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Multimodality Imaging for Spontaneous Coronary Artery Dissection in Women

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ABSTRACT

Spontaneous coronary artery dissection (SCAD) has gained attention as a key cause of acute coronary syndrome and sudden cardiac death among women. Recent advancements in cardiac imaging have improved identification and accelerated awareness of SCAD. Accurate diagnosis of SCAD through use of imaging is critical, as emerging evidence suggests that the optimal short- and long-term management strategies for women with SCAD differs substantially from that of women with atherosclerotic coronary disease. This review summarizes the application of both invasive and noninvasive imaging for the diagnosis, assessment, surveillance, and treatment of women affected by SCAD.
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Spontaneous coronary artery dissection (SCAD) is emerging as a much more common cause of acute coronary syndrome and sudden cardiac death than previously recognized, especially in women. The primary event at the level of the arterial wall may involve hematoma formation in the media, separation of the intimal layer, or both. These processes are distinct from atherosclerosis. Antegrade coronary blood flow may be directly impeded by an intimal dissection plane within the lumen or indirectly compromised by compression from medial hematoma. The consequence of the dissection process may be myocardial ischemia, myocardial infarction (MI), or sudden cardiac death. Unlike atherosclerotic coronary artery disease, SCAD has a propensity for young women without traditional risk factors (1), some of whom are highly fit (2).

SCAD was previously considered a rare event, a belief reinforced by published reports of only isolated cases, small single-center case series, and postmortem descriptions. Due to a combination of heightened awareness of the impact of heart disease in women and SCAD in particular, the availability of sensitive biomarker assays, the advent of adjunctive intravascular imaging during coronary angiography (CA), and prevalent noninvasive vascular imaging (3), SCAD is now more commonly recognized. Subsequently, this has amplified familiarity of the SCAD angiographic patterns in patients who may have previously been misdiagnosed as atherosclerosis, spasm, or vasculitis. Despite recent estimates of higher prevalence than previously reported (1,4), SCAD remains incompletely understood. Ongoing efforts aim to further define SCAD and delineate the environmental, molecular and genetic contributors to incident SCAD. Both invasive and noninvasive cardiovascular imaging techniques are integral to the diagnosis, characterization, and management of patients with SCAD. Moreover, multimodality imaging is critical for ongoing efforts to elucidate the cause and mechanism of SCAD. This report reviews the role of multimodality imaging in the diagnosis, evaluation, surveillance, and treatment of women with this condition (**Central Illustration**).

EPIDEMIOLOGY, ETIOLOGY, AND DEMOGRAPHICS

SCAD prevalence remains uncertain and in retrospective studies has been reported as 0.07% to 1.1% (1,5–7). With the exception of 1 community-based study, these studies determined prevalence via angiographic databases, some of which included forensic databases and/or patients who had dissection associated with

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atherosclerosis. In addition, these studies included patient cohorts undergoing diagnostic angiography prior to the use of intravascular imaging such as intravascular ultrasound (IVUS) or optical coherence tomography (OCT), which are pertinent for distinguishing SCAD when obvious features are not apparent on angiography (8). Forensic databases may be limited as SCAD can be missed at autopsy if it is not specifically considered in the differential at the time of postmortem examination, and fatal dissections can occur in the distal coronary arteries not be routinely examined (9). Therefore, these aforementioned studies likely under-appreciate the frequency of SCAD (8,10,11) with SCAD being more prevalent, chiefly in women, than previously reported when intravascular imaging is employed (12).

The first description of coronary dissection is credited to Dr. Pretty (13) in 1931, who described the sudden death of a 42-year-old multiparous and otherwise healthy woman following chest pain, nausea and vomiting. Autopsy revealed dissection and rupture of the right coronary artery. Since then, autopsy series have observed SCAD in young women but have not provided conclusive evidence of its underlying pathogenesis. In 1982, Robinowitz et al. (14) evaluated the coronary arteries in 8 women with SCAD and range of 26 to 47 years of age, 75% of whom presented with sudden cardiac death. In these patients, dissection occurred primarily in the outer third of the media with inflammatory infiltrates, predominantly eosinophilic granulocytes, noted in the adventitia. Out of these observations came a proposed mechanism for dissection whereby localized eosinophilia with hormone-enhanced release of lytic collagenase, peroxidase, acid phosphatase, and major basic protein lead to erosion of the coronary media and adventitia (15,16). However, another series found that eosinophilic infiltrates are not consistently present and may be a reactionary mechanism (17). Inherent abnormalities and neovascularization of the underlying vasa vasorum are thought to increase susceptibility for plaque rupture in typical atherosclerosis (18); one may hypothesize that dysfunctional vasa vasorum could contribute to the underlying pathogenesis of SCAD.

In recent series, patients with SCAD are most often women (74% to 92%) with mean age ranging from 42 to 52 years (1,19). Patients present with acute coronary syndrome including ST-segment elevation myocardial infarction, non-ST-segment elevation myocardial infarction, and sudden cardiac death (1,19). Predisposing factors may include female sex; extracoronary vascular abnormalities (EVA) such as fibromuscular dysplasia (FMD), coronary/arterial tortuosity, peripartum state, recent episode of extreme emotion or exertion, and connective tissue disorders (1,19,20). Among women with SCAD, up to 18% of dissections are associated with the peripartum/pregnant state (1). The presence of any EVA in patients with SCAD is high and includes aneurysms, noncoronary dissections and most commonly, FMD. The prevalence of FMD among SCAD patients has been observed as high as 25% to 86% (21–24).

Discriminating SCAD from atherosclerotic disease in the acute setting is particularly important given the markedly elevated risks of percutaneous coronary intervention with SCAD (1,25). In patients presenting with definite MI, SCAD is most often diagnosed by characteristic findings on CA. Selective utilization of adjunctive imaging techniques, specifically intravascular imaging, may increase diagnostic sensitivity (26). In uncertain cases of SCAD, such as those patients with a borderline troponin elevation and nonspecific findings on electrocardiogram, other imaging modalities such as coronary computed tomography angiography (CTA), echocardiography, myocardial perfusion imaging (MPI), or cardiac magnetic resonance (CMR) may suggest myocardial ischemia or infarction, which may then lead to CA and accurate diagnosis. Examples of this include regional wall motion abnormalities on echocardiography, decreased myocardial perfusion on coronary CTA, or regional endocardial or late gadolinium enhancement on CMR.

In light of the fact that SCAD vessels can heal with conservative management alone (1,19,27,28) in contrast to the approach to those with atherosclerotic obstruction, interventionalists are increasingly taking a “less is more” approach. Thus, percutaneous coronary intervention is avoided in stable SCAD patients who have preserved flow, even in the presence of significant obstruction or infarction (27). However, percutaneous coronary intervention or coronary artery bypass graft surgery remains appropriate for those patients who are clinically unstable or demonstrate compromised coronary blood flow, even if pregnant (**Figure 1**) (27). There are insufficient data to favor one revascularization technique over another, so decisions should be made based on individual patient characteristics and availability of surgical and interventional expertise.

The rate and frequency of healing and SCAD extension is not yet known. However, an important subset of patients initially treated conservatively may experience clinically significant SCAD progression

ABBREVIATIONS AND ACRONYMS

CA = coronary angiography

CMR = cardiac magnetic resonance

CTA = computed tomography angiography

EVA = extracoronary vascular abnormalities

FMD = fibromuscular dysplasia

IVUS = intravascular ultrasound

MPI = myocardial perfusion imaging

OCT = optical coherence tomography

PET = positron emission tomography

SCAD = spontaneous coronary artery dissection