Catheterization and Cardiovascular Interventions 84:1115–1122 (2014)

Interventional Rounds

Coronary Angiogram Classification of Spontaneous Coronary Artery Dissection

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Spontaneous coronary artery dissection (SCAD) is under-diagnosed and the true prevalence is underestimated. Unfortunately, SCAD is frequently missed on coronary angiogram since the arterial wall is not imaged with this test. Optical coherence tomography or intravascular ultrasound should be the true gold-standard to diagnose SCAD. Given the elusive angiographic diagnosis of SCAD and the lack of familiarity with angiographic variants of SCAD, a diagnostic algorithm and angiographic classification for SCAD is proposed in this article.

Key words: angiography; coronary; catheterization; diagnostic; IVUS-imaging; intravascular ultrasound; imaging; optical coherence tomography

INTRODUCTION

Spontaneous coronary artery dissection (SCAD) is under-diagnosed and the true prevalence is underestimated. The first SCAD was reported in 1931, and since then <800 cases were published to-date. Understandably, the medical community labeled this condition as rare. Among patients undergoing coronary angiography, SCAD was diagnosed in only 0.2%–1.1% of angiograms; and it was also reported to be a rare cause of acute coronary syndrome (ACS) and sudden cardiac death, accounting for 0.1%–4% and 0.4%, respectively [1]. However, the prevalence is much higher in young women with ACS, accounting for ~25% of women aged ≤50 presenting with myocardial infarction (MI) in our series [2]. Many of these SCAD cases were actually misdiagnosed due to the imperfections of coronary angiography.

SCAD is a spontaneous, non-traumatic, and non-iatrogenic separation of the coronary artery wall by intramural hemorrhage, which can occur with or without an inciting intimal tear. The creation of a false lumen with intramural hematoma (IMH) can propagate antegrade and retrograde, compressing arterial lumen to varying degrees, and causing ischemia or infarction according to the degree of arterial occlusion. There are two proposed mechanisms for SCAD. The first is an intimal tear that leads to hemorrhage into the media, causing medial dissection and creation of a false lumen. This may appear angiographically as the pathognomonic multiple radiolucent lumen, contrast dye stains in the arterial wall, or slow clearing or hang-up of contrast dye in the lumen. The second mechanism is less well known, and is proposed to be initiated by the rupture of vaso vasorum causing spontaneous bleed into arterial wall producing IMH [3]. This may appear only as luminal compression, and this angiographic appearance is more commonly seen than the classic arterial wall stains [4]. Most angiographers are not familiar with this non-classic angiographic appearance of SCAD, contributing to why SCAD is often missed on angiograms. Other reasons include mild stenosis, smooth-walled stenosis, and involvement of distal and small arteries.

Coronary angiography is a 2-dimensional luminogram that does not image arterial walls. Since SCAD

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Conflict of interest: Nothing to report.

Additional Supporting Information may be found in the online version of this article.

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Received 13 August 2013; Revision accepted 10 November 2013

DOI: 10.1002/ccd.25293
Published online 13 November 2013 in Wiley Online Library (wileyonlinelibrary.com)

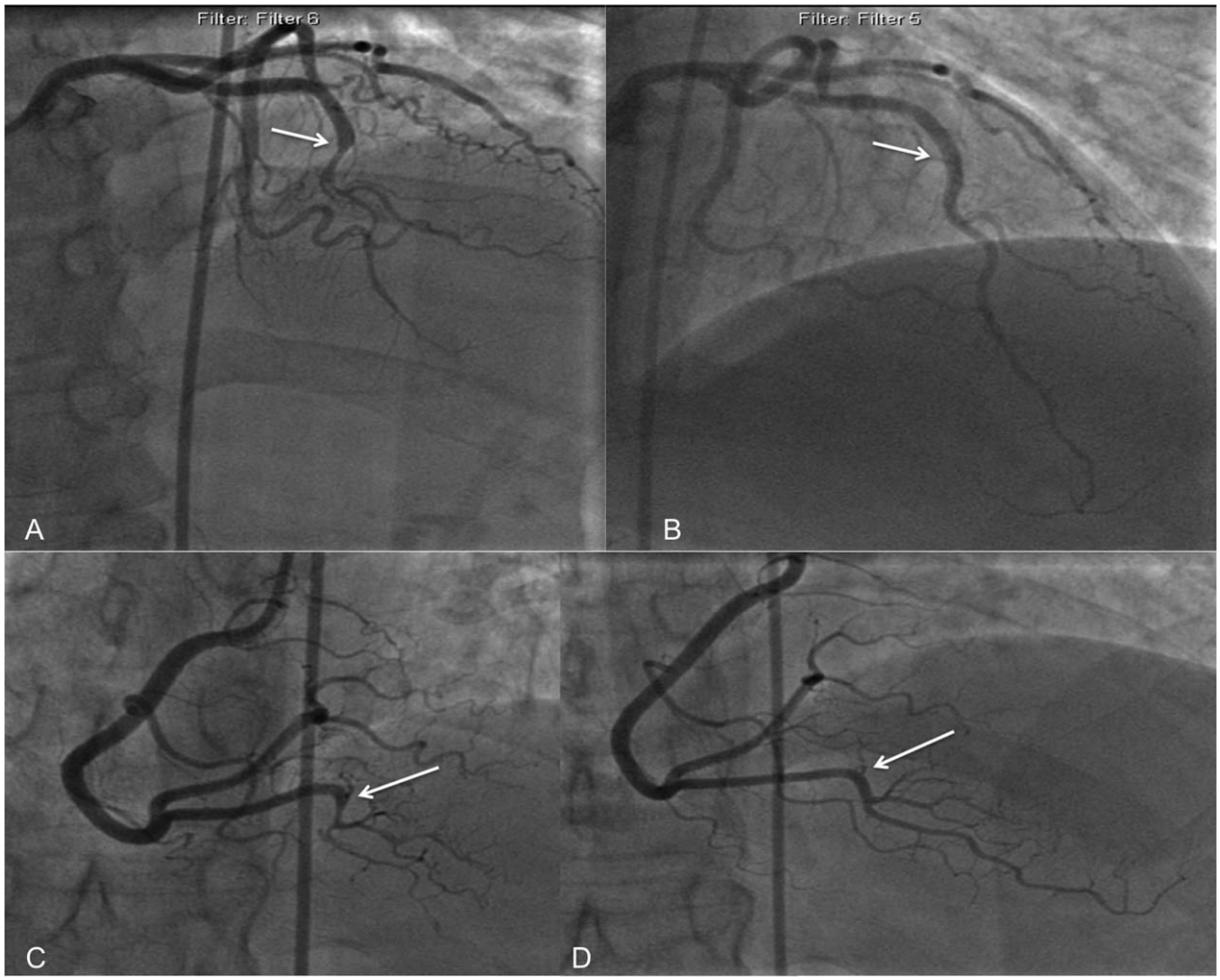

Fig. 1. Type 1 SCAD angiographic appearance: (A) SCAD of the mid left anterior descending (LAD) artery with multiple lumen and contrast staining of the arterial wall (starting from arrow), with complete occlusion of the distal LAD. B: Repeat angiogram 2 months later showing angiographic healing of the LAD. C: SCAD of the posterior descending artery (PDA) of a different patient with multiple radiolucent lumen from contrast staining of arterial wall (starting from the arrow). D: Repeat angiogram showing angiographic healing of the PDA 6 weeks later.

predominantly affects the arterial wall, dedicated intracoronary imaging modality like optical coherence tomography (OCT) and intravascular ultrasound (IVUS) that focus on the arterial wall should be the true gold-standard. In particular, OCT has an unmatched spatial resolution of 10–20 μm, whereas IVUS has lower spatial resolution (∼150 μm) but better penetration. Both these technologies provide complementary details on SCAD imaging [5], with OCT being superior in visualizing intimal tears, intraluminal thrombi, false lumen, and IMH, but due to inadequate optical penetration and shadowing, it may not visualize the full extent of the IMH. On the other hand, IVUS is adequate to visualize IMH and false lumen, but the lumen–intimal interface is not as distinctly delineated as OCT, and thus inferior in visualizing intimal tear. However, IVUS allows more complete and deeper vessel visualization, and thus better appreciation of the extent of IMH. Due to cost constrains, most catheterization laboratories can only utilize one intracoronary technology for SCAD diagnosis. OCT is preferred in such cases because of the superiority in spatial resolution and clarity in diagnosing the presence of IMH and intimal tear. Furthermore, OCT is superior in visualizing malapposed stent struts and thus optimizing stent results if intervention is pursued. Although IVUS allows better appreciation of the depth and bulk of IMH, this detail is not necessary in the diagnosis or management of SCAD.

Cardiac CT angiography (CCTA) has gained important strides in evaluating atherosclerotic stenosis;

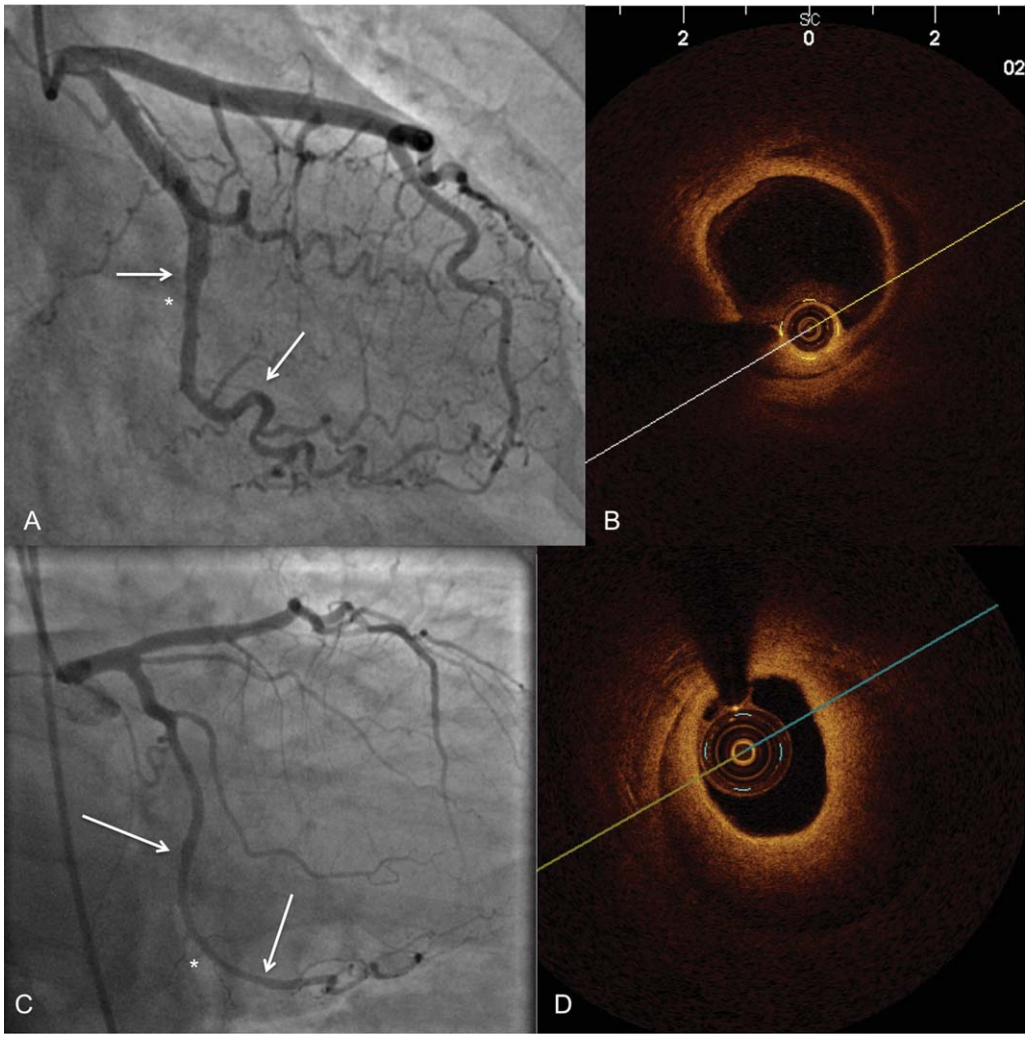

Fig. 2. Type 2 SCAD angiographic appearance with diffuse mild stenosis: (A) SCAD of the mid circumflex artery with diffuse mild stenosis only (delineated by the arrows), with sharp demarcation in the proximal segment of the stenosis from "normal" to "abnormal" segment (Supporting Information Video 2A). B: Corresponding OCT of the mid circumflex artery (*) showing IMH. C: SCAD of the mid to distal circumflex artery of a different patient, with diffuse smooth stenosis (between the arrows) (Supporting Information Video 2C). D: Corresponding OCT of distal circumflex (*) artery showing IMH.

however, it has limited use in the diagnosis of SCAD due to lower spatial resolution compared to conventional angiography [1]. A large proportion of SCAD affect small side-branches and distal coronary arteries [6] that are not well visualized with CCTA. Moreover, contrast staining of arterial wall may not be visualized in the absence of intimal tear, and subtle diffuse lumen compression by IMH may not be detected by CCTA.

Early angiographic diagnosis of SCAD is pertinent as it may preempt the use of potentially harmful therapy such as fibrinolysis [1], or unnecessary percutaneous coronary intervention (PCI) with either angioplasty or stenting. The decision to revascularize with PCI or coronary artery bypass surgery should depend on the clinical status, hemodynamic stability, and angiographic characteristics. A conservative approach is typically favored [1,7,8] unless patients have ongoing ischemia, recurrent chest pains, hemodynamic instability, or left main dissection. Such a prudent approach is recommended in stable SCAD patients as their natural history appears favorable, with the vast majority of patients with repeat angiograms demonstrating spontaneous arterial healing [6,8]. Conversely, the challenges with PCI for SCAD have been described, in particular, inability to gain true lumen access, extension of dissection/IMH, need for multiple and long stents, and subsequent restenotic and stent thrombosis risks [1]. In fact, the Mayo Clinic series documented a technical success

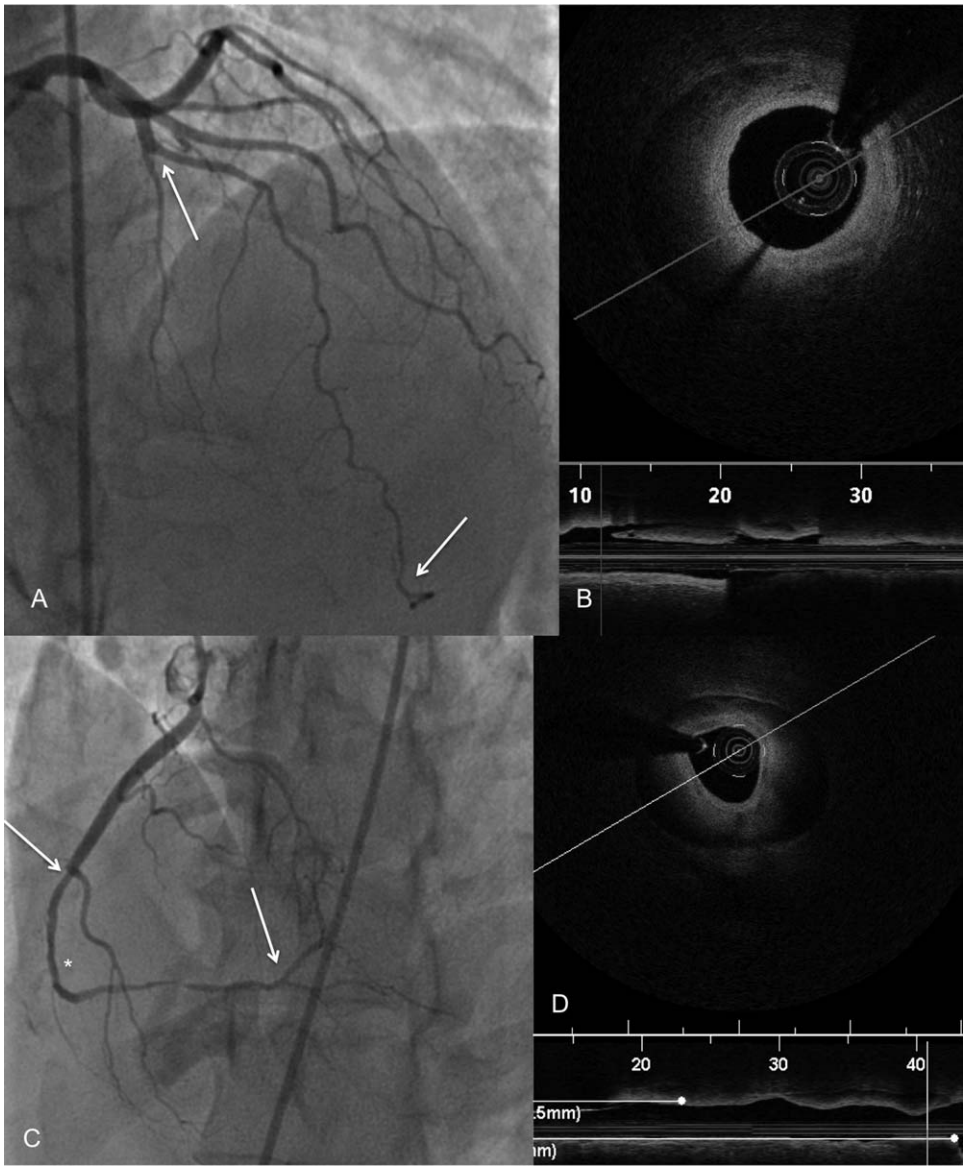

Fig. 3. Type 2 SCAD angiographic appearance of moderate to severe diffuse stenosis: (A) Diffuse and smooth moderate stenosis of the mid to apical LAD (between the arrows) due to SCAD (Supporting Information Video 3A). B: Corresponding OCT image of LAD showing IMH. C: Diffuse moderate to severe stenosis of the right coronary artery (RCA) of a different patient, from mid RCA to the PDA (between the arrows), with varying stenosis severity due to SCAD (Supporting Information Video 3C). D: Corresponding OCT of the RCA (*) showing IMH.

rate of only 65% with PCI; furthermore, ~25% of successful PCI had propagation of the dissection requiring additional stents [9]. Therefore, accurate diagnosis of SCAD is germane as the management of SCAD is different from atherosclerotic lesions. If PCI is necessary, bioabsorbable stents have theoretical advantage as the struts are eventually resorbed, rendering potentially obsolete the long-term issues of malapposition (that occurs with resorption of IMH) and extensive stent lengths.

ANGIOGRAPHIC VARIANTS OF SCAD APPEARANCE

There are three distinct angiographic appearances and patterns of SCAD that can be characterized:

1. Type 1 (evident arterial wall stain): This is the pathognomonic angiographic appearance of SCAD with contrast dye staining of arterial wall with multiple radiolucent lumen (Fig. 1).

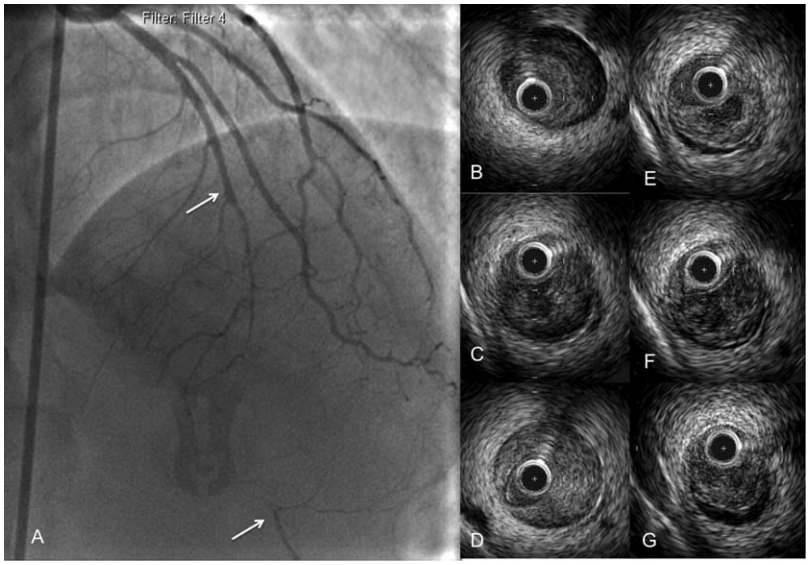

Fig. 4. Type 2 angiographic SCAD: (A) Diffuse stenosis and occlusion of the mid to apical LAD due to SCAD (between the arrows) (Supporting Information Video 4A). B–G: Corresponding IVUS images showing the marked variability in echogenicity of the IMH depending on blood flow and thrombosis of false lumen at different segments of the LAD: (B) hypoechogenic crescent-shaped IMH surrounding true lumen, (C) IMH of varying hyperechogenicity, (D) hyperechogenic false lumen from IMH, (E) IMH with peripheral hyperechogenicity and central hypoechogenicity, (F) IMH with inhomogenous echogenicity, (G) outer circumference hypoechogenicity of intramural blood, and more central hyperechogenic hematoma.

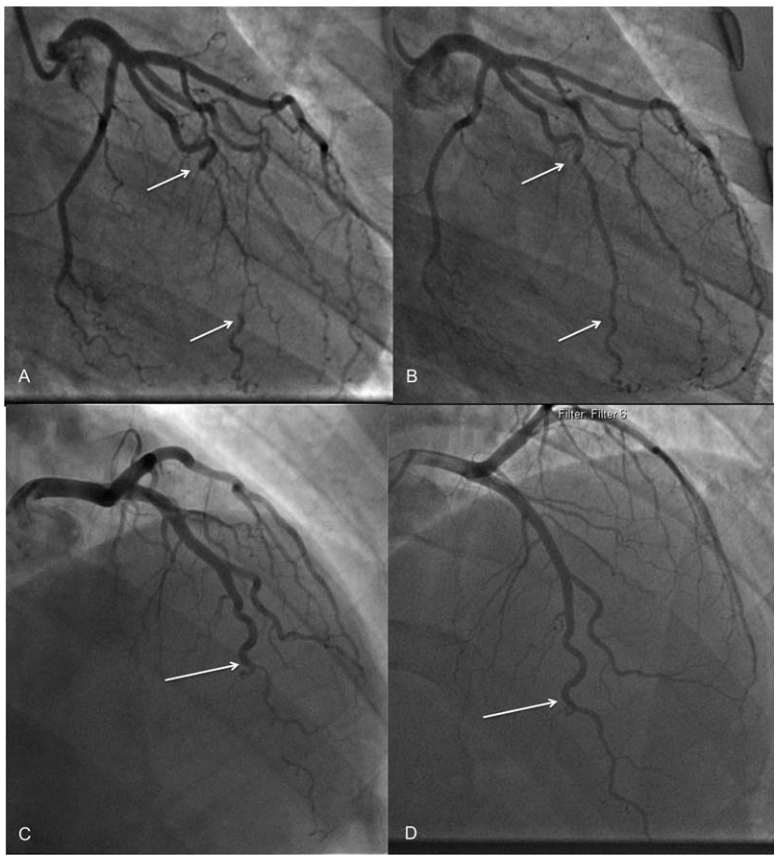

Fig. 5. Type 2 angiographic SCAD with repeat angiograms showing healing: (A) SCAD of the ramus intermedius artery with diffuse severe stenosis from mid to distal segment in a different patient (between arrows) (Supporting Information Video 5A). B: Repeat angiogram 1 year later showing angiographic healing (Supporting Information Video 5B). C: SCAD of the mid to apical LAD starting from the arrow with severe diffuse stenosis in another patient. D: Repeat coronary angiogram 3 months later showing angiographic healing.

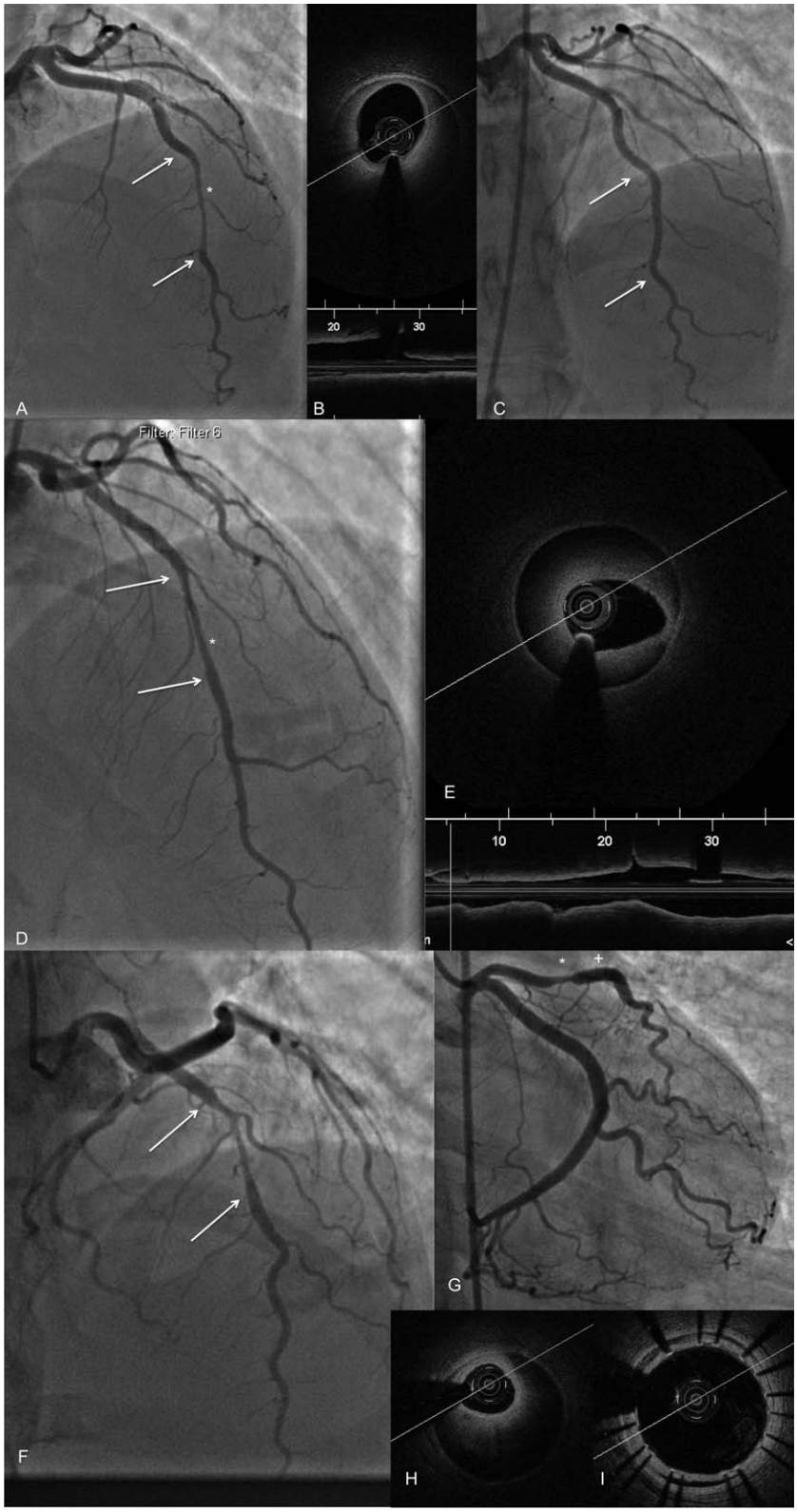

Fig. 6. Type 3 angiographic SCAD mimicking atherosclerosis: (A) Long smooth mid LAD stenosis due to SCAD (between the arrows), mimicking atherosclerotic lesion. B: Corresponding OCT image of mid LAD (*) showing IMH. C: Repeat coronary angiogram 17 months later showing angiographic healing. In another patient (D) long smooth stenosis of the mid LAD due to SCAD (between the arrows), mimicking atherosclerotic lesion. E: Corresponding OCT image of the mid LAD (*) showing IMH. In a different patient (F) long mid LAD hazy and smooth stenosis due to SCAD that mimicked atherosclerosis (between arrows). G: Repeat angiogram 1 week later for chest pain showed severe stenosis (*) proximal to the stent (+). H: OCT of (*) showed IMH. I: OCT of the stented segment (+) showed residual "tacked-up" IMH from SCAD a week prior.

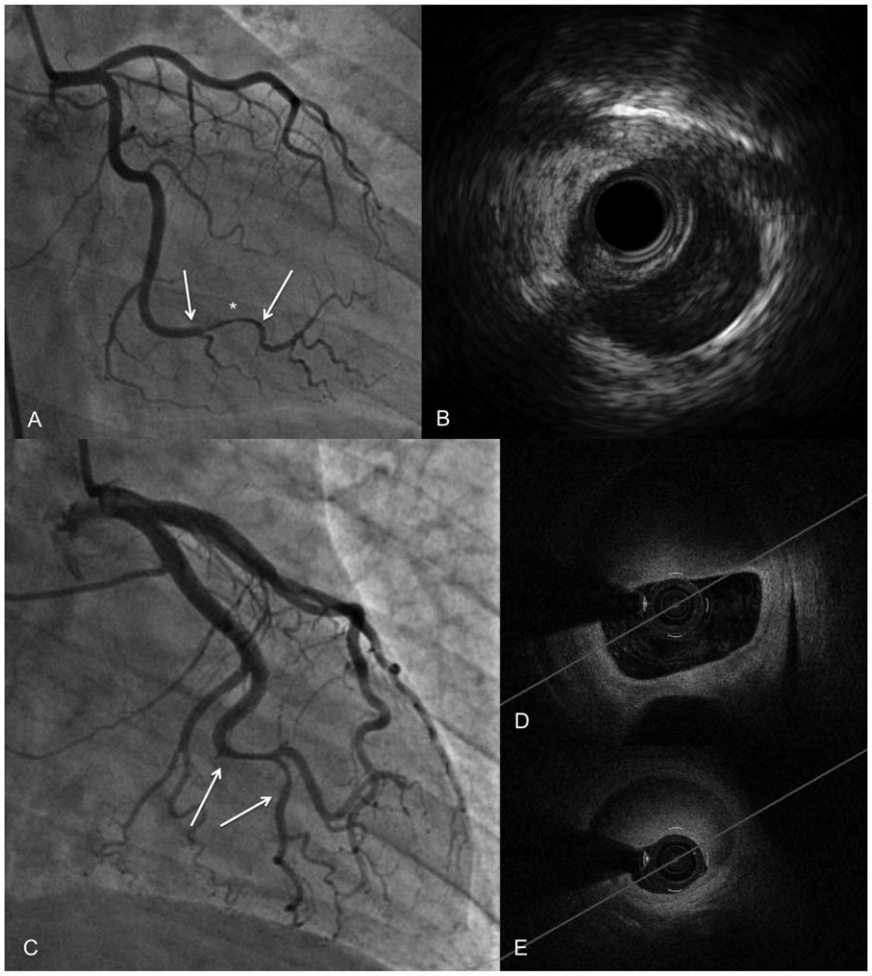

Fig. 7. Type 3 angiographic SCAD mimicking atherosclerosis: (A) SCAD of the obtuse marginal artery with relatively linear stenosis (between the arrows). B: Corresponding IVUS image of obtuse marginal artery (*) showing large crescent-shaped hypoechogenicity of IMH. In another patient (C) SCAD of the obtuse marginal artery showing linear stenosis between the arrows. D: Corresponding OCT image of the obtuse marginal artery in the proximal segment of the dissection, showing a linear compression of the arterial lumen by IMH, giving the appearance of linear stenosis on coronary angiogram. E: OCT of the obtuse marginal artery more distally showing crescent-shaped IMH compressing the arterial lumen.

TABLE I. Clinical Features That Raise of Suspicion for SCAD in MI Patients

Features that raise a high clinical index of suspicion for SCAD
1. Myocardial infarction in young women (especially age $\leq$50)
2. Absence of traditional cardiovascular risk factors
3. Little or no evidence of typical atherosclerotic lesions in coronary arteries
3. Peripartum state
4. History of fibromuscular dysplasia
5. History of relevant connective tissue disorder or systemic inflammatory condition (e.g. Marfan's syndrome, Ehler Danlos syndrome Type 4, Loeys-Dietz syndrome, cystic medial necrosis, systemic lupus erythematosus, Crohn's disease, ulcerative colitis, polyarteritis nodosa, sarcoidosis, Churg-Strauss syndrome, Wegener's granulomatosis, rheumatoid arthritis, giant cell arteritis)
6. Recent intensive exercise or emotional stress

2. Type 2 (diffuse stenosis of varying severity): This angiographic appearance is not well appreciated and is often missed or misdiagnosed. SCAD commonly involves the mid to distal segments of coronary arteries, and can be so extensive that it reaches the distal tip. There is an appreciable (often subtle) abrupt change in arterial caliber, with demarcation from normal diameter to diffuse narrowing. This

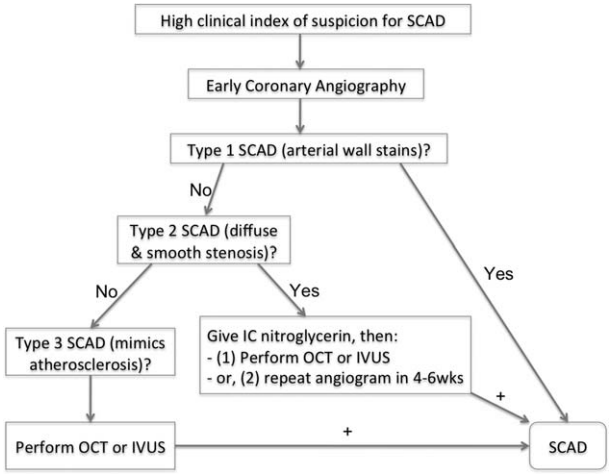

Fig. 8. Simple algorithm for the angiographic diagnosis and confirmation of SCAD.

diffuse (typically >20 mm) and usually smooth narrowing can vary in severity from an inconspicuous mild stenosis to complete occlusion (Figs. 2–5).

3. Type 3 (mimic atherosclerosis): This appearance is the most challenging to differentiate from atherosclerosis (Figs. 6 and 7) and most likely to be misdiagnosed. Angiographic features that favor SCAD are: (a) lack of atherosclerotic changes in other coronary arteries, (b) long lesions (11–20 mm), (c) hazy stenosis, and (d) linear stenosis. Angiographer needs to have a high index of suspicion for SCAD (Table I) and liberally use intracoronary imaging for such cases.

DIAGNOSING SCAD

A stepwise algorithm to diagnose non-atherosclerotic SCAD is proposed (Fig. 8). Clinicians should have a high index of suspicion for SCAD especially in young women presenting with MI without traditional cardiovascular risk factors (Table I). An early invasive coronary angiography for these patients is recommended. If type 1 SCAD appearance is evident, then there is little controversy and the diagnosis of SCAD can be made. Angiographers should then assess for the presence of atherosclerotic changes in other coronary arteries, and utilize intracoronary imaging if there is uncertainty as to non-atherosclerotic SCAD. If type 1 SCAD appearance is not evident, SCAD diagnosis would be most objectively confirmed by OCT or IVUS. For diffuse (>20 mm) and smooth stenosis (type 2 SCAD), intracoronary nitroglycerin should be administered to relieve potential overlying spasm. If the lesion remains after nitroglycerin, then OCT or IVUS should be pursued. If there are concerns of compromising arterial flow with intracoronary imaging, then repeat angiogram >4 weeks later should be pursued to reassess the stenosis, as SCAD typically resolves spontaneously. If the appearance mimics atherosclerosis (type 3 SCAD) and there is a high clinical index suspicion for SCAD (Table I), then OCT or IVUS should be pursued. This proposed simple algorithm will hopefully help improve the diagnosis of SCAD.

CONCLUSIONS

In summary, the diagnosis of SCAD on coronary angiography is elusive. Most angiographers are not familiar with angiographic variants of SCAD. The use of intracoronary imaging (OCT or IVUS) for non-classic angiographic forms of SCAD is important. The proposed simple algorithm to investigate SCAD will hopefully help improve the diagnosis of SCAD.

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