Spontaneous Coronary Artery Dissection: Presentation of Three Cases and Short Review of the Literature

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Introduction

Spontaneous (not iatrogenic) coronary artery dissection (SCAD) is a rare cause of acute coronary syndromes (ACS), including cardiogenic shock and sudden cardiac death. Except common atherosclerosis, peripartum period is a documented predisposing factor, as well as arteritis, usually as a result of auto-immune response. This diagnosis is typically presented in a diagnostic coronary angiography. All therapeutic options are acceptable, mainly percutaneous coronary intervention (PCI) but also drug therapy and surgical treatment. We present three cases of SCAD at our institution, with a short review of the literature.

Case Presentation

Patient 1

A 54 year-old female with no previous history of heart disease and no risk factors for coronary artery disease, was transferred to our hospital from a peripheral Health Center due to acute anterior myocardial infarction (STEMI). Angina occurred 2 hours after her participation in a Marathon race. Coronary angiography revealed SCAD of distal LAD with TIMI-II peripheral flow (Figure 1). Direct stenting was performed, with good angiographic result. Four days later, the patient was discharged after a mild elevation of cardiac enzymes and an uncomplicated clinical course. She is asymptomatic, two years after the acute coronary syndrome.

Patient 2

A 76 year-old hypertensive and hyperlipidemic male, with history of atherosclerotic coronary artery disease (two percutaneous coronary interventions 3 and 4 years ago) was admitted due to unstable angina. Coronary angiography in combination with Optical Coherence Tomography (OCT) revealed SCAD of mid-LAD with TIMI-III peripheral flow (Figure 2). The culprit lesion was not in contiguity with the location of the previous interventions. Direct stenting was performed, with good angiographic result (confirmed with OCT), followed by uncomplicated clinical course. The patient was discharged two days later. He is asymptomatic, two years after the acute coronary syndrome.

Figure 1: a) Spontaneous dissection in the distal LAD after strenuous effort (arrow), b) Contrast staining at the dissection site in the distal LAD (arrow), c) Successful treatment with stent implantation.
Patient 3

A 32 year-old female with no previous history of heart disease and no risk factors for coronary artery disease, was transferred to our hospital from a provincial Hospital in cardiogenic shock due to acute anterior myocardial infarction (STEMI). Thrombolysis was administered at the provincial Hospital 6 hours before transfer, followed by severe electrocardiographic and clinical deterioration (air-transfer was delayed due to no medical reasons). Urgent coronary angiography revealed extended dissection of left coronary artery (Figure 3). Angioplasty was performed with angiographic improvement of TIMI flow in LAD, but no significant clinical improvement. The patient died five hours later.

Discussion

SCAD is an underdiagnosed (due to asymptomatic cases) but still rare condition, which usually causes ACS of various severity. The non-traumatic nature of SCAD should be emphasized, in contrast with the most common iatrogenic coronary artery dissection during PCI. In general, SCAD is diagnosed in less than 1% of patients with ACS, although routine use of OCT reveals 4% incidence of SCAD in such patients [1]. Women are affected twice as men, with especially high prevalence of SCAD (up to 24%) among women younger than 50 year-old presenting with STEMI [2-4]. Left Anterior Descending Artery (LAD) accounts for approximately 60% of cases [1-4]. Its presence can be suspected in relatively young patients without (or with minimal) risk factors for atherosclerotic coronary artery disease, especially in pregnant women with ACS. Growing utilization of diagnostic coronary angiography with the contribution of intravascular ultrasound (IVUS) and OCT, is responsible for increased recognition of SCAD.

Regarding pathogenesis, two well described pathophysiologic mechanisms are implicated: atherosclerotic plaque rupture resulting in an intimal flap which extends distally, as well as local inflammatory
process which breaks down the medial-adventitial layers, causing rupture of the vasa vasorum. Notably, eosinophilic infiltrates with lytic substances like collagenase, contribute to the above mentioned pure "inflammatory process", i.e. inflammation without apparent atherosclerotic disease [5]. During pregnancy or in the first weeks after delivery, hormonal and hemodynamic factors may affect elastic and collagen fibers of arterial wall [6]. Theoretically, elevated estrogen and progesterone levels during pregnancy may induce eosinophils to release lytic substances, contributing to the predisposition to dissection in the peripartum period [7].

Increased shear stress, intensified by local arterial spasm or high blood pressure, may contribute to dissection [4]. Cystic medial necrosis, provoked by chronic arterial hypertension, may also weaken the arterial wall and predispose to dissection [8]. Connective tissue disorders (Marfan’s syndrome, Ehlers-Danlos syndrome, fibromuscular dysplasia) vascularitis like polyarteritis nodosa, extreme physical exertion, cocaine use and specific drugs (oral contraceptives, cyclosporine, 5-fluorouracil) have all been associated with SCAD [9-13]. SCAD has also been reported in patients without any risk factors [14]. Similar to our third patient, thrombosis in patient with SCAD has been reported to cause clinical deterioration on the base of extension of the dissection [15].

SCAD is a rare condition without randomized trials of treatment. Empirically, the indications of treatment of SCAD are similar to those of atherosclerotic disease, based on both clinical and angiographic factors. When conservative treatment is preferred, usual antithrombotic and antiischemic therapy of ACS should be applied (except thrombolysis). Treatment of SCAD with PCI or coronary artery bypass graft surgery, may confront specific considerations and difficulties. In percutaneous intervention, special consideration should be given in placing the guidewire in the true lumen. Before stenting, the evaluation of the length of dissection and the vessel's size may be challenging. OCT or IVUS may contribute in both fields, by confirming the desirable positioning of the guidewire and estimating precisely the exact dimensions of the dissected part of the vessel. Patients referred to cardiac surgeons for urgent or elective surgical treatment are usually these with quite long dissections. As a result, surgical grafting to the true lumen may also confront specific difficulties [16]. Limited data suggest favorable outcome of SCAD after initial treatment, with low recurrent rates of 5-10% [17-20], although PCI failure can be relatively high, in comparison with ACS due to common atherosclerotic coronary artery disease. Conservative management in SCAD cases with preserved vessel flow without ongoing ischemia, may have similar outcome with revascularization, giving a reliable alternative in such cases [19]. Administration of antiplatelets aims to prevent total vessel occlusion while negative chronotropic agents (mainly B-blockers) are also quite useful as they support coronary perfusion by prolonging the diastolic period [21]. Elevated heart rate can also induce and promote the atherosclerotic process by mechanisms involving endothelial shear stress (which is a possible determinant for SCAD) and biochemical activities [22]. Multivessel spontaneous dissection of the left coronary tree (like our third patient) is an even less frequent and devastating condition, with limited reports in medical literature [23]. After acute treatment, the possibility of underlying connective-tissue or inflammatory disease should be examined in selected patients with clinical suspicion.

In our three cases with SCAD, we faced three different presentations from the spectrum of ACS (uncomplicated STEMI, unstable angina and STEMI complicated with cardiogenic shock). The unfavorable outcome of the third patient was practically inevitable due to the devastating delay of the air-transfer, despite the subsequent PCI.

The awareness of SCAD as an uncommon pathophysiological substrate of ACS is quite important for its proper diagnosis and management. In patient with STEMI and clinical suspicion of SCAD, thrombolysis should not be an alternative to urgent transfer (in acceptable time-delay) for primary PCI, but should be considered only in patients without any possibility for primary PCI.

In conclusion, SCAD is a rare and challenging angiographic finding in patients with ACS. Peripartum period is the strongest prognostic factor of its presence. It can also be suspected in relatively young patients with connective tissue disorders or chronic inflammatory diseases, with no (or minimal) risk factors for coronary artery diseases. PCI, conservative or surgical therapy can be applied in individualized basis in patients with SCAD, with favorable long-term prognosis and low recurrent rate. Regarding the relatively high failure rate of revascularization in comparison with common atherosclerotic disease, conservative treatment is a good alternative in patients with SCAD and preserved vessel flow, without ongoing ischemia. Intravascular imaging (OCT, IVUS) may improve not only the diagnostic accuracy of coronary angiography, but also the outcome of PCI in such patients.

References


