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CASE REPORT

INTERMEDIATE

CLINICAL CASE: TCT 2022

Chest Pain and the Dynamic Evolution of Spontaneous Coronary Artery Hematoma

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ABSTRACT

Spontaneous coronary intramural hematoma (SCIH) is a rare but underdiagnosed condition, with dynamic evolution. We present a patient with acute chest pain and normal coronary angiogram undergoing work-up for myocardial infarction with nonobstructive coronary arteries. Cardiac magnetic resonance revealed an ischemic pattern, and subsequent angiography revealed coronary occlusion by SCIH. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2023;6:101673)

HISTORY OF PRESENTATION

A 45-year-old woman was admitted to the emergency department with chest pain, and fever (peak 38 °C) in the previous days. Markers of myocardial injury were elevated (high-sensitivity troponin T 222 pg/mL at baseline and 582 pg/mL 3 hours later), white blood cell count and C-reactive protein were mildly elevated, and D-dimer, chest X-ray, and electrocardiography were normal (**Figure 1**). The patient was admitted to the cardiac intensive care unit.

LEARNING OBJECTIVES

- To understand the diagnostic pitfalls and the danger of spontaneous coronary intramural hematoma.
- To understand the importance of cardiac magnetic resonance in the differential diagnosis of myocardial infarction with nonobstructive coronary arteries.

Transthoracic echocardiography showed inferior wall hypokinesia (Video 1). Urgent coronary angiography showed no evidence of obstructive coronary artery disease (**Figure 2,** Videos 2 to 5).

PAST MEDICAL HISTORY

The patient had hypothyroidism and obesity (body mass index 32 kg/m^2). No previous history of cardiovascular events, bleeding, or coagulopathy was reported. One previous pregnancy was carried over without any issues. No history of previous miscarriages was reported.

DIFFERENTIAL DIAGNOSIS

The initial working diagnosis was myocardial infarction with nonobstructive coronary arteries (MINOCA). Further tests were needed to rule out other conditions such as acute myocarditis, coronary spasm, plaque erosion, and spontaneous coronary artery dissection (SCAD).

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

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ABBREVIATIONS AND ACRONYMS

CMR = cardiac magnetic resonance

CT = computed tomography

IVUS = intravascular ultrasound

MI = myocardial infarction

MINOCA = myocardial infarction with nonobstructive coronary arteries

SCAD = spontaneous coronary artery disease

SCIH = spontaneous coronary intramural hematoma

INVESTIGATIONS

Cardiac magnetic resonance (CMR) was performed 2 days later, showing inferior wall ischemic pattern (**Figure 3,** Video 6), and electrocardiography (ECG) showed changes in the inferior leads with T-wave inversion (**Figure 4**).

A second coronary angiogram with planned intravascular imaging was than performed and showed a critical stenosis of the mid-distal right coronary artery, determining functional vessel occlusion (**Figure 5,** Videos 7 and 8). Coronary vasospasm was ruled-out after intracoronary nitrates infusion (200 μg nitroglycerine). Intravascular ultrasound (IVUS) was than performed, showing diffuse intramural hematoma of the ostial, proximal, and middle segment of the right coronary artery with subocclusive stenosis at the middle segment with no evidence of atherosclerosis (**Figure 6,** Video 9).

MANAGEMENT

Considering the clinical and radiologic evidence of evolving myocardial injury, conservative management was excluded, and direct stenting of the lesion was performed with IVUS-guided implantation of 4 overlapping drug-eluting stents (Orsiro Mission 3.5/40 mm, 3.5/26 mm, 3.5/22 mm, and 4.0/13 mm) (**Figure 7**) distally to the culprit segment and up to the ostial segment. Angiography and IVUS confirmed optimal stent implantation and complete sealing of the wall hematoma (**Figures 8 and 9,** Video 10). The patient was transferred to the cardiology ward asymptomatic and was discharged uneventfully on day 7.

DISCUSSION

After the first coronary angiogram, which revealed no coronary lesions, our initial working diagnosis was MINOCA. Because no coronary hints of disease were present, and the clinical suspicion for coronary

FIGURE 1 Electrocardiography on Admission

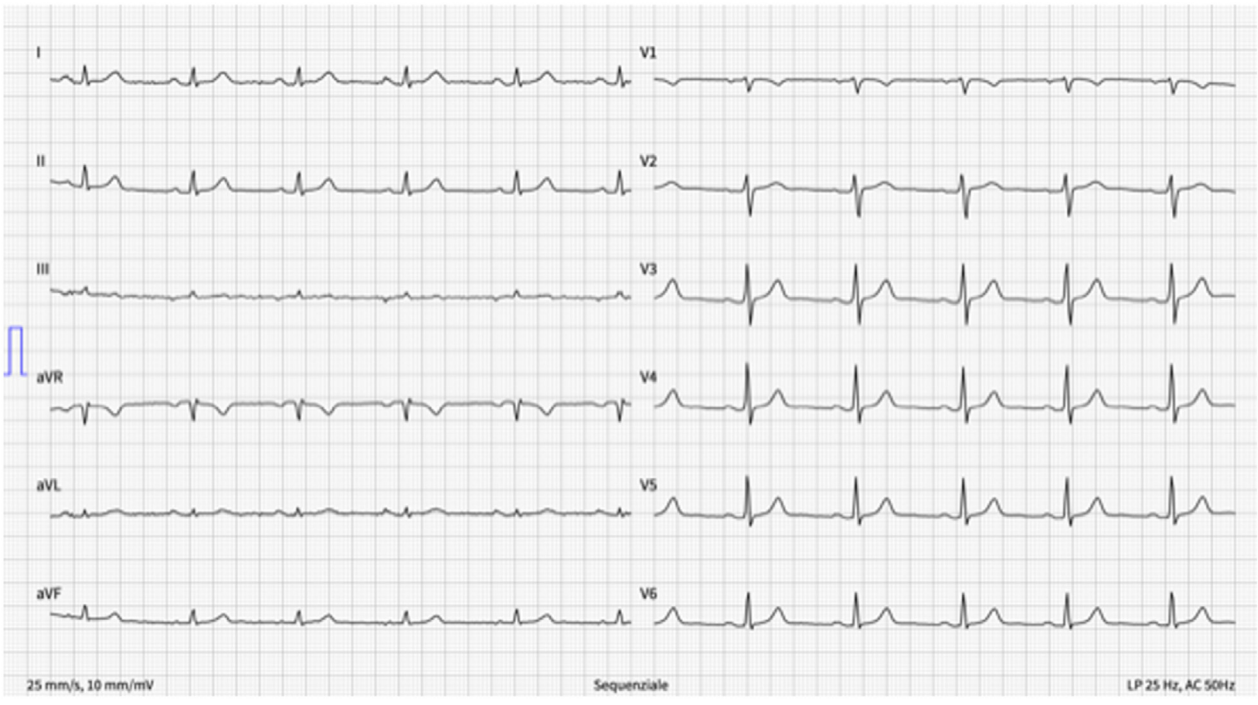


FIGURE 2 First Coronary Angiography

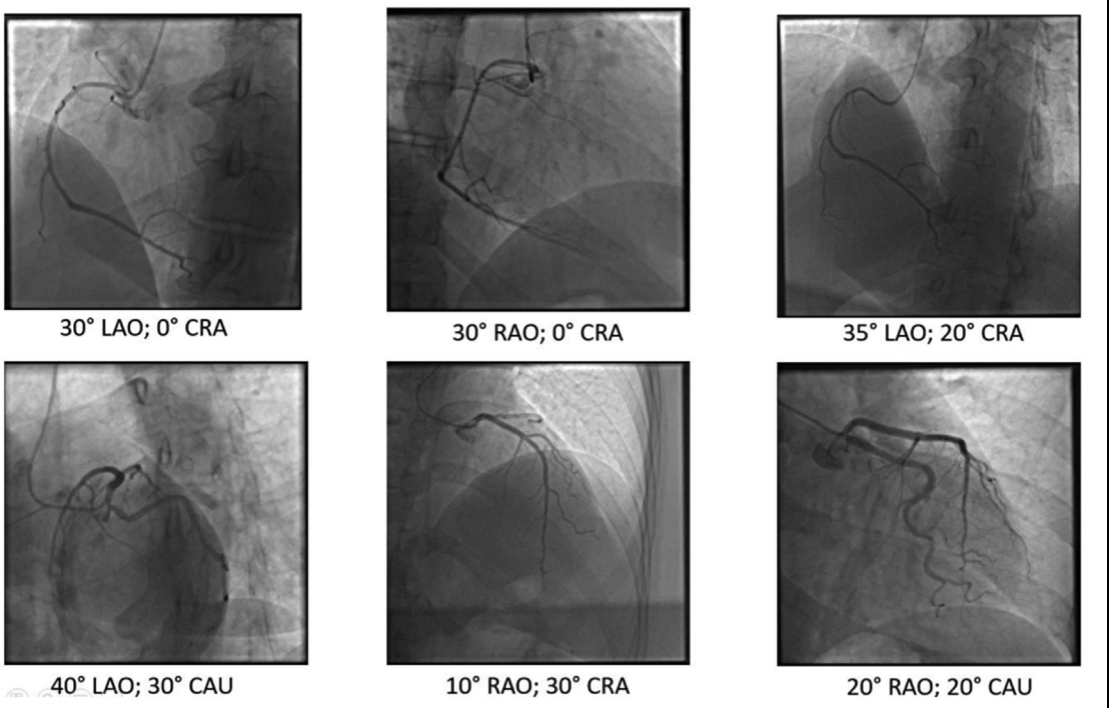
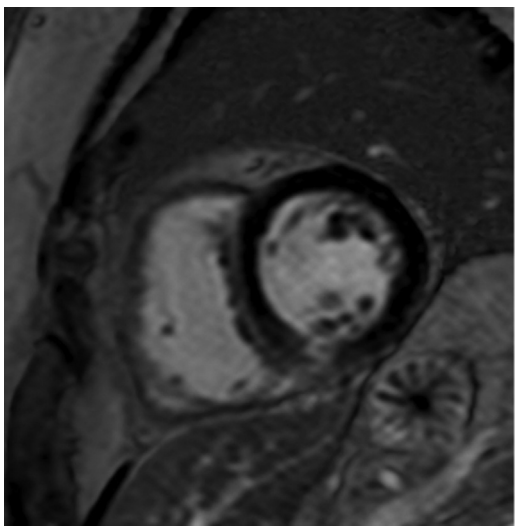


FIGURE 3 Cardiac Magnetic Resonance

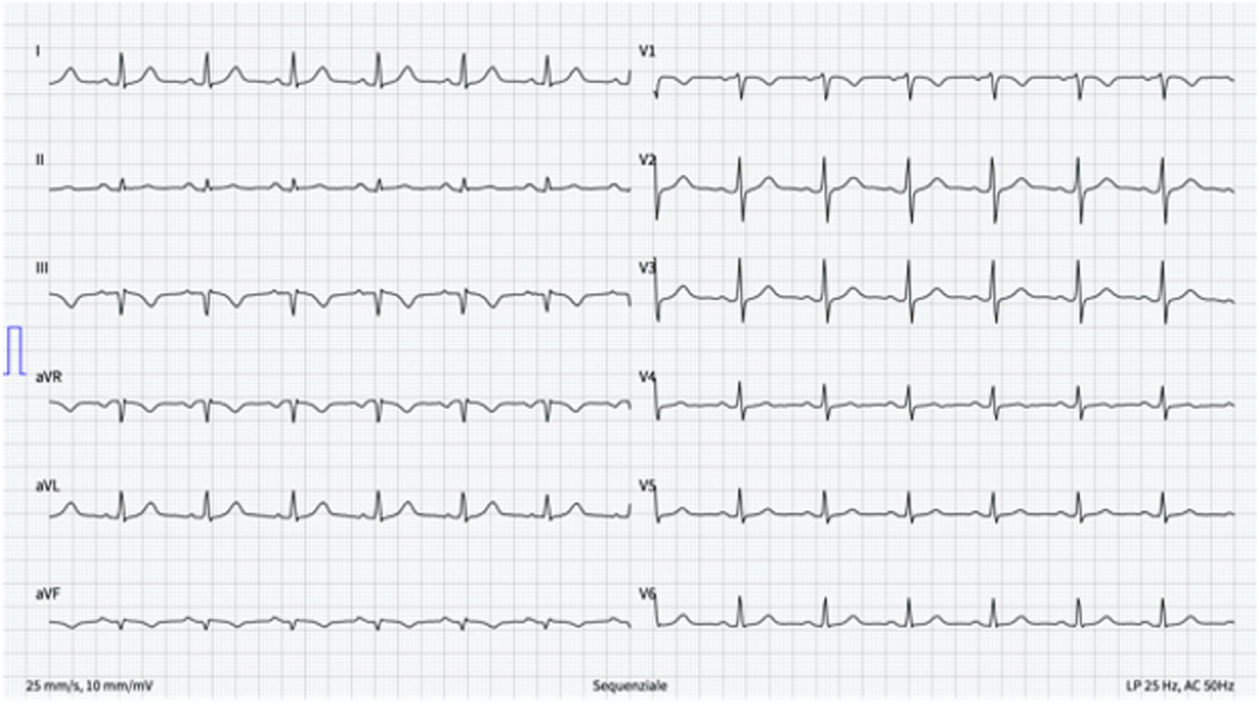


Two-dimensional phase-sensitive inversion recovery images show transmural late gadolinium enhancement in inferior wall.

vasospasm was low, we decided to perform CMR to discriminate inflammatory vs ischemic causes of myocardial injury. Once CMR excluded inflammatory disease and confirmed an ischemic pattern, a second coronary angiogram focused on a better evaluation with intracoronary imaging revealed the final cause of myocardial infarction: a SCIH.

MINOCA is reported in 6% to 15% of patients with acute myocardial ischemia and is generally observed in young patients not affected by traditional cardiovascular risk factors, women, and specific ethnicities. Several tools are needed to finally diagnose patients with MINOCA, including intracoronary imaging, coronary vasoreactive tests, and CMR. A recent diagnostic pathway was suggested by Occhipinti et al[1] for patients with MINOCA: In the absence of any hints of specific disease, such as SCAD or vasospasm, suspected during coronary angiogram, an initial approach with CMR is suggested to differentiate an inflammatory pattern from an ischemic pattern, or specific patterns associated with other conditions, such as cardiomyopathies. Following that pathway, we were able to exclude possible inflammatory causes of acute

FIGURE 4 Electrocardiographic Changes in the Inferior Leads



presentation in young women, such as acute myocarditis, and to confirm an ischemic origin of the disease. In fact, the HARP (Heart Attack Research Program; NCT02905357) MINOCA study showed that about 75% of patients with MINOCA, when undergoing CMR, could be successfully reclassified to a correct diagnosis.[2] In our case, CMR allowed narrowing of the field by excluding numerous differential diagnoses and defining the subsequent diagnostic step that provided the diagnosis of SCIH.

SCIH refers to a spontaneous bleeding occurring between the intima and media coronary layers, secondary to rupture of coronary vasa vasorum. This entity is currently included in the spectrum of SCAD[3,4] and is considered to be rare, even if it may be often misdiagnosed as atherosclerotic coronary disease. SCIH pathophysiology is not well described and

FIGURE 5 Subocclusive Stenosis of the Mid-Segment of the Right Coronary Artery

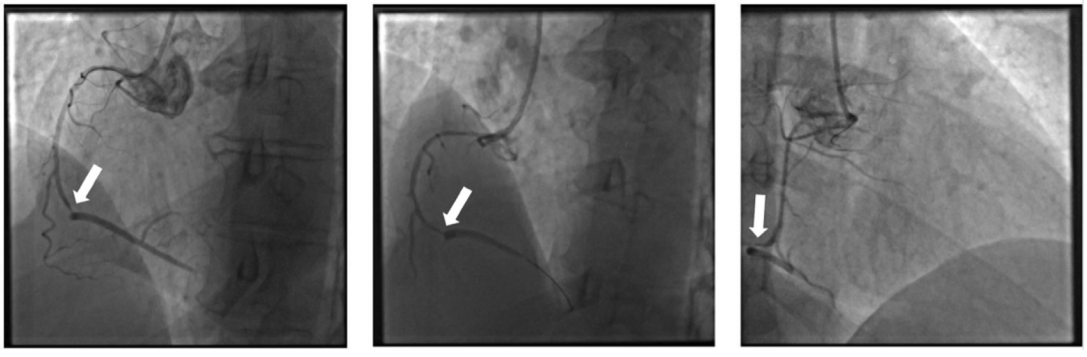
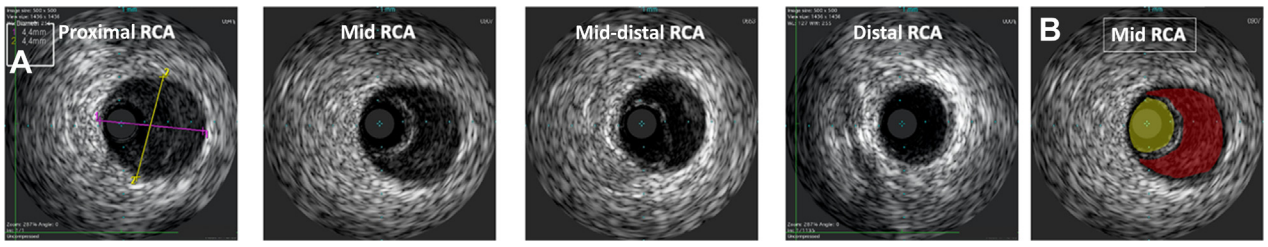


FIGURE 6 **Intravascular Ultrasound**



(A) Intramural hematoma. **(B)** In detail, the false lumen **(red)** and the true lumen **(yellow)** at the mid-segment of the right coronary artery (RCA).

has been hypothesized that it may represent an early phase of SCAD, with intramural hematoma eventually determining vessel obstruction or high wall stress and tearing of the intima determining secondary coronary dissection (the so-called "outside-in" hypothesis). This contrasts with the so-called "inside-out" (or intimal tear) hypothesis, which considers the primary mechanism of SCAD to be the initial disruption of the intimal luminal interface with penetration of blood inside the false lumen for intramural hematoma accumulation.[3,5] In our case, the absence of an intimal tear visualized by intracoronary imaging clearly supports the "outside-in" pathophysiologic hypothesis of SCAD.

Interestingly, we observed the potentially evolving nature of SCIH, which was probably not apparent on the first coronary angiogram and evident later with coronary occlusion on worsening of the wall hematoma. Even if this speculation could not be definitely demonstrated in our case, because no initial intracoronary imaging was performed, a worsening of the SCIH between the first and second coronary angiograms could have possibly been related to the more potent antithrombotic therapy given on hospital arrival. Previous studies have suggested worse outcomes of SCAD with more potent antithrombotic therapy, with a higher risk of nonfatal myocardial infarction and repeated percutaneous coronary intervention in those treated with dual antiplatelet therapy rather than single antiplatelet therapy.[6] Potent antithrombotic therapy may trigger a recurrent bleeding at the site of the SCIH and worsen the coronary obstruction.

Regardless of the underlying mechanism, we observed the dynamic and treacherous nature of SCIH, which poses a high risk of under- or

FIGURE 7 **Coronary Stents Implantation**

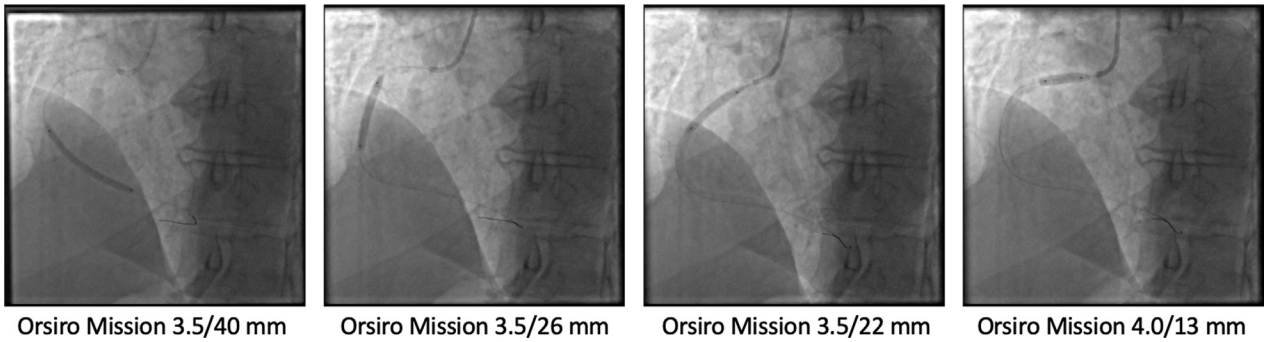
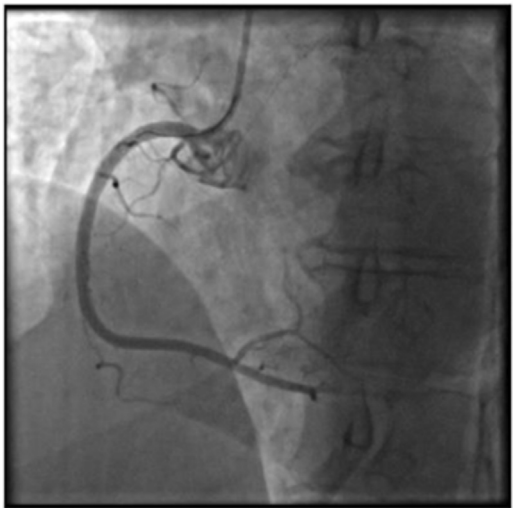


FIGURE 8 Final Angiography



misdiagnosis at an early stage. In this case, a misdiagnosis or even early discharge might have led to catastrophic consequences.

Finally, the optimal interventional treatment of SCIH and SCAD is still debated.[4] The outcomes of percutaneous coronary intervention in these situations are less predictable, with higher rates of complications. The majority of SCAD spontaneously recovers to normal vessel wall architecture within months when conservatively managed, so the general consensus is to conservatively treat hemodynamically and clinically stable patients with SCAD.[4] In our case, the revascularization strategy was deemed necessary owing to the ischemic damage observed on CMR and the rapidly worsening signs of myocardial ischemia, and it was deemed safe after confirmation of correct wiring with the use of IVUS.

FOLLOW-UP

The patient had no further admissions or adverse events after hospital discharge. One-month follow-up echocardiography revealed normal ejection fraction. Brain and lower abdomen magnetic resonance imaging was performed to rule out fibromuscular dysplasia. Six-month coronary CT showed optimal result of coronary stenting with no evidence of disease recurrence or progression.

CONCLUSIONS

This case highlights the dynamic and treacherous nature of SCIH, causing initial symptoms of myocardial ischemia without evident coronary obstruction, and then rapidly evolving in a severe and life-threatening coronary occlusion on hematoma expansion. Higher-level diagnostic testing, such as CMR and IVUS, was essential for correct diagnosis and treatment in this complex scenario.

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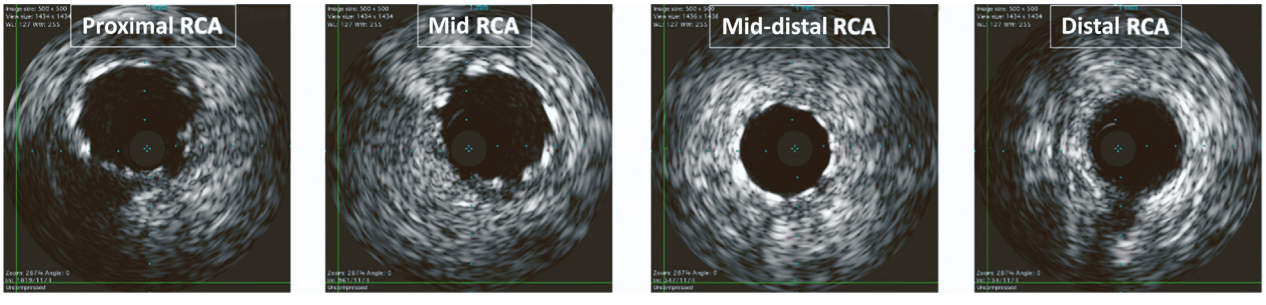
FIGURE 9 Final Control Intravascular Ultrasound



RCA = right coronary artery.

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KEY WORDS acute coronary syndrome, cardiac magnetic resonance, IVUS, MINOCA, SCAD, spontaneous coronary intramural hematoma

APPENDIX For supplemental videos, please see the online version of this paper.