

BRIEF SUMMARY OF PATHOGENESIS, DIAGNOSIS AND MANAGEMENT OF SPONTANEOUS CORONARY ARTERY DISSECTION

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Keywords: coronary artery dissection, spontaneous, acute coronary syndrome, intramural hematoma.

Summary

Spontaneous coronary artery dissection (SCAD) is a rare cause of acute coronary syndrome (ACS). It was first described 80 years ago. Pathogenetic mechanisms are most likely to be associated with intima tear or bleeding vasa-vasorum, which resulting in intramural haemorrhage. SCAD typically occurs in young women who do not have coronary heart disease risk factors and who have acute coronary syndrome. Half of all SCAD presents with ST - elevation myocardial infarction (STEMI), while the rest with non - ST - elevation myocardial infarction (NSTEMI). The gold standard method for diagnosis is interventional coronary artery angiography. After the acute ischemic onset syndrome, most patients have a stable, benign clinical course, and eventually experience spontaneous vessel wall healing. Therefore, conservative treatment (a watchful strategy) is recommended as the initial treatment. For the majority of cases as interventional and surgical treatment in most cases seems to be suboptimal. In this extremely complex situation, several novel and attractive coronary interventions have been proposed. The risk factors, pathogenesis theories, diagnosis, management, prognosis of SCAD will be summarized in this review.

Introduction

As first described in 1931, spontaneous coronary artery dissection (SCAD) is a non-atherosclerotic coronary artery disease that manifests as acute coronary syndrome (ACS) or death (1). Primary or isolated SCAD is a disorder due to intramural hematoma (IMH) and / or dissection that obstructs coronary flow, which is not related to trauma nor is iatro-

genic. During pregnancy and soon after childbirth, women are more susceptible to developing SCAD. According to Mayo clinic Registry data, about 17 % of SCAD is associated with pregnancy (2). SCAD, previously considered rare, is responsible for 24-36% of myocardial infarction (MI) in women under the age of 50 and is now recognized as the most common cause of pregnancy-related MI (3). SCAD mainly affects women (92-98% of cases), who are relatively young, 42-52 years old, and do not have traditional cardiovascular risk factors (1,2). The prevalence of SCAD was 0,49 % according to National Inpatient Sample database, presenting more than 13 million patients admitted due to ACS between 2005 and 2015 (4,5). However, these estimates probably not represent the real incidence of SCAD, as the disease could be often misdiagnosed, especially in the early era.

The aim of this review was to evaluate the risk factors, pathogenesis, diagnosis, management and prognosis of spontaneous coronary artery dissection.

Methodology

Data search in electronic scientific databases PubMed, Embase in the period 2016-2019 was performed. Search words: coronary artery dissection, spontaneous, acute coronary syndrome, intramural hematoma. A review of twenty-nine literature sources was conducted.

Risk factors and pathogenesis. SCAD has no pathogenic risk factors. It was noticed that, SCAD more often affects young women who do not have cardiovascular factors for cardiovascular disease (6-8). It is currently thought that risk factors for SCAD are multiple pregnancies (more than 4 times), administration of hormone replacement therapy, infertility therapy, connective tissue diseases, in particular, fibromuscular dysplasia, vascular Ehlers-Danlos syndrome, Marfan syndrome, Loeys-Dietz syndrome, emotional and physical stress (2,9-12). Two major pathogenesis mecha-

nisms of SCAD are currently described: (a) hypothesis of medial haemorrhage which indicates that haemorrhage in the artery wall is due to spontaneous rupture and intramural bleeding from vaso-vasorum is a major pathogenic mechanism; and (b) an intimal tearing hypothesis that disruption of the intimal interface creates an entry point for intramural hematoma accumulation in the false lumen and leads to arterial wall separation (1,13–15).

Clinical presentation. Have been admitted SCAD patients complaints characteristic to ACS. Like cases of ACS, SCAD is characterized by chest, shoulder or upper abdominal pain, with or without radiation to the arm, nausea or vomiting, neck irradiation, sweating, dyspnoea, and rarely back pain, dizziness, fatigue, headache, fainting (1). Symptoms vary in severity from mild to severe, and cardiac dysfunction can cause arrhythmias, cardiogenic shock, heart failure (16,17).

Diagnosis. SCAD manifest as ACS and thus usually is preliminary diagnosed by ECG and laboratory tests (series of troponin tests). Half of all patients have been diagnosed with ST - elevation myocardial infarction (STEMI), the rest – with non - ST - elevation myocardial infarction (NSTEMI) (1,2,18). SCAD is diagnosed in most patients during coronary angiography (19). Indications for coronary artery angiography are ordinary. In 2014, Saw et al. (20) have developed 3 types of SCAD classification based on angiography (table 1). Non-invasive procedures, such as computed tomography and magnetic resonance coronary angiography, have limitations and are not used for SCAD diagnosis.

Management. Generally, the main treatment strategies for SCAD are coronary artery bypass surgery, percutaneous coronary intervention (PCI), medical therapy, and active surveillance. The choice of these treatment strategies still represents a debatable issue and may depend on individual clinical symptoms, the localization and spread of the lesion, the experience of the treating institution, and previous relapses.

Conservative medical therapy alone is preferred in stable patients. Medical therapy consisted of aspirin, P2Y12 inhibitors (clopidogrel) and beta blockers, followed by angiotensin converting enzyme inhibitors, calcium channel blockers, nitrovasodilators, and angiotensin II receptor blockers, for stent or bioresorbable vascular scaffold strategies, dual antiplatelet therapy is generally prescribed, usually for 12 months (21). In general, long-term results for SCAD patients are excellent, and most patients can safely receive medical therapy. Several retrospective case series have reported that spontaneous angiographic healing occurred in 73-97% of cases when repeated angiography was performed (1). However, SCAD can be life-threatening because some

patients experience early or late complications, including SCAD of other coronary vessels. Therefore, further testing and optimal monitoring strategies have to be defined (22,23). Currently, SCAD PCI and surgical treatment algorithm have been proposed to simplify the course of intervention (figure 1). Revascularization in SCAD patients is very difficult due to subcutaneous damage to coronary vessels (24). Although stenting is usually delayed, the urgency of revascularisation depends on clinical status and hemodynamic stability and urgent PCI is usually successful in life-threatening situations (25). Indications for revascularisation include complete vessel occlusion, left main involvement, ongoing ischemia, recurrent chest pain and haemodynamic instability, as well as sustained ventricular arrhythmias.

Coronary artery bypass grafting (CABG) is commonly used as a rescue strategy in failure of PCI for SCAD patients (24,26). In case of diffuse left main dissection the literature on the CABG approach is limited by clinical cases. The short-term outcomes of CABG treatment appear to be good, but the long-term coronary artery graft failure has been observed. Most likely due to spontaneous coronary artery wall healing. It has been suggested that both venous and arterial conduits may fail when the dissected artery has healed (2). Therefore, CABG is reserved for multivascular dissection and extreme cases of hemodynamic instability.

Alternative treatment for SCAD has been proposed performing fenestrations. Motreff et al. (27) conducted a study evaluating the efficacy of fenestrations. The intima was cut with a scoring or a cutting balloon inducing intramural hematoma decompression, excellent long-term results have been reported (27). It seems like cutting balloon angioplasty before stent placement prevents longitudinal extension of intramural hematoma and helps to avoid unnecessary stent implantation.

Prognosis. According to the largest contemporary cohort study, hospital mortality from SCAD was 4.2 percent (4). In a retrospective study including 87 patients, a SCAD relapse rate of 17 percent was observed (28). Hospital prognosis

Table 1. SCAD angiographic classification

Type 1	Type 1 shows a classic appearance with contrast staining of the arterial wall and multiple lumens
Type 2	Type 2 represents diffuse stenosis of different weights and lengths
Type 2A	Type 2A stenosis is restricted by the normal proximal and distal lumen
Type 2B	Type 2B stenosis extends to the distal tip of the artery
Type 3	Type 3 represents localized or tubular stenosis that is difficult to distinguish from atherosclerosis

is generally great for people undergoing conservative treatment, CABG or percutaneous coronary intervention (28,29).

Conclusion

Spontaneous coronary artery dissection defined as the non-traumatic and non-aetrogenic separation of the coronary artery wall. It is a rare cause of acute coronary syndrome. Although rare, SCAD could be considered in young women presenting with ACS, even if there is no cardiovascular risk factors. The gold standard for diagnosing SCAD remains percutaneous coronary artery angiography. Despite improved evidence, the management of SCAD remains based on individual experience considering patients status and haemodynamic stability.

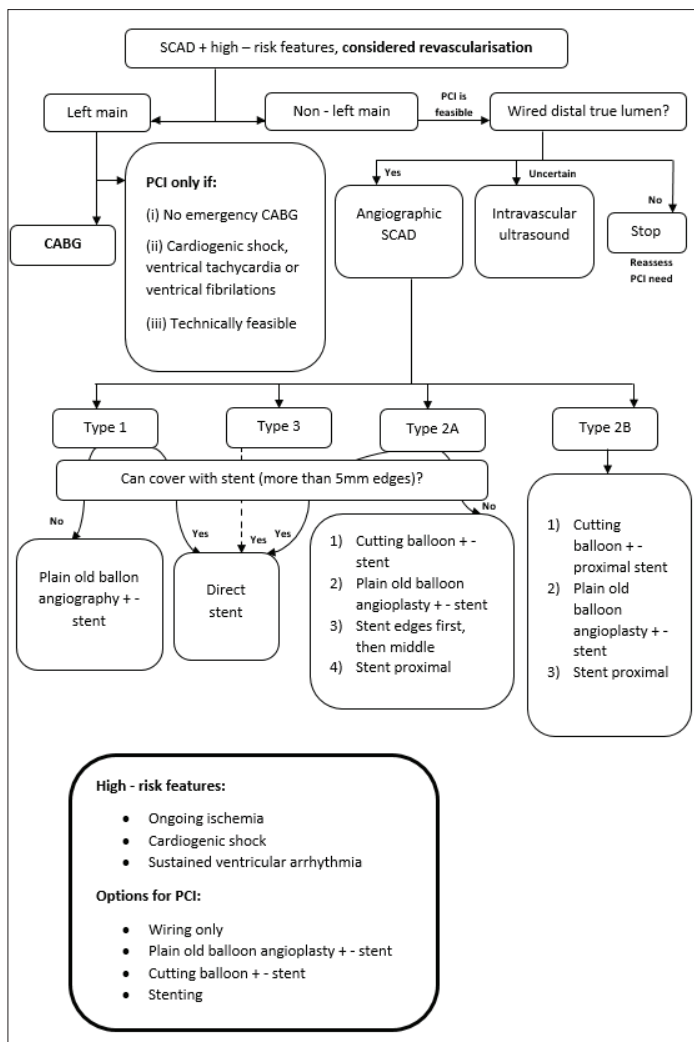


Figure 1. Suggested algorithm for SCAD PCI and CABG treatment. Adapted from: Jacqueline Saw. Natural history of spontaneous coronary artery dissection: to stent or not to stent?

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**SPONTANINĖS KORONARINIŲ ARTERIJŲ
DISEKACIJOS PATOGENEZĖS, DIAGNOSTIKOS IR
VALDYMO APŽVALGA**

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Raktažodžiai: vainikinių arterijų disekacija, spontaninis, ūmus koronarinis sindromas, intramuralinė hematoma.

Santrauka

Spontaninė vainikinių arterijų disekacija (SVAD) yra reta ūmaus išeminio sindromo priežastis. Pirmą kartą SVAD buvo aprašyta prieš 80 metų. Patogeneziniai mechanizmai greičiausiai yra susiję su įplėšimu ar kraujavimu vasa-vasorum, dėl kurio atsiranda intramuralinis kraujavimas. SVAD dažniausiai nustatoma jaunoms moterims, neturinčioms koronarinės širdies ligos rizikos veiksnių ir kurioms yra ūminis koronarinis sindromas. Pusė SVAD atvejų nustatomas miokardo infarktas su ST pakilimu, o likusiai daliai – be ST pakilimo. Auksinis standartinis diagnostinis metodas yra

intervencinė vainikinių arterijų angiografija. Po ūminio išeminio sindromo daugumos pacientų klinikinė eiga yra stabili ir gerybinė, stebimas spontaninis kraujagyslių sienelių gijimas. Taigi, kaip pradinį gydymo metodą rekomenduojama taikyti konservatyvų medicininį gydymą (budrus stebėjimas). Daugeliu atvejų intervencinis ir chirurginis gydymas atrodo suboptimalus. Esant tokiai nepaprastai sudėtingai situacijai siūlomi keli nauji ir patrauklūs koronariniai intervenciniai gydymo metodai. Šioje apžvalgoje apibendrinami rizikos veiksniai, patogenezės teorijos, diagnostikos ir valdymo metodai bei SVAD prognozė.

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Gauta 2020-02-18