

A New Score System for Diagnosis of Spontaneous Coronary Artery Dissection

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Introduction

Back in 1931 Pretty has published the first reported case of spontaneous coronary artery dissection (SCAD) in a 42-year old woman suddenly death due to a rupture of right coronary artery (RCA) atheroma [1]. Eighty-five years later, much has been known but there is much more to discover yet.

In the Literature, there are numerous cases of patient clinically presenting with myocardial infarction (MI), acute coronary syndrome (ACS) or vasospastic angina, in the meanings of "X syndrome", primary angina or Prinzmetal variant showing angiographic non-obstructive coronary artery disease (NOCAD). According to previous studies, NOCAD is associated with a rate of major adverse cardiovascular events equal to obstructive CAD [2,3].

NOCAD has been angiographically defined how less than 50% stenosis of luminal diameter [2]. Previous authors have reported that as many as 10% of myocardial infarction (MI) patients have evidence of NOCAD [2-4]. In this view, although many limitations and gaps are present in current knowledge how they have affirmed, much has been understood thanks to intravascular ultrasound (IVUS has a spatial resolution of 150 μm) as well as optical coherence tomography (OCT has a spatial resolution of 10-20 μm). An interesting type of NOCAD is the SCAD that is an extremely rare phenomenon that represents 0.1-1.1% in most angiographic series and 0.1%-4% of all ACS [5]. Nevertheless, these findings are greatly underestimated still today because of a not negligible numbers had sudden death, an extensive underuse of coronary angiography (CG) in young women and the well-documented limitations of CG to detect SCAD [5]. The SCAD etiopathogenesis remains unclear but some evidence correlate with connective/collagen tissue alteration or changes and higher vessels wall shear stress [4,5].

Clinical presentation may consist in angina pectoris, STEMI/NSTEMI, cardiogenic shock or, not so rare, sudden death [4,5]. According to our experience and Literature review, we have affirmed provocatively that vasospasm could have been overestimated in pre-intravascular imaging era, to explain what angiography was not able to detect, as reported in several studies [6,7] and our own experience [8].

Particularly, spontaneous coronary artery dissection (SCAD) as a cause of acute myocardial ischemia in women (80% of SCAD patients) [2,5]. In this view, we would like to do further considerations: more than 70% of SCADs are misdiagnosed by angiography [6] and in suspected cases only intravascular imaging may verify arterial wall integrity [6,7]. With these premises, IVUS or OCT analysis becomes of primary importance with an improvement in treatment and prognosis [4-7]. Here we want to go beyond the previous considerations: To the best of our knowledge, our group has published for the first time in the literature a flow-chart for a faster and efficient diagnosis and a proper treatment according to literature review [4-7] and our own experience [8-10].

According to our clinical/angiographic score [11] (Figure 1) we

have dedicated a large interest to women. We have assigned: 1 point for 1) female gender, 2) <50-year old, 2) peripartum period or with multiple prior pregnancies, 3) no classical coronary risk factors, 4) estrogenic therapy, 5) emotional/physical stress, 6) cocaine/amphetamines/vasospastic drugs abuse. Angiographic presentation: 7) diffuse, typically smooth arterial narrowing rather than pathognomonic contrast staining in the arterial wall, 8) one vessel disease (no typical atherosclerotic lesions in other coronary arteries), 9) long/tortuous suspected lesion. 2 points for 10) connective tissue disorder (Marfan Syndrome, Ehlers-Danlos Syndrome, cystic medial necrosis)/coronary artery tortuosity and extracoronary vascular abnormalities, (especially fibromuscular dysplasia), 11) systemic inflammatory disease (systemic lupus erythematosus, Crohn's disease, sarcoidosis, polyarteritis nodosa, vasculitis, Beçhet disease). 3 points for 12) history of coronary artery spasm, 13) frequent SCAD recurrences.

When there are at least 3 points in a patient presenting with chest pain, ECG/echocardiography abnormalities (i.e. transitory/permanent ST-segment elevation, wall motion anomalies) or troponin rise/fall, we suggest OCT/IVUS analysis to exclude SCAD as we successfully have shown [8-11]. Apropos of treatment, we think that a conservative approach should be considered only for asymptomatic patients, distal vessel SCAD or <3.0 mm diameter although, an invasive approach should be considered according to the clinical/angiographic characteristics of the patient [8-10]. On the other hand, BVS strategy should be preferred in case of proximal/middle vessel lesion, \geq 3.0 mm diameter or patient still symptomatic/hemodynamically unstable as recently reported [12]. Primary importance has a close follow-up, with or without invasive coronary imaging, to assess the risk of SCAD recurrence and the optimal sealing of the vessel over the time [8-10].

Surely, large clinical trials are needed to investigate this increasing disease affecting young, more often women. In this light, here we want to stress how physicians should keep in mind NOCAD and mostly SCAD first of all in young women presenting with ACS. From our point of view, in absence of international guidelines or universal expert consensus, our provocative approach could be a good starter point to avoiding misdiagnoses and to perform a faster and better therapy.

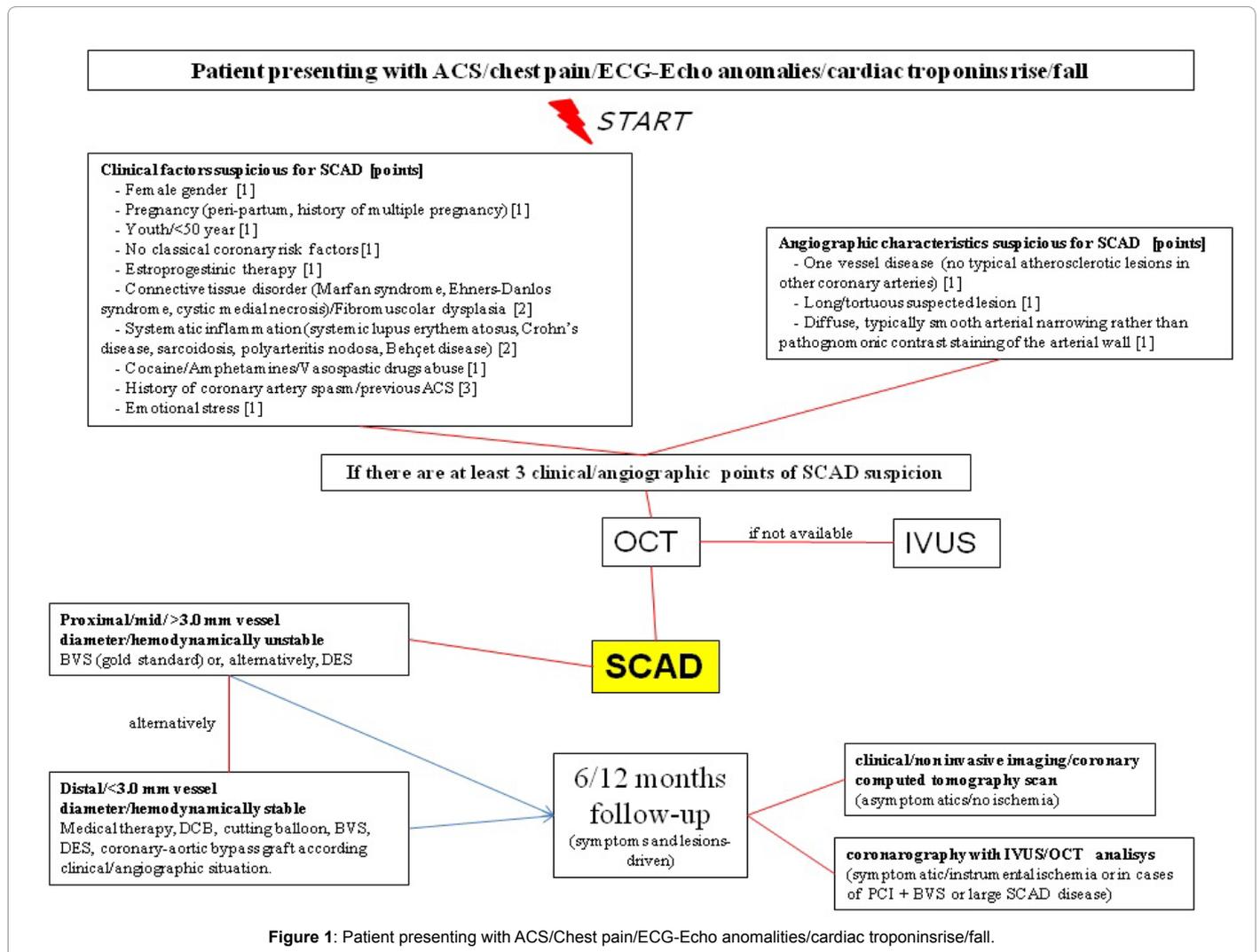
In conclusion, we want further to stress how in the last years the attention for NOCAD and SCAD has highly increased thanks to proper

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diagnosis by intravascular image technique (OCT/IVUS) furthermore, treatment strategy is nowadays strongly debatable because of favourable preliminary results regarding BVS therapy in this field which affects mainly young patients, especially women.

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