Spontaneous Coronary Artery Dissection in a Young Man with a Factor V Leiden Gene Mutation: A Case Report and Review of the Literature

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Abstract

Keywords

- ▶ spontaneous coronary artery dissection
- ▶ myocardial infarction
- ▶ acute coronary syndrome
- ▶ factor V Leiden gene mutation

Spontaneous coronary artery dissection is a rare but increasingly recognized cause of acute myocardial ischemia in young adults, especially in women. We report a case of spontaneous coronary dissection in a young healthy man who was also a carrier of the factor V Leiden gene mutation.

Spontaneous coronary artery dissection (SCAD) is a rare cause of acute coronary syndromes (ACS). It is reported more commonly in young women, with less than one-third of cases reported in men.¹ We report a case of a young male presenting with ACS due to SCAD on coronary angiography associated with factor V Leiden mutation. To our knowledge, this is the first case of SCAD reported in association with factor V Leiden mutation.

Case Report

A 31-year-old man presented with intermittent substernal chest pressure and exertional dyspnea for 12 hours. His medical history was significant for paroxysmal atrial tachycardia status postablation during childhood. He was not taking any medications, smoked tobacco occasionally, and had no history of illicit drug use.

Examination revealed a blood pressure of 157/70 mm Hg, pulse of 65 beats per minute, oxygen saturation of 100% on

room air, and no fever. Lungs were clear to auscultation, and heart sounds were normal.

Initial electrocardiogram revealed sinus rhythm with less than 1 mm ST-segment elevations in the inferior leads. Repeat electrocardiogram showed greater than 1 mm ST-segment elevation in the inferior leads. Cardiac enzymes at admission, including creatine kinase (CK), CK MB isoenzyme (MB), and troponin I were within normal limits. Repeat cardiac markers 2 hours after presentation to the emergency department were positive. His CK was 344 IU/mL (reference range, 21–232 IU/mL), MB was 20.7 ng/mL (reference range, 0.5–3.6 ng/mL), and troponin I was 0.46 ng/mL (reference range, 0.02–0.49 ng/mL). His troponin I peaked at 24.11 ng/mL, while CK and MB peaked at 1,118 and 124 ng/mL, respectively, on the second day of admission. His lipid panel demonstrated total cholesterol of 245 mg/dL, low-density lipoprotein cholesterol 175 mg/dL, high-density lipoprotein cholesterol 30 mg/dL, and triglyceride level of 241 mg/dL. A urine drug screen was negative.

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Fig. 1 (a) Coronary angiography in left anterior oblique projection showing dissection of the mid-right coronary artery (black arrow) extending to the crux (white arrow); (b) postprocedural result after insertion of four bare metal stents; (c) repeat angiography 3 months later after patient presented with recurrent symptoms showed proximal in-stent stenosis (black arrow); the right posterior descending artery is visible beyond the edge of the original dissection (white arrow); (d) postprocedural result after laser debulking and balloon angioplasty treatment of in-stent stenosis.

Emergent cardiac catheterization demonstrated right coronary artery mid-vessel plaque rupture with thrombus formation and dissection extending to the crux. The patient was treated with four bare metal stents with Thrombolysis in Myocardial Infarction 3 flow and no angiographic evidence of any residual stenosis (►Fig. 1a, b).

A hypercoagulable work-up was positive for heterozygous factor V Leiden gene mutation. A vasculitis work-up was negative including normal erythrocyte sedimentation rate, C-reactive protein, rapid plasma reagin, and antinuclear antibody. A transthoracic echocardiography demonstrated left ventricular ejection fraction of 55%, no wall motion abnormalities, and mild-to-moderate mitral regurgitation.

The patient was discharged home on aspirin, clopidogrel, β blockers, and statin. He presented again 3 months later with exertional chest pain. A repeat cardiac catheterization demonstrated marked in-stent restenosis treated with laser debulking and balloon angioplasty (►Fig. 1c, d). At 1 year follow-up, the patient was symptom free and doing well.

Discussion

SCAD is an infrequent but well-recognized clinical entity which can present as ACS, syncope, or heart failure. The prevalence of SCAD is reported to be 0.10 to 0.24% of all patients undergoing coronary angiography for stable coronary artery disease or ACS.² Most SCAD, however, is thought to present as sudden cardiac death. Although in recent decades both the number of reported cases and the survival have increased, SCAD is likely underreported and underdiagnosed.³

The pathophysiology of SCAD involves formation of a hematoma in the outer layer of media forming a false lumen with or without the formation of intimal tear.⁴ Etiologies include atherosclerosis, cocaine use, vasculitis, fibromuscular dysplasia, or vigorous exercise (►Table 1). Two-thirds of cases are reported in women.⁵ The right coronary artery is more frequently involved in men, while in women the left anterior descending artery is mostly involved.^{3,6} Heterozygous factor V Leiden gene mutation has been reported in young patients with myocardial infarction.^{7,8} There are no published case reports of SCAD with factor V Leiden mutation.

Table 1 Etiology of spontaneous coronary artery dissection

Pregnancy	Coronary artery spasm
Atherosclerosis	Remote aortic valve replacement
Cocaine use	Acute emotional stress
Retching	Contraceptives
Vigorous exercise	Septic emboli
Marfan syndrome	Triptan use
Ehlers–Danlos IV syndrome	Hyperthyroidism
Systemic lupus erythematosus	Autoimmune thyroiditis
Kawasaki disease	Prednisone use with rheumatoid arthritis
Takayasu arteritis	Drug hypersensitivity
Angitis	Active hepatitis C with cryoglobulinemia
Coronary ectasia	Renal transplantation
Apical hypertrophic cardiomyopathy	Bromocriptine
Fibromuscular dysplasia of coronary arteries	Rheumatic mitral stenosis

Our case is unique in that SCAD occurred in an otherwise young healthy male with factor V Leiden mutation which is a potential new risk factor for SCAD. We can only hypothesize the existence of a causal association between the factor V Leiden mutation and the spontaneous coronary dissection in our patient, but we cannot prove it. However, two previously published case reports describe an association of factor V Leiden mutation with carotid artery dissection.^{9,10}

Treatment options for SCAD include medical management, early revascularization with percutaneous intervention (PCI), or coronary artery bypass grafting (CABG). CABG is preferred for patients with left main coronary artery or multivessel dissection. PCI is preferred for isolated single vessel dissection. In cases of inoperable left main or multivessel dissection, bridging to transplant can be considered.

Suggested medical management include nitrates, β blockers, and calcium channel blockers to reduce coronary vasospasm. Aspirin, clopidogrel, and low-molecular-weight heparin have been recommended to limit the extension of hematoma. Medical treatment is preferred in cases with less than 50% luminal stenosis and Thrombolysis in Myocardial Infarction 2 to 3 flow.¹ The use of glycoprotein IIb/IIIa inhibitors has been reported.¹¹ Thrombolytics have been used for ST-segment elevations that eventually were diagnosed as SCAD, however, more adverse side effects including bleeding and dissection extension have been reported.¹² In one case, immunosuppression (with prednisone and cyclophosphamide) was reported for the treatment of SCAD.¹³

Early revascularization has better outcomes.⁵ Twenty-one percent of patients develop a second dissection within 2 years. One-fifth of patients treated conservatively eventually require revascularization.

Conclusion

SCAD is a rare disease that can present as ACS, syncope, or heart failure in young patients. The long-term outcome is better in patients treated with early revascularization.

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