The Texas Heart Institute® Journal

Case Reports

Spontaneous Coronary Artery Dissection in a Male Patient With Fibromuscular Dysplasia

Artur Schneider, DO[1]; S. Michael Gharacholou, MD[2]

[1]Department of Internal Medicine, Mayo Clinic, Jacksonville, Florida
[2]Department of Cardiovascular Medicine, Mayo Clinic, Jacksonville, Florida

Abstract

A male patient presented with cardiac arrest attributed to anterior ST-segment elevation myocardial infarction from type 1 spontaneous coronary artery dissection. Subsequent imaging confirmed fibromuscular dysplasia in noncoronary arterial segments. The patient was started on guideline-directed medical therapy and referred to cardiac rehabilitation, showing substantial improvements in clinical status. With greater awareness and advancements in imaging, spontaneous coronary artery dissection has been more frequently recognized, and although as many as 81% to 92% of all cases occur in female patients, it can be seen among men, as well. Adjunctive imaging for arteriopathies may help establish the diagnosis for equivocal causes of acute coronary syndrome in women and men.

Keywords: Dissection, blood vessel; coronary vessels; fibromuscular dysplasia; acute coronary syndrome; ST elevation myocardial infarction

Case Report

Presentation and Physical Examination

A 57-year-old man presented to the emergency department by ambulance after he experienced sudden onset of crushing chest pain that radiated to his jaw and left arm. An electrocardiogram taken before his arrival confirmed anterior ST-segment elevation myocardial infarction (MI), which quickly degenerated to ventricular fibrillation, prompting defibrillation and cardiopulmonary resuscitation.

Physical examination revealed a well-developed, ill-appearing, diaphoretic male patient in acute distress. Vital signs revealed a body temperature of 36.1 °C, a heart rate of 112/min, a respiratory rate of 20/min, blood pressure of 97/61 mm Hg, and oxygen saturation of 100%. Pulmonary examination revealed an appropriate respiratory rate, nonlabored breathing, no use of accessory muscles, and clear lungs to auscultation bilaterally in all peripheral lung fields. Cardiovascular examination showed a regular heart rate and rhythm, normal S_1 and S_2 sounds, strong pulses bilaterally, and no elevation in jugular venous pressure.

Medical History

The patient's medical history was clinically significant for hyperlipidemia, anxiety, depression, and α-1 antitrypsin deficiency, but he had no known cardiac history. Of note, the patient was experiencing a great deal of emotional hardship: He was residing with his mother, who had recently enrolled in hospice care.

Citation: Schneider A, Gharacholou SM. Spontaneous coronary artery dissection in a male patient with fibromuscular dysplasia. *Tex Heart Inst J.* 2024;51(1):e238358. doi:10.14503/THIJ-23-8358
Corresponding author: Artur Schneider, 4500 San Pablo Rd, Jacksonville, FL 32224 (schneider.artur@mayo.edu)

Differential Diagnosis

Given the sudden onset of crushing chest pain in this patient along with electrocardiographic changes observed en route to the hospital, ST-segment elevation MI was high on the list of differential diagnoses, but one could not exclude the possibility of his symptoms being secondary to coronary vasospasms or spontaneous coronary artery dissection (SCAD). Further workup was required to narrow the diagnosis.

Technique

The patient's laboratory findings were clinically significant for high-sensitivity troponin: 12 ng/L at presentation and 55 ng/L at 1 hour (>51 ng/L signifies a high risk for myocardial injury). Complete blood cell count and kidney function panel results were within normal limits. Chest radiographic findings were clinically significant for low lung volumes and for patchy opacities suggestive of atelectasis in the setting of an elevated hemidiaphragm. Point-of-care cardiac ultrasound imaging showed anterior apical wall motion abnormalities. The interpretation of an electrocardiogram in the emergency department showed ST-segment elevations in the anterior leads consistent with an anterolateral MI. The patient's initial presentation was to a facility that did not perform percutaneous coronary interventions, so thrombolysis was administered along with a loading dose of clopidogrel 300 mg. He was transferred to a facility capable of percutaneous coronary intervention, where he underwent urgent angiography. This procedure demonstrated several radiolucent lumens in the mid–left anterior descending coronary artery, with moderate stenosis likely from intramural hematoma (Fig. 1). Given that the vessel had grade 3 thrombolysis in MI flow, the intervention was deferred, and the patient was treated conservatively. He was started on aspirin 81 mg/d, atorvastatin 80 mg/d, clopidogrel 75 mg/d, and metoprolol 50 mg/d. Initiation of an angiotensin-converting enzyme inhibitor and an angiotensin II receptor blocker was deferred given his borderline low blood pressure readings on presentation.

Outcome

The patient was discharged on his third hospital day, and his left ventricular function was normal at the time of discharge. Given his index SCAD event, the patient underwent brain-to-pelvis imaging to identify possible underlying arteriopathies. A computed tomographic angiogram of the head and neck was clinically significant for fibromuscular dysplasia (FMD) involving the mid–cervical right internal carotid artery (Fig. 2) as well as for focal involvement of the left vertebral artery's V3 segment at the C2 level (Fig. 3). Computed tomograms of the abdomen and pelvis revealed FMD of the bilateral renal arteries (Fig. 4). The patient was evaluated by a neurologist, who recommended avoiding activities that would result in neck manipulation or prolonged hyperextension or flexion.

Key Points

- Although rare, SCAD can occur in men and can present in the setting of arteriopathies and emotional triggers.
- Early diagnosis, conservative management, and avoidance of percutaneous intervention lead to meaningful recovery and favorable outcomes in patients presenting with SCAD.
- Patients who present with SCAD, regardless of their sex, should undergo further workup to evaluate for underlying arteriopathies.

Abbreviations and Acronyms

ACS	acute coronary syndrome
FMD	fibromuscular dysplasia
MI	myocardial infarction
SCAD	spontaneous coronary artery dissection

Latest Follow-Up

The patient completed cardiac rehabilitation following acute coronary syndrome (ACS) and has done well with conservative management.

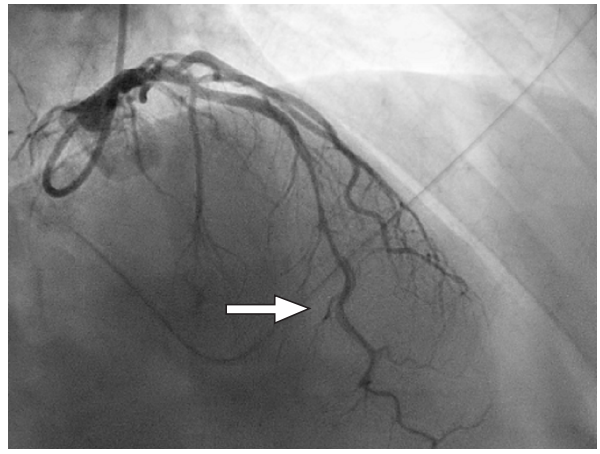

Fig. 1 Coronary angiogram during left heart catheterization demonstrates several radiolucent lumens in the mid–left anterior descending coronary artery (arrow), with moderate stenosis likely from intramural hematoma.

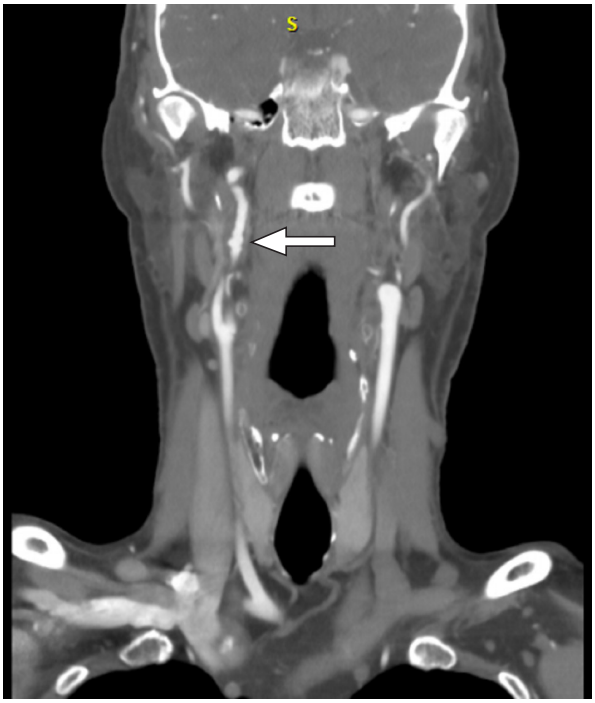

Fig. 2 Computed tomographic angiogram (coronal view) shows the focal, beaded appearance of the left vertebral artery's V3 segment at the C2 to C3 level (arrow).

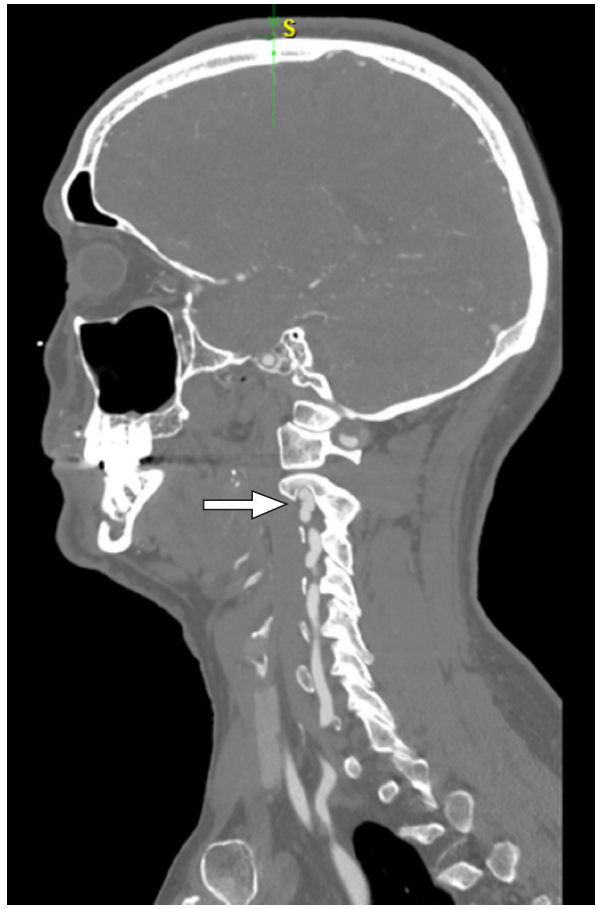

Fig. 3 Computed tomographic angiogram (sagittal view) shows the beaded appearance of the mid–cervical right internal carotid artery (arrow) consistent with fibromuscular dysplasia extending over an approximately 2.5-cm segment.

Discussion

Spontaneous coronary artery dissection has previously been thought to be a rare diagnosis, but with advances in intracoronary imaging, SCAD has been increasingly identified as the cause for ACS in a small number of patients overall, though it may account for up to 30%[1] of ACS cases in women younger than 60 years of age.[2] Of note, 43% of all pregnancy-related MIs are secondary to SCAD.[3]

Spontaneous coronary artery dissection in men continues to be a rare finding. Historically it has been reported that as many as 81% to 92% of all SCAD cases occur in female patients.[4] Spontaneous coronary artery dissection can affect all coronary arteries; however, the left anterior descending coronary artery appears to be affected most, and multivessel SCAD occurs in only 9% to 23% of cases.[5] It is hypothesized that in SCAD, an intramural hematoma spontaneously forms in the outer one-third of the tunica media.[6,7] Though the condition's pathophysiology is currently not fully understood, 2 leading theories describe possible mechanisms of SCAD development. One theory suggests that a disruption in the vessel wall, an intimal tear, is the primary mechanism that allows an intramural hematoma to form. The second theory proposes that the initiating event leading to an intramural hematoma is a hemorrhage from the vasa vasorum within the vessel wall.[5,8] Although further research is required to identify the true cause of these spontaneous events, it is evident that the formation of an intramural hematoma at any level may lead to decreased blood flow, ischemia, and clinical symptoms.

Several conditions have been associated with SCAD and are thought to contribute to the disease process in select cases. Systemic arteriopathies—in particular, FMD—have most commonly been associated with the formation of SCAD. Fibromuscular dysplasia affects medium-sized arteries and causes fibrous tissue to build up, leading to luminal narrowing. In addition to narrowing, patients may develop small arterial aneurysms, tortuosities, or dissections. Arterial "buildup" in FMD can be distinguished from atherosclerotic plaque by its more distal development along the vessel.[9] Several case

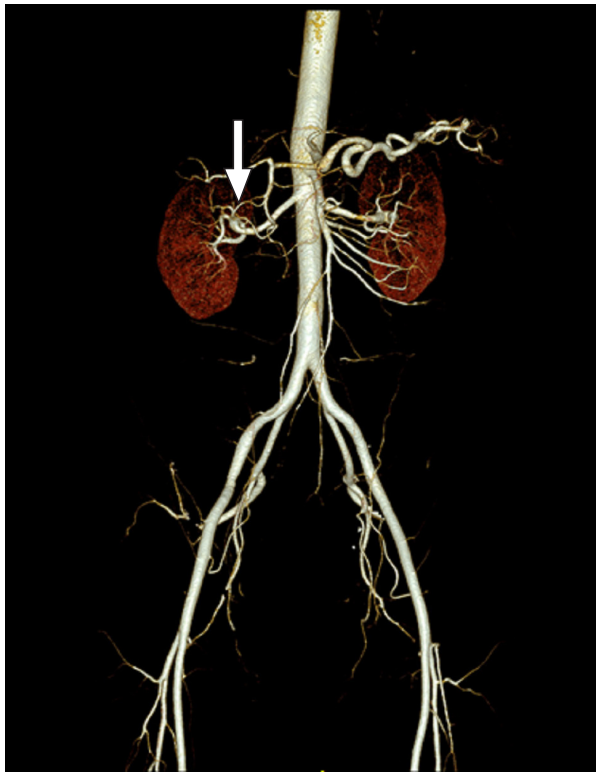

Fig. 4 Computed tomogram (coronal view) shows the beading of the distal right main renal artery (arrow) with aneurysmal dilatation of the segmental right renal arteries, consistent with fibromuscular dysplasia.

reports have discussed the likely correlation between SCAD and FMD. The phosphatase and actin regulator 1 (*PHACTR1*) gene, a pleiotropic locus found in several vasculopathies, has been closely associated with FMD. This locus codes for a phosphatase and actin–binding protein that allows for cellular mobility and organization, and its disruption may lead to disorganized vascular formation. Of note, this same pleiotropism has been associated with SCAD, coronary artery disease, and MI.[10]

Angiographically, 3 variations of SCAD have been identified. Type 1 SCAD is seen in approximately 25% of patients and is defined by typical contrast dye staining of the arterial wall along with multiple visible radiolucent lumens. As many as 70% of patients have been found to have type 2 SCAD, in which diffuse stenosis causes coronary arterial narrowing. Because of its wide range of presentations, type 2 SCAD is further differentiated into type 2A and type 2B. The difference between the 2 subtypes is that a patient with type 2A SCAD continues to have normal coronary arterial segments distal to the occlusion, while a patient with type 2B typically does not. Finally, type 3 SCAD is most similar to atherosclerotic lesions and is therefore difficult to recognize. This variation of the condition often requires intracoronary imaging to reconcile its symptoms with its diagnosis. The absence of atherosclerotic disease in unaffected coronary arteries can be a hint at type 3 SCAD.[11,12]

Any patient presenting with signs and symptoms of ACS should be treated according to evidence-based guidelines. Once SCAD is diagnosed in a patient, however, treatment management slightly differs from that of atherosclerotic coronary artery disease. Though the mainstay therapy of atherosclerotic disease involves revascularization through percutaneous intervention, SCAD is primarily managed medically.[13] Findings suggest that a majority of dissected vessels heal completely over time. Revascularization has also been associated with higher risks of failure and poor outcomes because of arterial instability and friability secondary to the dissection. Mechanically speaking, an arterial dissection poses several risks when trying to place or maintain a stent: The guidewire can be placed in a false lumen, causing worsening of the dissection. In cases of proximal left anterior descending coronary artery or left main SCAD, this error can be catastrophic. Balloon dilatation may dislodge or extend the intramural hematoma and cause obstruction proximally or distally to the dilated segment. Eventual healing of the intramural hematoma may lead to late-acquired malposition of a stent[3,14]; however, high-risk features, including left main coronary artery dissections, ongoing ischemia, hemodynamic instability, low-grade (0-1) thrombolysis in MI flow, and refractory arrhythmias, are treated with revascularization procedures because of their high mortality rates without intervention.[13]

Medical therapy begins with the use of antiplatelet therapies, per ACS guidelines.[15] Anticoagulation is generally avoided because it poses a theoretical risk of worsening intramural hematoma.[16] β-Blockers represent a major component of medical therapy because they help control heart rate, decrease sheer stress, reduce blood pressure, and clinically are associated with a reduced risk of recurrent SCAD.

The current case highlights the importance of maintaining a high index of suspicion for SCAD in patients presenting with signs and symptoms of ACS, even in the absence of classic risk factors, as well as awareness that the presentation may happen in men. The likely trigger

in the current patient was emotional stress brought on by his mother's enrollment in hospice care. This case supports the idea that early diagnosis and conservative management have positive outcomes in SCAD. This patient's diagnosis of FMD also suggests that SCAD can be a first presenting sign in this patient population. All individuals found to have SCAD should undergo further testing to identify comorbidities that may contribute to or predispose them to the development of vascular injury. Further studies are required to better understand the pathophysiology of SCAD and to propose more targeted therapies.

Article Information

Published: 9 May 2024

Author Contributions: We have only included authors who have made an important contribution to the study and are thoroughly familiar with the case, therefore warranting their contribution to the editor. Artur Schneider and S. Michael Gharacholou contributed equally to the development, writing, and editing of this article.

Conflict of Interest Disclosure: The authors have no financial relationships to disclose.

Funding/Support: None.

Disclaimer: The views expressed in this article are those of the authors.

References

1. Saw J, Humphries K, Aymong E, et al. Spontaneous coronary artery dissection: clinical outcomes and risk of recurrence. *J Am Coll Cardiol.* 2017;70(9):1148-1158. doi:10.1016/j.jacc.2017.06.053
2. Gulati R, Behfar A, Narula J, et al. Acute myocardial infarction in young individuals. *Mayo Clin Proc.* 2020;95(1):136-156. doi:10.1016/j.mayocp.2019.05.001
3. Elkayam U, Jalnapurkar S, Barakkat MN, et al. Pregnancy-associated acute myocardial infarction: a review of contemporary experience in 150 cases between 2006 and 2011. *Circulation.* 2014;129(16):1695-1702. doi:10.1161/CIRCULATIONAHA.113.002054
4. Shufelt CL, Pacheco C, Tweet MS, Miller VM. Sex-specific physiology and cardiovascular disease. *Adv Exp Med Biol.* 2018;1065:433-454. doi:10.1007/978-3-319-77932-4_27
5. Saw J, Mancini GBJ, Humphries KH. Contemporary review on spontaneous coronary artery dissection. *J Am Coll Cardiol.* 2016;68(3):297-312. doi:10.1016/j.jacc.2016.05.034
6. Hill SF, Sheppard MN. Non-atherosclerotic coronary artery disease associated with sudden cardiac death. *Heart.* 2010;96(14):1119-1125. doi:10.1136/hrt.2009.185157
7. Desai S, Sheppard MN. Sudden cardiac death: look closely at the coronaries for spontaneous dissection which can be missed. A study of 9 cases. *Am J Forensic Med Pathol.* 2012;33(1):26-29. doi:10.1097/paf.0b013e3181e29598
8. Kwon TG, Gulati R, Matsuzawa Y, et al. Proliferation of coronary adventitial vasa vasorum in patients with spontaneous coronary artery dissection. *JACC Cardiovasc Imaging.* 2016;9(7):891-892. doi:10.1016/j.jcmg.2015.11.030
9. Khoury MH, Gornik HL. Fibromuscular dysplasia (FMD). *Vasc Med.* 2017;22(3):248-252. doi:10.1177/1358863X17700716
10. Georges A, Yang ML, Berrandou TE, et al; FEIRI investigators; International Stroke Genetics Consortium (ISGC) Intracranial Aneurysm Working Group; MEGASTROKE; ARCADIA Investigators. Genetic investigation of fibromuscular dysplasia identifies risk loci and shared genetics with common cardiovascular diseases. *Nat Commun.* 2021;12(1):6031. doi:10.1038/s41467-021-26174-2
11. Saw J. Coronary angiogram classification of spontaneous coronary artery dissection. *Catheter Cardiovasc Interv.* 2014;84(7):1115-1122. doi:10.1002/ccd.25293
12. Saw J, Mancini GBJ, Humphries K, et al. Angiographic appearance of spontaneous coronary artery dissection with intramural hematoma proven on intracoronary imaging. *Catheter Cardiovasc Interv.* 2016;87(2):E54-E61. doi:10.1002/ccd.26022
13. Pristera N, Chaudhury P, Van Iterson EH, Cho LS. Spontaneous coronary artery dissection: principles of management. *Cleve Clin J Med.* 2021;88(11):623-630. doi:10.3949/ccjm.88a.20162
14. Main A, Saw J. Percutaneous coronary intervention for the treatment of spontaneous coronary artery dissection. *Interv Cardiol Clin.* 2019;8(2):199-208. doi:10.1016/j.iccl.2018.11.008
15. Jia S, Liu Y, Yuan J. Evidence in guidelines for treatment of coronary artery disease. *Adv Exp Med Biol.* 2020;1177:37-73. doi:10.1007/978-981-15-2517-9_2
16. Adlam D, Alfonso F, Maas A, Vrints C; Writing Committee. European Society of Cardiology, acute cardiovascular care association, SCAD study group: a position paper on spontaneous coronary artery dissection. *Eur Heart J.* 2018;39(36):3353-3368. doi:10.1093/eurheartj/ehy080