

Multifaceted Presentation of Recurrent Spontaneous Coronary Artery Dissection Angiography and Optical Coherence Tomography Findings

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A 58-year-old woman with a previous history of hypertension, hypercholesterolemia, and cigarette smoking presented with a non-ST-segment-elevation myocardial infarction with transient (nonpersistent) anterior ST-segment-elevation suggesting ischemia on the territory of the left anterior descending coronary artery. The coronary angiogram showed no signs of atherosclerosis but depicted severely tortuous coronary arteries with a focal lumen narrowing in the mid-segment of the left anterior descending coronary artery (Figure 1A). In addition, a diffuse lesion with a double lumen was demonstrated at the mid-segment of the posterior descending coronary artery branch of the right coronary artery (Figure 1B). Moreover, the distal left circumflex coronary artery also showed an image of a double lumen (Figure 1C). These findings were highly suggestive of 3-vessel spontaneous coronary artery dissection (SCAD). As the patient was stable and asymptomatic with no signs of ongoing ischemia, a conservative medical approach was selected, and she was eventually discharged on a low dose of aspirin. A control angiogram scheduled at 12 months depicted *resitutio ad integrum* of the left anterior descending coronary artery and posterior descending coronary artery lesions with a completely normal vessel appearance (Figure 1D and 1E). Nevertheless, the image of double lumen on the distal circumflex coronary artery remained unchanged.

Three years later, the patient was admitted again with a new non-ST-segment-elevation myocardial infarction, this time with electrocardiographic signs of inferior ischemia. The coronary angiogram revealed a new image of mild lumen narrowing on the most proximal segment of the left anterior descending coronary artery (Figure 2A). Optical coherence tomography (OCT) confirmed the presence of an intramural hematoma without any intimal tear at this level (Figure 2D). The circumflex coronary artery showed the persistence of the double lumen image, consistent with a chronic nonsealed SCAD (Figure 2B). This was confirmed by OCT that nicely displayed the double lumen and the intimal flap (Figure 2E). Importantly, a mild but extremely long lumen narrowing was detected on the right coronary artery diffusely involving its

proximal and mid-segments (Figure 2C). This image persisted after repeated intracoronary nitroglycerin administration. On intravascular ultrasound, the typical features of a huge intramural hematoma were unravelled (Figure 2F). OCT imaging of this segment confirmed the existence of a long intramural hematoma but also disclosed a confined intimal tear that intravascular ultrasound failed to visualize (Figure 2G). Because the patient was at that point asymptomatic, without signs of acute ischemia, a conservative medical management was indicated. A head-to-pelvis computed tomography scan was scheduled as part of the screening of fibromuscular dysplasia and other extracoronary vascular abnormalities as currently recommended.¹ Unfortunately, 48 hours after discharge, the patient came back to the Emergency Department with acute onset of left hemianopia. An urgent cranial computed tomography scan confirmed a small area of intracranial bleeding at the right portion of the occipital lobe, suggestive of a hemorrhagic transformation of ischemic stroke, and she was admitted to the stroke unit. Selective angiography of the supra-aortic arcs and intracerebral circulation was performed, revealing the presence of large bilateral aneurysms at both intracranial internal carotid arteries and also signs of fibromuscular dysplasia on both internal carotid arteries with the typical string-of-beads pattern (Figures 3A and 3B). Neurological impairment progressively improved, and the patient could be discharged 7 days later with only mild visual sequelae on physical examination. At 3-month clinical follow-up, she remained uneventful and asymptomatic.

SCAD remains a rare cause of acute coronary syndrome. Classically, this clinical entity has been related to young females without coronary risk factors frequently in relation with specific stress periods as peripartum or menopause. Recent large series, however, have demonstrated that a great majority of patients are in fact mid-aged women with a high prevalence of classic coronary risk factors.^{2,3} Moreover, recent studies suggest that intracoronary imaging (intravascular ultrasound and OCT) may play a major role in the accurate diagnosis of this elusive unique condition.^{2,3} Current evidence supports an initial conservative medical approach (watchful waiting strategy) as the optimal initial management for most of these patients,

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limiting revascularization to cases with recurrent and ongoing ischemia or hemodynamic instability.⁴ Last, but not least, it has become evident the existence of a strong association between SCAD and other systemic conditions as fibromuscular dysplasia and other extracoronary vascular abnormalities.⁴

Here we report a unique patient with recurrent episodes of SCAD presenting with the entire spectrum of the morphological manifestations produced by this entity. The distinct dynamic morphological features were unravelled by angiography and confirmed by intravascular ultrasound and OCT. Acute intimo-medial flaps and intramural hematomas could be demonstrated at different stages. Moreover, during follow-up, evidence of healed dissections coexisting with images of chronic nonsealed flaps were detected. Finally, striking images of associated fibromuscular dysplasia and other extracoronary vascular abnormalities were readily visualized.

This patient nicely exemplifies the dynamic and multifaceted clinical presentation and the diverse underlying substrate of SCAD involving not only the coronary arteries but also other large arteries.

Disclosures

None.

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KEY WORDS: atherosclerosis ■ fibromuscular dysplasia ■ intravascular ultrasound ■ optical coherence tomography ■ spontaneous coronary dissection

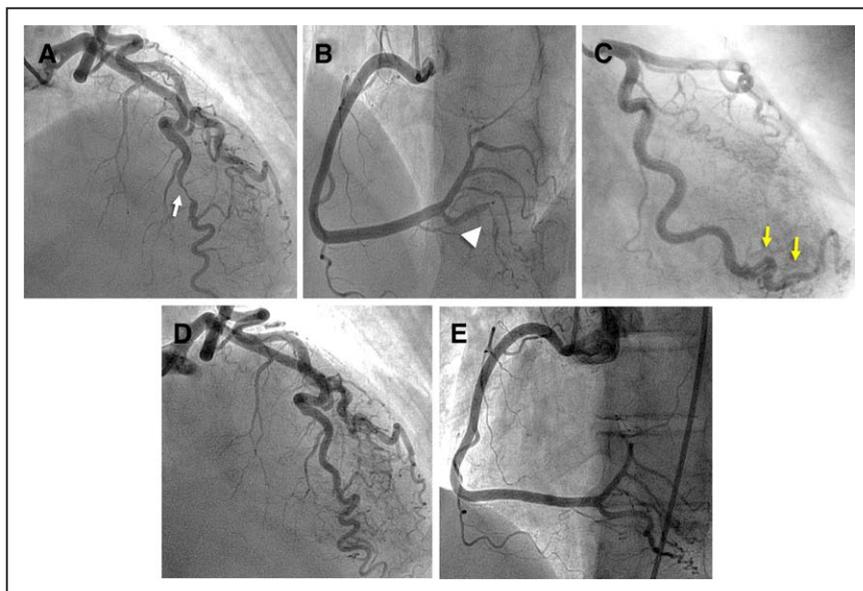


Figure 1. Coronary angiogram showing (A) focal lesion (white arrow) at the mid-segment of left anterior descending coronary artery (LAD). (B) Mid-portion of the posterior descending coronary artery (PDA) with focal stenosis and double-lumen image (white arrowhead) and TIMI 2 (Thrombolysis in Myocardial Infarction 2) distal flow. (C) Distal segment of left circumflex coronary artery (LCX) with double-lumen morphology suggesting spontaneous coronary artery dissection (SCAD; yellow arrows). (D) and (E) Control angiogram 12 months later showing complete resolution of both lesions at the mid-LAD (D) and PDA (E).

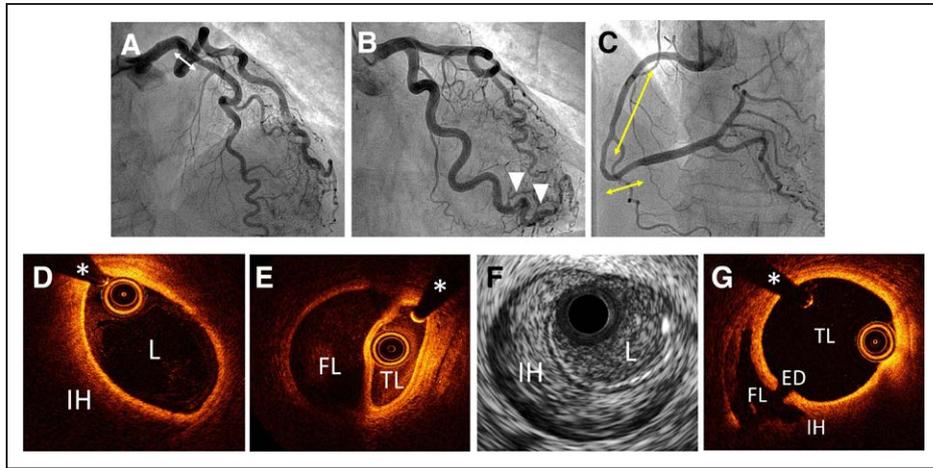


Figure 2. **A**, Cranial projection of left anterior descending coronary artery (LAD) depicting a mild proximal lumen narrowing (double-head white arrow). **B**, Caudal projection showing persistence of a double-lumen image (white arrowheads) at the distal portion of left circumflex coronary artery (LCX). **C**, Cranial projection demonstrating a long segment of mild diffuse narrowing at the proximal and mid right coronary artery (RCA; limited by the yellow lines). **D**, Optical coherence tomography (OCT) run at the proximal LAD disclosing an image suggestive of intramural hematoma (IH) that displaces the lumen (L). **E**, OCT at distal LCX shows a double-lumen pattern suggesting nonsealed chronic spontaneous coronary artery dissection (SCAD). **F**, Intravascular ultrasound at the mid-segment of the RCA showing an IH involving 3 quarters of the circumference of the true lumen. **G**, OCT was able to identify the entry door (ED) of the dissection at the mid-distal segment of RCA. FL indicates false lumen; and TL, true lumen. *denotes wire artifact.

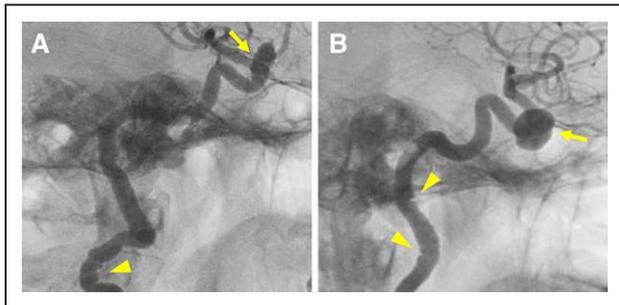


Figure 3. Angiogram of the supra-aortic arcs and intracerebral circulation showed large bilateral aneurysms affecting the intracranial portion of both internal carotid arteries (yellow arrows) and also signs of fibromuscular dysplasia in both extracranial left (**A**) and right (**B**) internal carotid arteries with the typical string-of-beads pattern (yellow arrowheads).

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