

Role of Coronary CT Angiography in Spontaneous Coronary Artery Dissection

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Conflicts of interest are listed at the end of this article.

Radiology: Cardiothoracic Imaging 2020; 2(6):e200364 • <https://doi.org/10.1148/ryct.2020200364> • Content codes: **CA** **CT**

Spontaneous coronary artery dissection (SCAD) is more common than previously thought and is present in up to 4% of patients presenting with acute coronary syndrome. SCAD predominantly occurs in relatively young women and is an important cause of myocardial infarction in young patients without traditional risk factors of atherosclerotic coronary artery disease. There have been substantial improvements in spatial and temporal resolution and reduction in ionizing radiation dose with new generation scanners. The risk of dissection propagation with an invasive coronary angiogram, improved CT scanner parameters, and predominantly conservative management of SCAD make coronary CT angiography a useful noninvasive imaging modality for the assessment of SCAD.

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Spontaneous coronary artery dissection (SCAD) is defined as a noniatrogenic dissection of an epicardial coronary artery not associated with atherosclerosis or trauma (1). SCAD can be a cause of sudden cardiac arrest and myocardial infarction in young persons without traditional risk factors of atherosclerotic coronary artery disease (CAD) (2,3). Even though SCAD was first reported at autopsy by Pretty in 1931 (4), it has remained poorly understood and, although once considered a rare disease, is often misdiagnosed and underdiagnosed.

Invasive coronary angiography (ICA) has traditionally been used for the assessment of patients with suspicion of SCAD and is often the initial diagnostic modality. Despite its high spatial resolution, ICA only visualizes the lumen of coronary vessels and has limitations in detecting SCAD. Recently, there has been increasing consideration of noninvasive coronary CT angiography as an alternative imaging modality for stable patients suspected of having SCAD. The use of coronary CT angiography has been fostered by technological advancements allowing improvement in spatial and temporal resolution and increased recognition and awareness of the risk of ICA. In addition, coronary CT angiography is an appealing option because most patients with SCAD are treated conservatively. This review highlights the role of coronary CT angiography in the diagnosis of SCAD and summarizes the importance of extracoronary vascular imaging and other imaging modalities used in the assessment of SCAD. The goal of this review is to raise awareness of this important diagnosis to imagers who may encounter SCAD at coronary CT angiography.

Epidemiology and Pathophysiology

The true prevalence of SCAD remains unknown, as the condition is often misdiagnosed and underdiagnosed. More recent studies estimate the prevalence of SCAD to be 1%–4% of patients presenting with acute coronary

syndrome (1,5). SCAD is a common cause of myocardial infarction in pregnancy and in young and middle-aged women (6). Up to 43% of pregnancy-associated myocardial infarction (7) and 35% of myocardial infarction in women younger than 50 years is attributed to SCAD (5,6,8).

SCAD is characterized by spontaneous formation of an intramural hematoma within the wall of the coronary artery either due to tear in the intima or rupture of vasa vasorum (9,10). Formation of this hematoma results in compression of the true lumen and obstruction of blood flow through the coronary artery, leading to myocardial ischemia and infarction (1). There can be thrombus formation within the true or false lumen, which contributes to the pathophysiologic features of acute coronary syndrome due to SCAD (9,11). Coronary arteries that have weakened arterial wall architecture are commonly affected by SCAD (12).

The etiology of SCAD is multifactorial, and predisposing factors include arteriopathies, environmental precipitators, or stressors (1). Fibromuscular dysplasia is the most common coexisting condition (13) and can be present in up to 86% of cases of SCAD (1,14). Fibromuscular dysplasia is a nonatherosclerotic non-inflammatory vascular disease that is more common in middle-aged women and typically involves renal and carotid arteries, although any medium-sized artery can be involved (15,16). Inherited arteriopathies and connective tissue disorders, including Ehlers-Danlos syndrome type IV, pseudoxanthoma elasticum, Marfan syndrome, and Loeys-Dietz syndrome, are also associated with SCAD in fewer than 5% of cases (14,17). Hormonal influences during pregnancy, infertility treatment, and use of oral contraceptives can lead to weakened arterial media (18). Pregnancy-induced hemodynamic changes may increase shear stress within the arterial wall, predisposing to

Abbreviations

CAD = coronary artery disease, ICA = invasive coronary angiography, SCAD = spontaneous coronary artery dissection

Summary

Coronary CT angiography is a useful noninvasive imaging modality for the initial assessment and follow-up of spontaneous coronary artery dissection.

Essentials

- Coronary CT angiography findings in spontaneous coronary artery dissection (SCAD) include (a) lack of atherosclerotic plaque, (b) tapered luminal stenosis, (c) abrupt luminal stenosis, (d) luminal occlusion, (e) intramural hematoma, (f) dissection flap, and (g) perivascular epicardial fat stranding.
- SCAD-associated arteriopathies can be concurrently assessed with CT angiography.
- Multimodality imaging can complement coronary CT angiography in the assessment of SCAD.
- Coronary CT angiography has adequate resolution in the proximal and middle portions of most coronary vessels, but excluding dissection in small distal vessels is limited.

SCAD (2). Intense Valsalva-like activities (vomiting, coughing, or childbirth), physical effort, emotional stress, and sympathomimetic drugs may also precipitate SCAD (1).

Imaging of SCAD

Echocardiography

Echocardiography is a readily available noninvasive imaging technique that is typically used for the initial evaluation of left ventricular function and regional wall motion abnormalities due to ischemia or infarction in patients with acute SCAD. Echocardiography may be helpful in subsequent serial imaging in SCAD to monitor left ventricular functional recovery, as it does not expose the patient to ionizing radiation (1). Given that SCAD typically affects young and previously healthy individuals, elevated troponin level may be mistakenly explained by diagnoses such as myocarditis or pericarditis. The presence of a focal wall motion abnormality at echocardiography in the setting of chest pain is helpful in raising suspicion for coronary causes including SCAD, coronary embolism, and premature atherosclerosis.

Nuclear Myocardial Perfusion Imaging

Myocardial perfusion imaging with SPECT or PET is not the initial imaging modality used for the assessment of SCAD. These modalities may prove useful in risk stratification of asymptomatic patients with SCAD (19), assessment of ischemia in patients presenting with recurrent chest pain, and assessment of myocardial viability prior to revascularization if required (20).

ICA with Intracoronary Imaging

The classic appearance of SCAD at ICA is the presence of multiple radiolucent lumens and extraluminal contrast mate-

rial staining (21,22), which are classified as type 1 SCAD and are observed in only a small proportion of cases (21). Other appearances of SCAD at ICA include diffuse stenosis of the coronary artery usually more than 20 mm in length (type 2) and focal or tubular stenosis usually less than 20 mm in length (type 3). The mean length of dissection has been reported to be more than 45 mm at ICA (9). Similar to coronary CT angiography, the most common appearance of SCAD at ICA is type 2 seen in up to 67.5% of dissected arteries, while type 1 and type 3 SCAD were seen in 29.1% and 3.4% of dissected coronary arteries, respectively (23). As the classic appearance of SCAD with multiple lumens and contrast staining of the arterial wall is present in fewer than one-third of cases, there is a potential for the diagnosis to be missed. Moreover, type 3 SCAD at ICA is similar in appearance to atherosclerotic CAD. Identification of intimomedial disruption and intramural hematoma with intravascular US and optical coherence tomography facilitates the diagnosis of angiographically ambiguous SCAD cases (24). Another type of SCAD, type 4, has been described in literature, though not routinely used in clinical practice. Type 4 lesions are associated with complete, usually distal, vessel occlusion (25). ICA is the currently recommended imaging modality of choice for patients with SCAD manifesting as acute coronary syndrome (1).

Cardiac MRI

Cardiac MRI is a noninvasive test with no risk of ionizing radiation, which can accurately assess left ventricular function, regional wall motion abnormalities, myocardial perfusion, and areas of infarction or scarring. Cardiac MRI is not the initial modality for assessment of patients with SCAD but can provide value in clarifying diagnosis in ambiguous cases and assessment of adverse sequelae, such as microvascular obstruction after acute SCAD (26). Cardiac MRI may also be useful in the assessment of ischemia in patients presenting with recurrent chest pain and assessment of myocardial viability.

Coronary CT Angiography

Technique.—Coronary CT angiography has an established role in noninvasive assessment of atherosclerotic CAD, with sensitivity up to 97% and specificity up to 78% when compared with ICA (27). There have been recent improvements in CT technology, including (a) improved temporal resolution due to the introduction of dual-source CT and whole-heart imaging with wide detector array CT, (b) improved spatial resolution, (c) ability to scan patients with high heart rate or arrhythmia, and (d) reduction in radiation dose (28–30). Such improvements make coronary CT angiography a useful noninvasive modality for imaging in SCAD.

Better spatial and temporal resolution has dramatically improved our ability to evaluate major epicardial vessels; however, the evaluation of small distal coronary arteries remains beyond current CT resolution. At our institution, multiphase coronary CT angiography using prospectively electrocardiographically

Table 1: Examples of Coronary CT Angiography Literature on SCAD

Reference	No. of Patients with SCAD	Comments
Sun et al (50), 2015	36 (34 controls)	CCTA performed better than ICA in SCAD detection in a retrospective review by four independent observers
Roura et al (47), 2016	34 (24 with CCTA)	CCTA at 3–6 months showed complete healing of SCAD in 83%, with excellent prognosis at long-term (20-month) follow-up
Hedgire et al (37), 2018	2*	Perivascular epicardial fat stranding helpful in identification of atherosclerotic CAD culprit lesions, SCAD, and patients with increased risk of ACS.
Eleid et al (51), 2018	3	SCAD may be missed on CCTA in patients presenting with signs and symptoms of ACS
Pozo-Osinalde et al (34), 2019	11	CCTA able to identify presence of SCAD in an acute setting. Distinct CCTA features include diffuse luminal narrowing with a “sleeve-like” wall thickening (most common), “plaque-like” appearance, dissection flap within the coronary lumen, and total occlusion
Capretti et al (52), 2019	3	Case series demonstrates SCAD not detected at ICA subsequently seen at CCTA.
Tweet et al (33), 2019	14	Four pertinent CCTA features of SCAD included abrupt luminal stenosis (most common), intramural hematoma, tapered luminal stenosis, and dissection†

Note.—ACS = acute coronary syndrome, CAD = coronary artery disease, CCTA = coronary CT angiography, ICA = invasive coronary angiography, SCAD = spontaneous coronary artery dissection.
 * There were a total of 11 patients with perivascular fat stranding and 208 controls without perivascular fat stranding.
 † Other findings from this study included epicardial fat stranding, coronary tortuosity, and coronary bridge. Coronary CT angiography appearance of intramural hematoma was similar to that of noncalcified atherosclerotic plaque.

triggered or retrospectively electrocardiographically gated imaging is performed for patients at low-to-intermediate risk who are referred from the emergency department with suspicion for acute coronary syndrome or who are suspected of having wall motion abnormality (28). Coronary CT angiography in patients at high risk is only performed if deemed necessary after initial cardiology consultation (31). Coronary CT angiography enables reconstruction of different images from the axial (source) data set, including multiplanar reformats, maximum intensity projections, minimum intensity projections, curved planar reformats, and volume-rendered three-dimensional images. Minimum intensity projection images aid the detection of myocardial hypoperfusion. Concurrent static perfusion image analysis can be performed in all coronary CT angiographic studies as detailed in a recent expert consensus document published by the Society of Cardiovascular Computed Tomography (32). Multiphase image acquisition allows assessment of myocardial function and regional wall motion abnormality and also mitigates the effects of motion artifact in the assessment of coronary arteries (28).

Coronary CT angiography findings in SCAD.—The value of coronary CT angiography in identifying acute SCAD has been shown by multiple case series and case reports (Table 1) (33,34). Various cardiac and extracardiac CT angiography features of SCAD are shown in schematic illustration (Fig 1) and discussed in detail below.

Coronary CT angiography findings in SCAD (33,34) include (a) lack of atherosclerotic plaque; (b) tapered luminal stenosis (Table 2; Figs 1, A, 2a, 3); (c) abrupt luminal stenosis (Table 2; Figs 1, B, and 4a), which is described as a sharp demarcation between contrast-opacified patent coronary artery lumen and distal luminal narrowing; (d) luminal occlusion (Table 2; Figs 1, C, and 5); (e) intramural hematoma (Table 2; Figs 1, D, and 6) with hemorrhage within the wall of the coronary artery (35,36); (f) dissection flap (Table 2; Figs 1, E, and 7); and (g) perivascular epicardial fat stranding (37) (Figs 1, F, 3, 4a). The presence of myocardial hypoperfusion (Fig 5b) in a corresponding vascular territory at coronary CT angiography can also improve confidence in diagnosing SCAD; however, such finding is not specific to SCAD and can also be seen in other acute

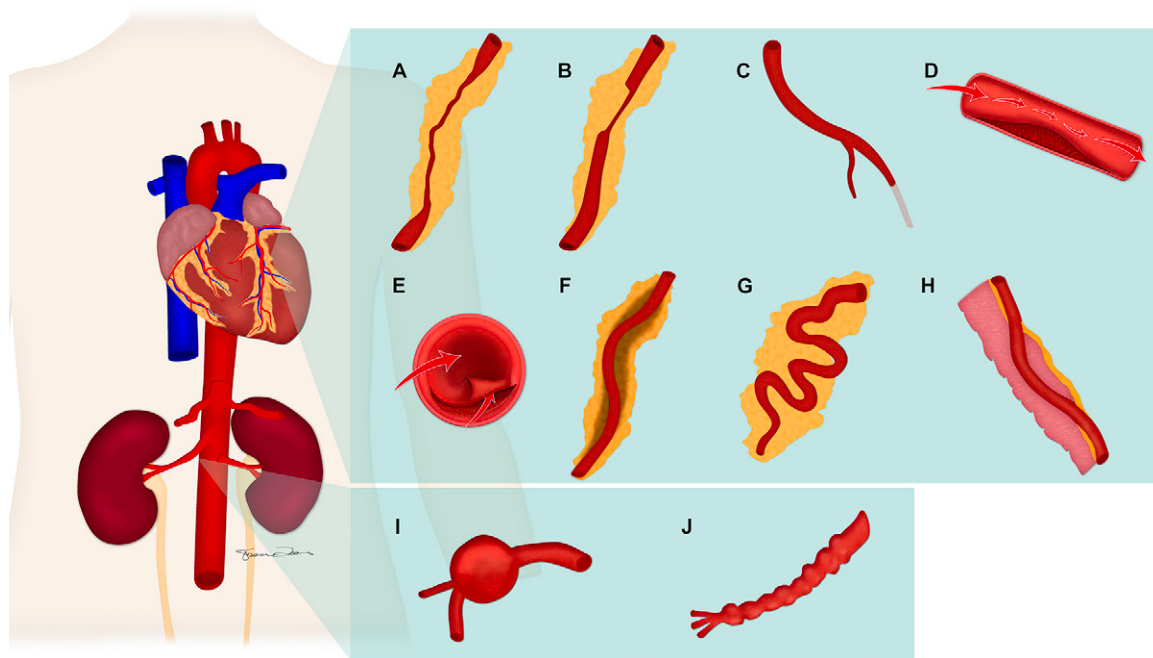


Figure 1: Schematic illustration demonstrates cardiac and extracardiac CT features of spontaneous coronary artery dissection (SCAD). Coronary artery imaging features of SCAD include, A, tapered luminal stenosis, B, abrupt luminal narrowing, C, luminal occlusion, D, intramural hematoma resulting in luminal stenosis, E, dissection flap, F, perivascular fat stranding, G, tortuosity of the coronary artery, and, H, myocardial bridging. Associated fibromuscular dysplasia demonstrates, I, aneurysm and, J, beading of the renal or other visceral arteries.

Table 2: Coronary CT Angiography Findings in SCAD and Mimics

CCTA Finding	Mimics	CCTA Features Indicating SCAD
Tapered luminal stenosis	Coronary vasospasm Noncalcified atherosclerotic plaque	Intramural hematoma Perivascular fat stranding Absence of atherosclerotic CAD in other coronary arteries
Abrupt luminal stenosis	Motion artifact Coronary vasospasm Noncalcified atherosclerotic plaque	Persistence on multiple phases Concurrent wall motion abnormality Concurrent myocardial perfusion defect Intramural hematoma Perivascular fat stranding Absence of atherosclerotic CAD in other coronary arteries
Luminal occlusion	Motion artifact Coronary embolism	Persistence on multiple phases Concurrent wall motion abnormality Concurrent myocardial perfusion defect Intramural hematoma Perivascular fat stranding Involvement of single coronary artery
Intramural hematoma	Noncalcified atherosclerotic plaque	Absence of atherosclerotic CAD in other coronary arteries High attenuation (~50 HU) on non-contrast material-enhanced CT scan
Dissection flap	Motion artifact Slab artifact	Persistence on multiple phases Concurrent wall motion abnormality Concurrent myocardial perfusion defect Intramural hematoma Perivascular fat stranding

Note.—CAD = coronary artery disease, CCTA = coronary CT angiography, SCAD = spontaneous coronary artery dissection.

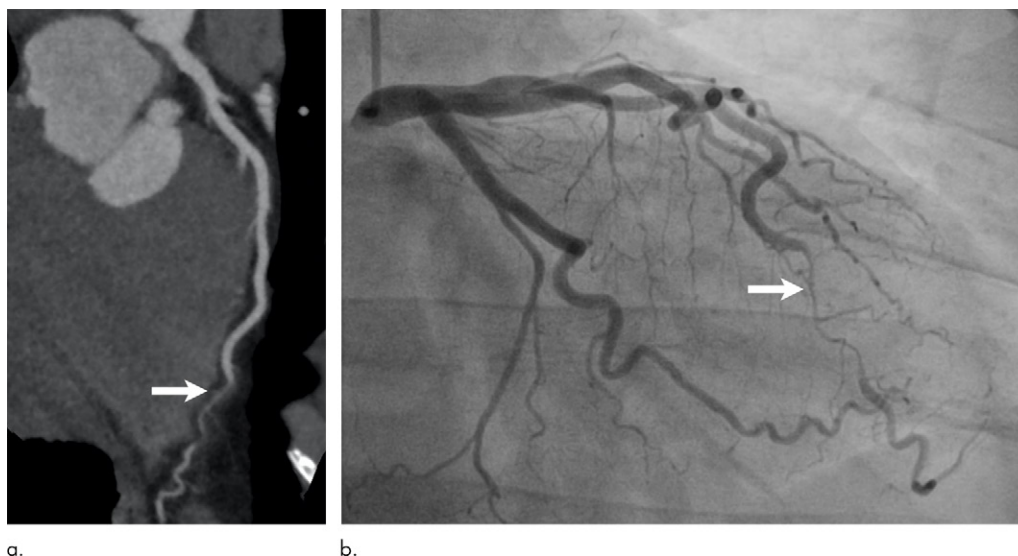


Figure 2: (a) Curved planar reformatted coronary CT and (b) coronary angiograms in a 35-year-old woman who presented with chest pain show tapered, smooth, and diffuse narrowing of distal left anterior descending coronary artery (arrow).

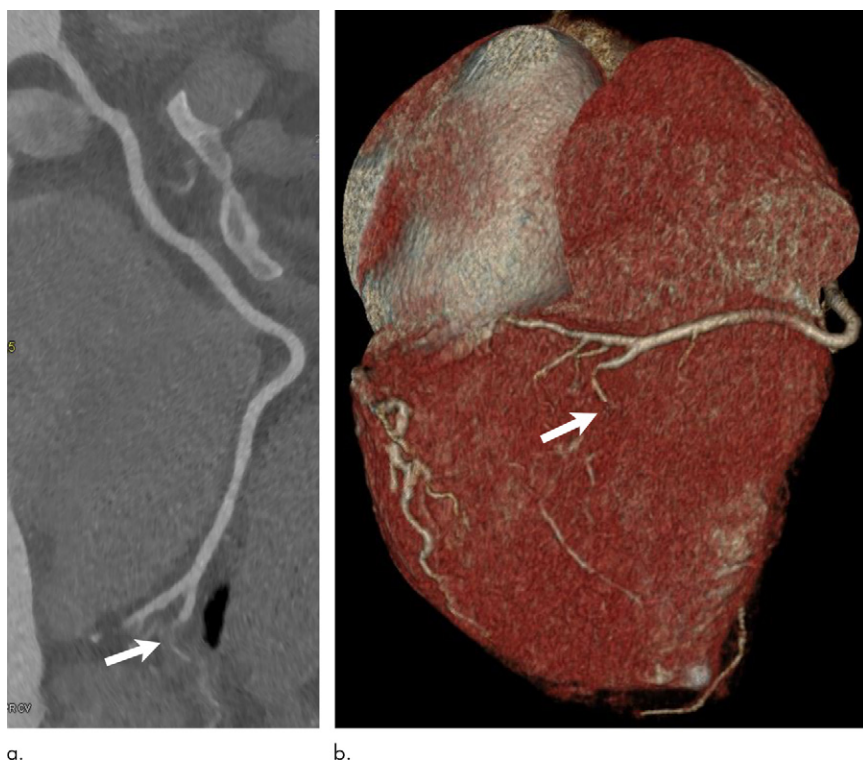


Figure 3: (a) Curved planar reformatted and (b) volume-rendered three-dimensional coronary CT angiography images in a 42-year-old woman with prior history of apical infarct, who presented to emergency department with chest pain, show irregular tapered severe narrowing of the proximal posterior descending artery with adjacent epicardial fat stranding (arrow).

coronary syndromes. Patients with SCAD have a high prevalence of coronary artery tortuosity (14,38) (Figs 1, *G*, and 8) and myocardial bridging (39) (Figs 1, *H*, and 8), features that can readily be identified with coronary CT angiography and may define high-risk phenotype. Using a semiquantitative coronary artery tortuosity score, Eleid et al found that tortuosity was more

common in patients with SCAD (78% vs 17% in control) (38).

A discrete intimal-medial dissection flap, analogous to type 1 SCAD at ICA, can be seen at coronary CT angiography particularly in larger vessels, but increasingly visible in smaller diameter vessels with an improved spatial and temporal resolution of CT scanners (29,30). Cardiac motion or slab artifact can mimic an intimal-medial dissection flap (Table 2). Presence of the dissection flap on multiple cardiac phases and other concurrent abnormalities, such as intramural hematoma and perivascular fat stranding, favor diagnosis of SCAD. Small retrospective studies of patients with SCAD have shown that most common coronary CT angiography features are tapered or abrupt luminal stenosis and intramural hematoma (33,34). A long-segment tapered or abrupt luminal stenosis at coronary CT angiography is analogous to type 2 SCAD seen at ICA (Fig 2b). Atherosclerotic CAD may sometimes be difficult to distinguish from SCAD (Table 2). Focal or short-segment coronary artery stenosis at coronary CT angiography compatible with type 3 SCAD at ICA may mimic atherosclerotic CAD, particularly if only the morphology of coronary artery lumen is assessed.

Coronary CT angiography, in addition to the assessment of lumen, allows assessment of calcified and noncalcified plaque and intramural hematoma and helps distinguish atherosclerotic CAD from SCAD (Figs 4, 6). The combination of non-contrast-enhanced CT scan for assessment of coronary artery calcification and coronary CT angiography is sensitive for the detection of

minimal atherosclerotic CAD, which may not be detected at ICA. The presence of atherosclerotic CAD in a nonculprit vessel may therefore aid diagnosis in challenging cases, as at times it may not be possible to differentiate intramural hematoma from noncalcified plaque (33). High attenuation on non-contrast-enhanced CT images may also occasionally help differentiate coronary artery intramural hematoma from noncalcified plaque (Fig 6).

Coronary vasospasm may also mimic SCAD (Table 2) with diffuse luminal stenosis (type 2 SCAD) or focal luminal stenosis (type 3 SCAD). Concurrent presence of intramural hematoma (40) or perivascular fat stranding (37) points toward the diagnosis of SCAD. Another mimic of SCAD is coronary artery embolism (Table 2), which usually demonstrates abrupt luminal occlusion (41). Involvement of multiple coronary artery territories, luminal occlusion adjacent to branch points, and the presence of an embolic source favors the diagnosis of coronary artery embolism over SCAD.

Stress cardiomyopathy can mimic SCAD, as both entities have similar demographic profiles, predisposing stressors, clinical presentations, and distribution of left ventricular wall motion abnormality. Hausvater et al (42) found that retrospective assessment of ICA in 80 patients with a provisional diagnosis of stress cardiomyopathy revealed 2.5% with definite diagnosis of SCAD and 9% with ICA indeterminate for SCAD. Duran et al found concomitant stress cardiomyopathy in approximately 56% of patients with SCAD (43), suggesting a potential overlap of the two conditions (44,45). Mimics of coronary CT angiography findings in SCAD are discussed in Table 2.

Incremental value of coronary CT angiography.—Several different reconstructions of coronary CT angiography images enable a more detailed analysis of each coronary artery with innumerable projections, which allows separate views of overlapping vessels, which may not be possible with ICA. Intracoronary imaging and ICA confer a risk of causing iatrogenic dissection (46) in the coronary and extracoronary arteries due to vascu-



Figure 4: Spontaneous coronary artery dissection (SCAD) and coronary artery disease (CAD) cases that manifested as diagnostic conundrums. **(a)** Curved planar reformatted coronary CT angiography image in a 42-year-old woman, who presented with chest pain waking her from sleep, shows abrupt severe luminal narrowing (arrow) of distal left anterior descending artery with a notable absence of coronary calcium. Presence of tortuosity, perivascular fat stranding (magnified view inset), and review of previous MR angiography (not shown) demonstrating beading of renal arteries indicating fibromuscular dysplasia led to diagnosis of SCAD. **(b)** Curved planar reformatted coronary CT angiography image in a 69-year-old woman, who presented with several episodes of dull chest pain at exertion and rest, shows abrupt severe luminal narrowing (arrow) of proximal left anterior descending artery. Presence of noncalcified plaque (chevron; magnified view inset) at the level of stenosis and in right coronary artery (not shown) favored atherosclerotic CAD. Noncalcified plaques can mimic intramural hematoma; however, no high attenuation was seen on non-contrast material-enhanced scan to indicate intramural hematoma.

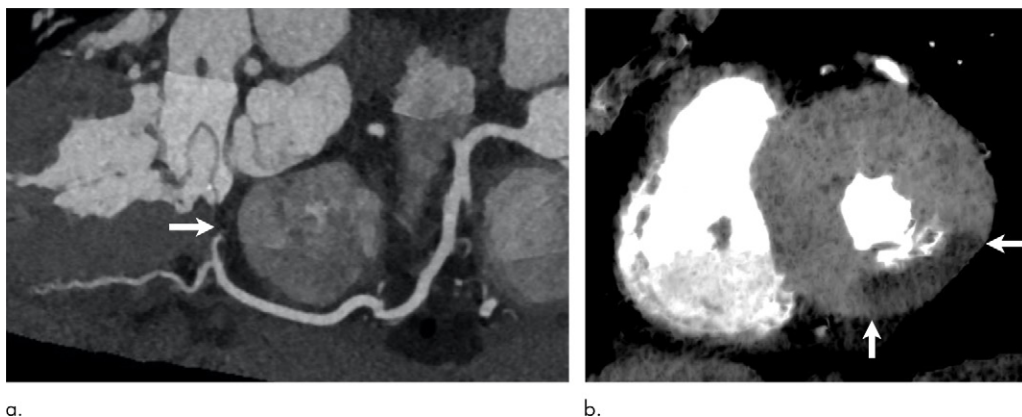
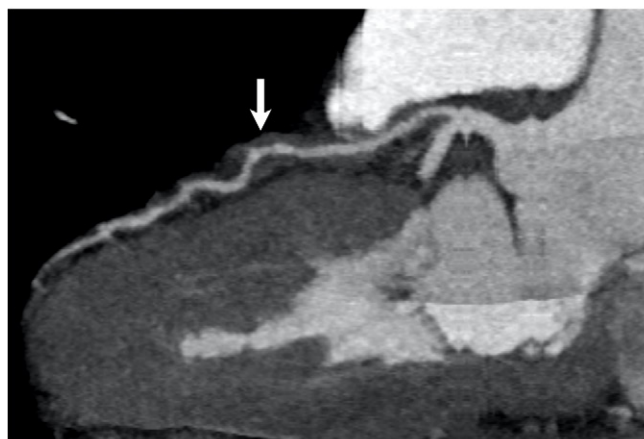


Figure 5: **(a)** Curved planar reformatted and **(b)** minimum intensity projection short-axis coronary CT angiography images in a 68-year-old woman, who presented with chest pain, show luminal occlusion of posterior left ventricular branch (arrow in **a**) with myocardial hypoperfusion in the corresponding territory (arrows in **b**). Involvement of single coronary artery territory and absence of an embolic source favored diagnosis of spontaneous coronary artery dissection over coronary artery embolism.



Figure 6: (a) Axial non-contrast material-enhanced CT, (b) axial coronary CT angiography, and (c) curved planar reformatted coronary CT angiography images in a 32-year-old woman, who was 2 weeks postpartum and presented with dyspnea and chest pain radiating to neck, back, jaw, and shoulder, show left anterior descending coronary artery intramural hematoma (arrow in c).



c.

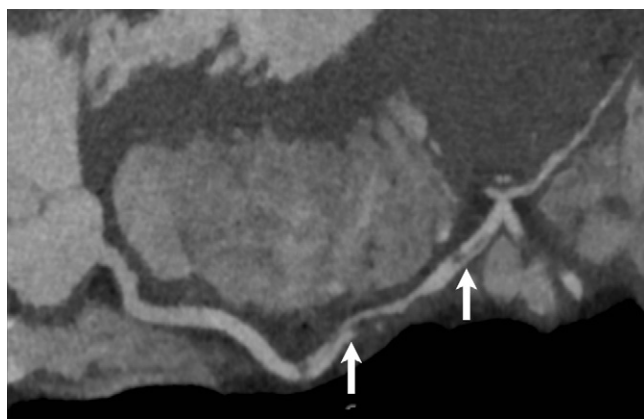


Figure 7: Curved planar reformatted coronary CT angiography image in a 29-year-old man with chest pain and elevated troponin level shows right coronary artery intimal disruption and dissection flap (arrows).

lar tortuosity and fragility. The risks from these other imaging techniques make coronary CT angiography the preferred modality for follow-up imaging in patients with SCAD experiencing recurrent symptoms and for surveillance. Coronary CT angiography offers added value in detecting non-flow-limiting coronary atherosclerosis that results in less than 50% luminal stenosis, which is often difficult to detect at ICA. Moreover, patients with an initial diagnosis of SCAD and no evidence of residual dissection at coronary CT angiography at 3–6 months have been shown to have excellent long-term prognosis (47).

Noncoronary vascular imaging.—All patients with SCAD should undergo additional cross-sectional imaging with either CT or MR angiography for the assessment of SCAD-associated arteriopathy (1,48). Fibromuscular dysplasia, the most common coexisting condition (13) with SCAD, can manifest as arterial stenosis, aneurysm, tortuosity, or dissection (1,16). The most common appearance in fibromuscular dysplasia is “string-of-beads” due to alternating areas of stenosis and dilatation (Figs 1, I and J, 9, 10). CT or MR angiography is useful in identifying noncoronary vascular abnormalities in inherited arteriopathies and connective tissue disorders associated with SCAD (14,48).

Limitations of coronary CT angiography in SCAD.—Use of coronary CT angiography is associated with risks of radiation exposure and iodinated contrast material administration, particularly in pregnant patients. Implementation of several dose reduction strategies has led to a substantial reduction in radiation dose from coronary CT angiography (28,49). Normal coronary CT angiography does not exclude SCAD, as the involvement of a distal vessel less than 1.5 mm in diameter may be beyond scanner resolution. In such ambiguous cases, the presence of other features such as intramural hematoma, perivascular fat stranding, myocardial hypoperfusion, or re-

gional wall motion abnormality may help. Often, resolution of diagnostic dilemma requires recognition of a distinct patient phenotype and use of a multimodality imaging approach, including use of ICA, which provides better resolution of smaller, more distal, coronary arteries (Fig 2b), and cardiac MRI, which enables assessment of myocardial scar and viability (Fig 11). It must be recognized by imagers that diagnosis of SCAD is difficult and at times can be indistinguishable from atherosclerotic CAD in terms of coronary CT angiography findings and clinical presentation. Although a high index of suspicion is required to recognize subtle cases of SCAD, this must be balanced to prevent erroneous diagnosis of SCAD in the setting of atherosclerotic CAD and other mimickers, including coronary CT angiography-related artifacts.

Conclusions

Coronary CT angiography is a useful noninvasive modality for comprehensive initial assessment and follow-up imaging of SCAD with concurrent detection of associated arteriopathies. Coronary CT angiography facilitates the assessment of coronary anatomy, luminal stenosis, intimal dissection flap, intramural hematoma, myocardial perfusion defects, and regional wall motion abnormality. Multimodality imaging complements coronary CT angiography for imaging SCAD in specific situations, including hemodynamic compromise at presentation, assessment of distal coronary arteries beyond the resolution of the CT scanner, and assessment of myocardial viability.

Disclosures of Conflicts of Interest: S.G. disclosed no relevant relationships. N.M.M. disclosed no relevant relationships. M.J.W. disclosed no relevant relationships. M.L.S. Activities related to the present article: disclosed no relevant relationships. Activities not related to the present article: disclosed money paid to author from Canon Medical-Vital Images for consultancy. Other relationships: disclosed no relevant relationships. R.B. disclosed no relevant relationships. B.B.G. Activities related to the present article: disclosed no relevant relationships. Activities not related to the present article: disclosed grants/grants pending to author's institution from Siemens and National Institutes of Health for unrelated projects; disclosed money paid to author from Siemens for minor travel reimbursement. Other relationships: disclosed no relevant relationships. S.S.H. disclosed no relevant relationships.

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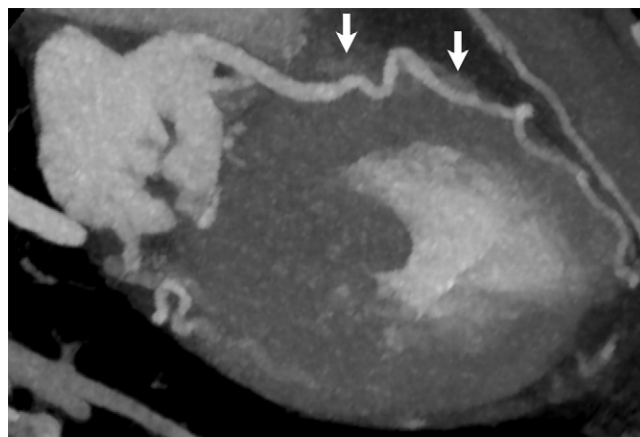


Figure 8: Maximum intensity projection reformatted coronary CT angiography image shows tortuous left anterior descending coronary artery with myocardial bridging (arrows) in a 68-year-old woman who was later found to have fibromuscular dysplasia.

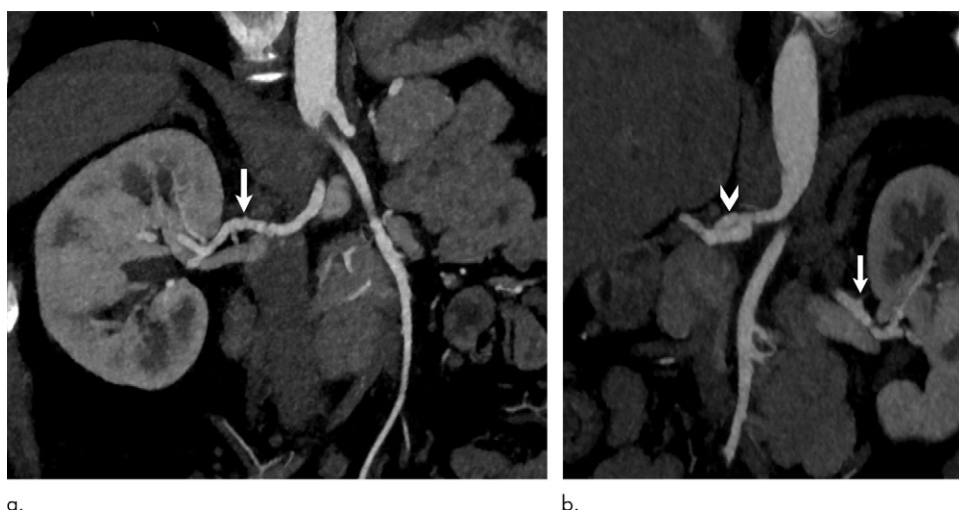


Figure 9: Maximum intensity projection images from CT angiogram of the same patient as Figure 3 (42-year-old woman) show beading of the (a) right and (b) left renal arteries (arrow). (b) Focal dissection and beading (chevron) of celiac artery is also seen.

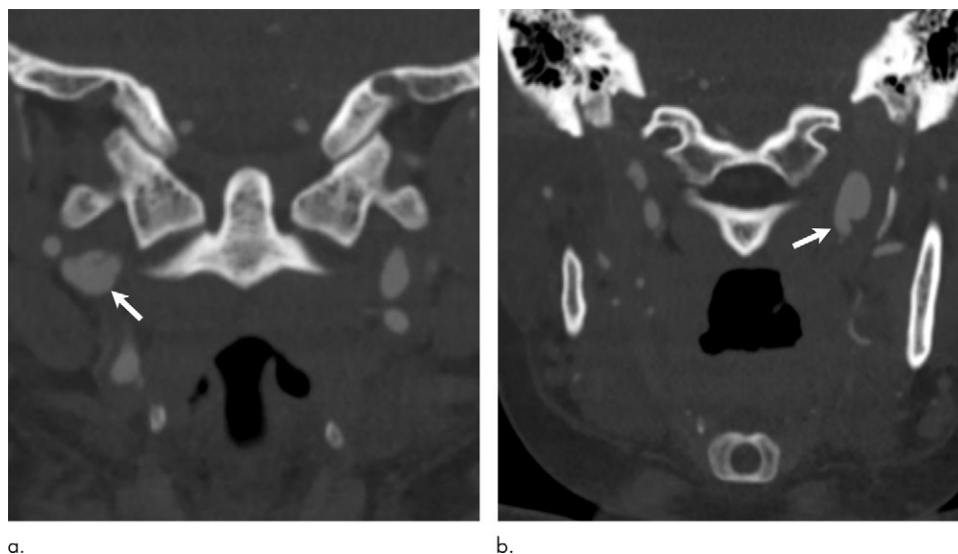


Figure 10: Coronal images from CT angiogram in the same patient as Figure 3 (42-year-old woman) show fusiform dilation of the (a) right and (b) left internal carotid arteries (arrow). Focal dissection of proximal right internal carotid artery is seen.

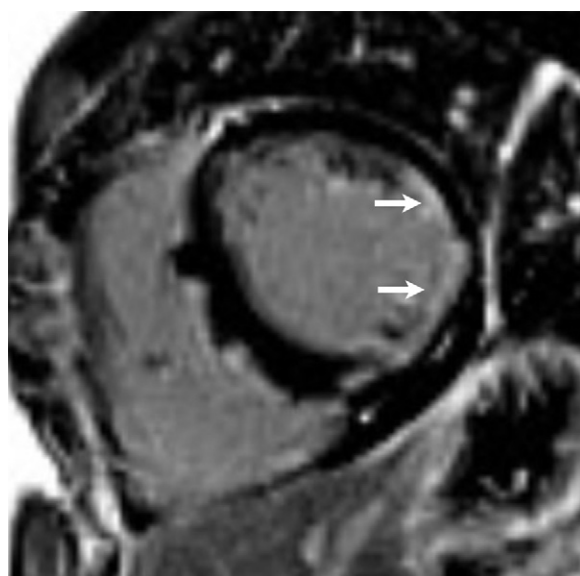


Figure 11: Phase-sensitive inversion recovery cardiac short-axis MR image in a 43-year-old woman with connective tissue disorder and multivessel spontaneous coronary artery dissection shows subendocardial late gadolinium enhancement in left circumflex coronary territory (arrows).

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