such as coronary catheterization, percutaneous coronary intervention (PCI), cardiac surgery, extended aortic root dissection or chest trauma (Fig. 1) [3–6].

SCADs generally present an intimal–media tear producing a communication between the vessel lumen and the intramural hematoma ("SCAD with entry door") (Fig. 2, left). Blood coming into the false lumen clots and spreads the flap of detached tissue. The dissection plane, located between the intima and the media or more commonly in the outer media, delimits a new hollow (false lumen) causing usually severe luminal narrowing and distortion [1,2,7]. However, several SCADs at least initially are only intramural hematomas that compress the lumen from the outside reducing the blood flow ("angiographically-invisible SCAD"). Subsequently some of these hematomas can evolve as a result of continuous intramural bleeding and/or structural abnormalities (ie., inflammatory infiltrates, cystic medial necrosis) with the final intimal–media disruption promoting suddenly a further path of growth [1,2,7–9] (Fig. 2, right).

The typical angiographic sign of coronary dissection is a radiolucent intimal–media flap (double lumen), frequently associated with persistent extra-luminal filling and/or delayed clearance of contrast media from the lumen, detected in at least 2 orthogonal projections [1,2,10] (Fig. 3A). However, as mentioned above, SCAD without luminal entry door frequently determines exclusively a smooth and regular coronary narrowing due to intramural hematoma eversion into the lumen [11,12]. These lesions not fulfilling the classical angiographic pattern of coronary dissection can be detected with tomographic imaging techniques, such as intravascular ultrasound (IVUS) and optical coherence tomography (OCT) (Fig. 3B, C) [11,12].

The aim of this article is to provide a comprehensive description of SCADs, review their presentation and outcomes from the available literature, and critically appraise current diagnostic and therapeutic options.

2. Epidemiology of SCAD

SCAD is a relatively rare presentation of coronary disease with an estimated prevalence of 0.07–0.28% [12–17] and an annual incidence of 0.26 cases per 100,000 persons (0.33 in women, 0.18 in men) among US subjects [18]. Since the first case described by Pretty in 1931 [19], a total of only ~1125 SCADs (Appendix A) have been reported in

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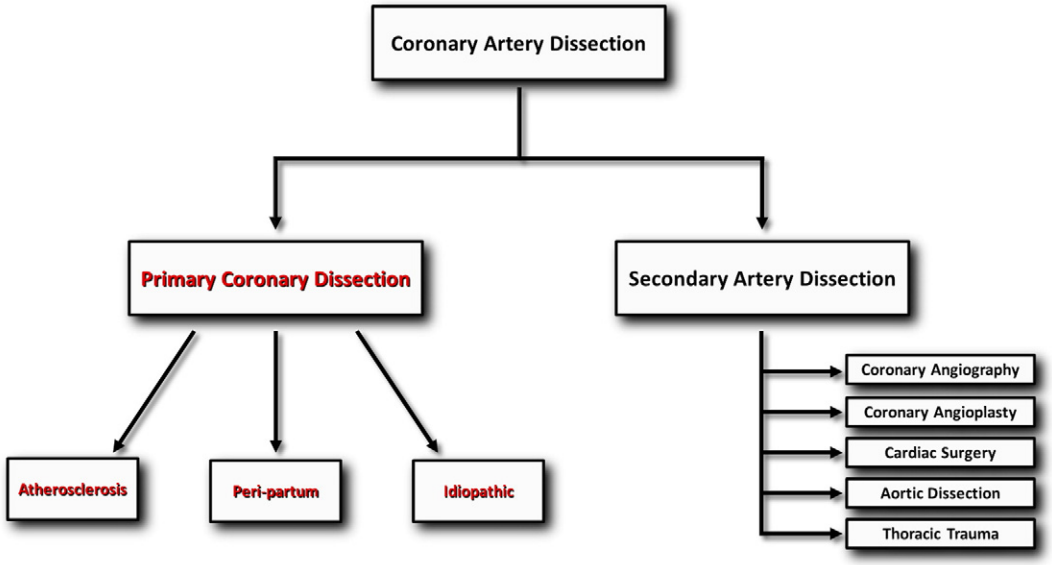

Fig. 1. Coronary artery dissections classification: Coronary artery dissections can be classified as "Primary", or spontaneous coronary artery dissections (SCADs), and "Secondary". By definition, SCAD requires the absence of a traumatic, iatrogenic and non-coronary process (aortic dissection). Secondary dissections are generally consequent to coronary angioplasty (wiring, balloon dilatation, stenting), coronary angiography (coronary catheterization), cardiac surgery, ascending aortic dissection and thoracic trauma.

literature consisting largely of individual cases or case series and rarely of small single-center registries (<90 patients).

Patients with SCAD generally present without cardiovascular risk factors or history of coronary events [13–17] and are typically female (58–79%) [1,12,13,20] and young, with a mean age of 41 ± 10.6 years in women and of 45.4 ± 14.4 years in men [21]. SCAD may account for more than 10% of STEMI in women <50 years of age [14]. Recently, SCAD presentations have been documented also in adolescents and in elderly [12,22–26]. The early assumption of a gender-specific location of SCAD, characterized by more frequent left coronary artery involvement in women (84–88%) and right coronary artery involvement in men (67–73%) [3,27], has been recently rejected, since the left anterior descending represents the most frequent site of SCAD both in males and in females (69% vs 72%; $p = 0.81$) [12,13,18]. Regardless of the coronary artery involved, the mid segment appears the preferential site [12,26].

Combining data extracted from existing small registries and case series (Table 1, see Appendix A for a description of the adopted methodology), the left anterior descending artery is the most commonly involved vessel (59.7% of patients) followed by the right coronary artery (26.5% of patients), the left circumflex (19.4% of patients) and the left main coronary artery (8.9% of patients) (Fig. 4, upper; Table 1). Multi-vessel SCAD is nearly three times less common than single-vessel SCAD (26.4% vs 76.3%; $p < 0.01$) [21]. Comparing multi-vessel involvement frequency between genders, the prevalence of SCAD is higher in women both in the two-vessel (17.5% vs 12.9%; $p = 0.022$) and in the three-vessel SCAD presentations (12.3% vs 5.3%; $p = 0.029$) (Fig. 4, mid). Conversely, men more often present with single-vessel SCAD (81.8% vs 70.1%; $p = 0.011$) (Fig. 4, mid) [21]. Reports of SCAD simultaneously involving all the three major epicardial arteries are anecdotal [28,29]. The prevalence of SCAD increases significantly in acute coronary syndromes (ACS) because the dissection is frequently responsible for an abrupt and severe flow limitation. However, there is a substantial discordance among reports in the frequency of each type of ACS. For instance, Mortensen et al. found the majority of patients presented with ST Elevation Myocardial Infarction (STEMI) at admission (84%), Tweet et al. indicated similar rates of STEMI (49%) and Non-ST Elevation Myocardial Infarction (NSTEMI) (44%), and Kansara et al. concluded that NSTEMI was the usual presentation of SCAD (85%) [13,18,30].

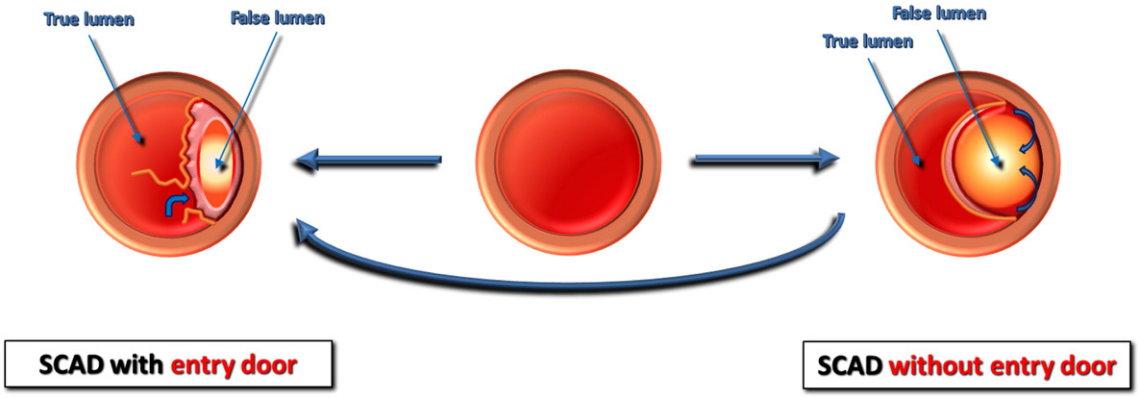

Fig. 2. Morphologic patterns: Spontaneous coronary artery dissection (SCAD) presents two patterns, one ("SCAD with entry door", left) characterized by a luminal entry door and a detached intimal–media membrane (radiolucent flap at angiography), the other (SCAD without entry door or "angiographically-invisible SCAD", right) characterized by an intramural hematoma without communication with coronary lumen. Probably most SCADs are at least initially SCAD without luminal entry door that subsequently break suddenly following the progressive enlargement of the intramural hematoma. The sudden blood inflow into the false lumen causes generally an abrupt vessel flow limitation and represents a further path of growth.