JACC: CARDIOVASCULAR INTERVENTIONS VOL. 6, NO. 1, 2013

ISSN 1936-8798/$36.00
PUBLISHED BY ELSEVIER INC. http://dx.doi.org/10.1016/j.jcin.2012.08.017

Spontaneous Coronary Artery Dissection

Prevalence of Predisposing Conditions Including Fibromuscular Dysplasia in a Tertiary Center Cohort

Jacqueline Saw, MD,* Donald Ricci, MD,* Andrew Starovoytov, MD,* Rebecca Fox, MSc,* Christopher E. Buller, MD†

Vancouver, British Columbia, and Toronto, Ontario, Canada

Objectives We sought to evaluate the prevalence of fibromuscular dysplasia (FMD) and other predisposing conditions among spontaneous coronary artery dissection (SCAD) patients.

Background Spontaneous coronary artery dissection is considered rare. However, we observed many young women with SCAD and concomitant FMD.

Methods Spontaneous coronary artery dissection patients were identified prospectively and retrospectively at Vancouver General Hospital over the past 6 years. Coronary angiograms were meticulously reviewed by 2 senior interventional cardiologists. Identified patients were contacted for prospective evaluation at our SCAD clinic, and screening for FMD of renal, iliac, and cerebrovascular arteries was performed with computed tomography angiography or magnetic resonance angiography, if not already screened during the index angiogram. Potential predisposing and precipitating conditions for SCAD were extracted from clinical history.

Results We identified 50 patients with nonatherosclerotic SCAD from April 2006 to March 2012. Average age was 51.0 years, and almost all were women (98.0%). All presented with myocardial infarction (MI), 30.0% had ST-segment elevation, and 70.0% had non–ST-segment elevation MI. Only 1 was postpartum, and 2 were involved in intense isometric exercises. Emotional stress was reported in 26.0% before the MI. Twelve percent had >1 dissected coronary artery. Most SCAD patients had FMD of ≥1 noncoronary territory (86.0%): 25 of 43 (58.1%) renal, 21 of 43 (48.8%) iliac, and 20 of 43 (46.5%) cerebrovascular (6 of 43, 14.0% had intracranial aneurysm). Five had incomplete FMD screening.

Conclusions Nonatherosclerotic SCAD predominantly affects women, and most have concomitant FMD. We suspect these patients have underlying coronary FMD that predisposed them to SCAD, but this requires proof from histology or intracoronary imaging of the affected coronary arteries. (J Am Coll Cardiol Intv 2013;6:44–52) © 2013 by the American College of Cardiology Foundation

From the *University of British Columbia, Vancouver General Hospital, Vancouver, British Columbia, Canada and †St. Michael's Hospital, Toronto, Ontario, Canada. The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

Manuscript received July 10, 2012; revised manuscript received August 20, 2012, accepted August 22, 2012.

Spontaneous coronary artery dissection (SCAD) has been reported to be a rare cause of acute coronary syndrome (ACS) and sudden cardiac death (SCD). The first autopsy report involved a 42-year-old woman with SCD in 1931 (1). Subsequently a pooled analysis of Medline published cases from 1931 to 2008 reported 440 cases of SCAD (2). We have previously suggested, however, that SCAD is underdiagnosed and might be more prevalent than previously recognized. Difficulty distinguishing dissection from atherosclerotic obstruction combined with a low clinical index of suspicion might account for under-diagnosis. Our single center retrospective review of young women (age ≤50 years) undergoing coronary angiography after myocardial infarction (MI) revealed that one-quarter had SCAD as the underlying mechanism (3).

Spontaneous coronary artery dissection is defined as a separation of the coronary arterial wall by hemorrhage, with or without an associated intimal tear. There are 2 proposed patterns of SCAD, the first initiated by an intimal tear that leads to propagating medial dissection, and the second by a dissecting medial hematoma, perhaps initiated by rupture of vaso vasorum (4). The former etiology might be visualized as multiple lumens (true and false), intimal flap, or slow clearing of contrast dye on angiograms. The latter might manifest as luminal narrowing or occlusion by intramural hematoma compression, thus mimicking stenosis due to atherosclerosis. Adjunctive intracoronary imaging with intravascular ultrasound or optical coherence tomography might be valuable in these instances to diagnose intramural hematoma.

Spontaneous coronary artery dissection is often classified according to the associated predisposing condition and has been broadly divided into atherosclerotic and nonatherosclerotic. Nonatherosclerotic associations have included peripartum state, connective tissue disorders, systemic inflammatory conditions, and coronary artery spasm. Those without identifiable predisposition have been labeled as idiopathic. Intense exercises with increased cardiocirculatory stresses and shear forces against the coronary arterial wall can precipitate SCAD among patients with or without these predisposing conditions.

Our group was the first to describe an association between fibromuscular dysplasia (FMD) and SCAD in a consecutive case series (5). The prevalence of FMD and other predisposing conditions among patients with SCAD is unknown. Thus, we seek to describe the relative prevalence of predisposing associations with SCAD and to evaluate their in-hospital outcomes.

Methods

We sought to identify all patients seen with SCAD at Vancouver General Hospital from April 1, 2006, to March 1, 2012, with both prospective and retrospective methods. Patients prospectively identified in our cardiac catheterization laboratory or referred to our SCAD outpatient clinic from local British Columbia hospitals constituted the primary source. We also searched for the keywords "coronary dissection," "FMD," and fibromuscular dysplasia in our cardiac catheterization database among coronary angiograms performed during the interval of interest to retrospectively identify additional cases. For the purpose of this report, we excluded patients judged to have atherosclerosis as the underlying condition causing SCAD.

The coronary angiograms were reviewed by 2 senior interventional cardiologists (J.S. and D.R.). In the absence of prior coronary intervention or trauma, SCAD was confirmed to be present when 1 of the following 3 criteria were met: 1) characteristic multiple radiolucent lines separating true and false lumens, often with contrast dye hang-up (or staining) (Fig. 1); 2) as previously described by Pate et al. (6), a long obstructive stenosis typically involving the mid to distal vessel with abrupt demarcation from normal proximal segments that did not respond to intracoronary nitroglycerin or that subsequently normalized on repeat angiograms and without atherosclerotic changes in other vessels (Fig. 2); or 3) adjunctive intracoronary imaging (intravascular ultrasound or optical coherence tomography) showing intramural hematoma (Fig. 3). The coronary segment involved with SCAD was defined as per the Bypass Angioplasty Revascularization Investigation classification (7). The number of coronary arteries dissected and the segments involved were recorded. Results from repeat coronary angiography or intracoronary imaging were recorded.

Abbreviations and Acronyms

ACS = acute coronary syndrome
CTA = computed tomography angiography
FMD = fibromuscular dysplasia
MI = myocardial infarction
MRA = magnetic resonance angiography
RCA = right coronary artery
SCAD = spontaneous coronary artery dissection
SCD = sudden cardiac death

Baseline characteristics of patients with confirmed SCAD were extracted from the cardiac catheterization database and from relevant clinical source documents. Conventional cardiovascular risk factors, medication on presentation, hospital presentation, electrocardiogram changes, in-hospital events, and angiographic and noninvasive imaging characteristics were recorded.

Combining extracted data with focused histories obtained in our SCAD clinic, we sought to identify the presence of conditions with previously recognized association with SCAD, including recent pregnancy, inherited connective tissue disorders, systemic inflammatory conditions predisposing to arteritis, and coronary artery spasm. We screened for noncoronary FMD by performing invasive or noninvasive angiography in up to 3 vascular territories: renal, iliac,