

Hereditary Spontaneous Coronary Artery Dissection

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Citation: Kumar M, Chaturvedula S, Meng J (2018) Hereditary Spontaneous Coronary Artery Dissection. J Case Rep Stud 6(3): 307

Received Date: May 29, 2018 Accepted Date: June 25, 2018 Published Date: June 27, 2018

Abstract

Introduction: SCAD with an estimated prevalence of 1.7% to 4% is an important cause of Myocardial infarction especially in middle-aged women with no or minimal cardiovascular risk factors. Diagnosis requires a high degree of suspicion and better coronary imaging. The hereditary/ genetic component has been postulated to play a role, but there is insufficient data on familial SCAD. We present a case of SCAD with probable genetic and autoimmune association.

Case Description: A 42-year-old female with past medical history of asthma and Hashimoto's thyroiditis presented to Emergency Department with sudden onset of intense left-sided chest heaviness, precipitated by emotional stress in a courtroom. Her family history was significant for vasospastic angina in her mother at the age of 38 years. Coronary angiography of her mother was normal. On examination, she was in moderate distress due to chest pain. She was tachycardic and hypertensive at the time of presentation. Neck veins were not distended. Cardiac examination revealed tachycardia with normal heart sounds without any murmurs or gallops. Rest of the examination was unremarkable. Initial Electrocardiogram revealed sinus tachycardia with no ischemic changes. She received aspirin and Cardiac enzyme panel showed Troponin of 15.87 ng/mL (0.05-0.49 ng/mL) and CK-MB of 89 ng/mL (0-8 ng/mL). She experienced the recurrence of pain in the emergency department, and repeated EKG showed ST-segment elevation in inferior leads. Immediate coronary angiography revealed dissection of the first obtuse marginal vessel, confirmed by Intravascular ultrasound (IVUS). Due to the presence of unopposed hematoma, distal and proximal vessels were stented using two drug-eluting stents with the restoration of brisk blood flow. Left ventriculography demonstrated inferior wall hypo-kinesis with low normal left ventricular ejection fraction. The patient was discharged after four days of observation on Aspirin, Ticagrelor, Metoprolol, and Atorvastatin.

Discussion: Diagnosis of SCAD is becoming more common due to increased awareness and better coronary imaging modalities. Our patient's mother had normal coronary angiography, and her symptoms were attributed to vasospastic angina in an era when knowledge of SCAD was limited due to non-availability of advanced coronary imaging. Both patient and her mother had a similar presentation without any traditional cardiovascular risk factors which raises suspicion of the genetic/ hereditary component. Her mother had Addison's disease, and the patient had autoimmune thyroiditis which also raises suspicion of autoimmune association. A high index of suspicion is required for diagnosis which can be confirmed by coronary imaging. Stenting is usually not performed as these heal spontaneously. Hypertension has been linked with recurrent SCAD.

Keywords: Hereditary; Auto-immune; Coronary Artery Dissection

Introduction

SCAD with an estimated prevalence of 1.7% to 4% is an important cause of Myocardial infarction especially in middle-aged women with no or minimal cardiovascular risk factors. Diagnosis requires a high degree of suspicion and better coronary imaging. The hereditary/ genetic component has been postulated to play a role, but there is insufficient data on familial SCAD. We present a case of SCAD with probable genetic and autoimmune association.

Case Report

A 42-year-old female presented to Emergency Department with sudden onset of intense, left-sided chest heaviness, precipitated by emotional stress in a courtroom. She reported paroxysms of intermittent chest pain with associated shortness of breath, back spasms, and diaphoresis in the week prior to this presentation. Her symptoms were not relieved by nitroglycerin but partially alleviated by opiates. Her medical history included well-controlled asthma and Hashimoto's thyroiditis but notably negative for traditional risk factors of atherosclerotic cardiovascular disease such as hypertension, hyperlipidemia, Diabetes mellitus, etc.

The patient did not use any prescription or over the counter medications or supplements on a regular basis. She denied use of tobacco, alcohol or any other illicit drugs. Her mother was diagnosed with vasospastic angina at the age of 38. Her father had an Acute Myocardial Infarction (MI) in his 50s.

On examination, she was in moderate distress due to chest discomfort. The Heart Rate was 117 /min, BP of 167/97 mm Hg, respiratory rate 20 breaths/ minute and 98% saturation on ambient air. The central venous pressure was not elevated, lungs were clear to auscultation. Cardiac examination revealed tachycardia with normal heart sounds without any murmurs or gallops. Rest of the examination was unremarkable.

Chest x-ray was normal. Electrocardiogram (EKG) was significant for sinus tachycardia with a normal rhythm, axis, intervals, and no ischemic changes. Complete blood count (CBC) and Basic metabolic profile (BMP) were within normal limits. Cardiac enzyme panel showed Troponin of 15.87 ng/mL (0.05-0.49 ng/mL) and CK-MB of 89 ng/mL (0-8 ng/mL).

She received aspirin and had a recurrence of similar chest pain in the emergency room. Repeat EKG was remarkable for ST-segment elevation in the lead II, III and aVF, V5 and V6 with T-wave inversion in the lead III (Figure 1). She received nitroglycerin, morphine, and heparin. However, she continued to have intractable chest pain prompting emergent cardiac catheterization.

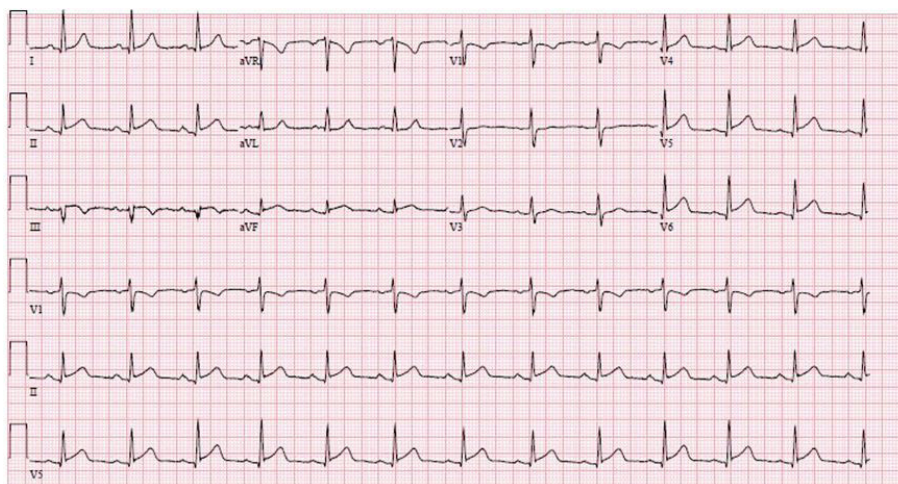


Figure 1: Repeat EKG showing ST segment elevation in lead II, III and aVF, V5 and V6 with inverted T-waves in lead III

Coronary angiogram revealed no atherosclerotic lesions but demonstrated a dissecting lesion in the first obtuse marginal vessel (Figure 2), confirmed by intravascular ultrasound (IVUS) (Figure 3). This lesion was treated with a drug-eluting stent (DES) and an additional stent was placed in the proximal artery as it had evidence of unopposed hematoma on IVUS. The procedure was successful in restoring brisk flow, and with no residual dissection, perforation (Figure 4). Left ventriculography demonstrated inferior wall hypokinesia with low normal left ventricular ejection fraction. The patient had an uneventful hospital stay. She remained symptom-free and was discharged after four days with a prescription for dual antiplatelet therapy (Aspirin, Ticagrelor), Metoprolol and Atorvastatin.



Figure 2: Coronary Angiography showing Coronary artery dissection in first obtuse marginal vessel

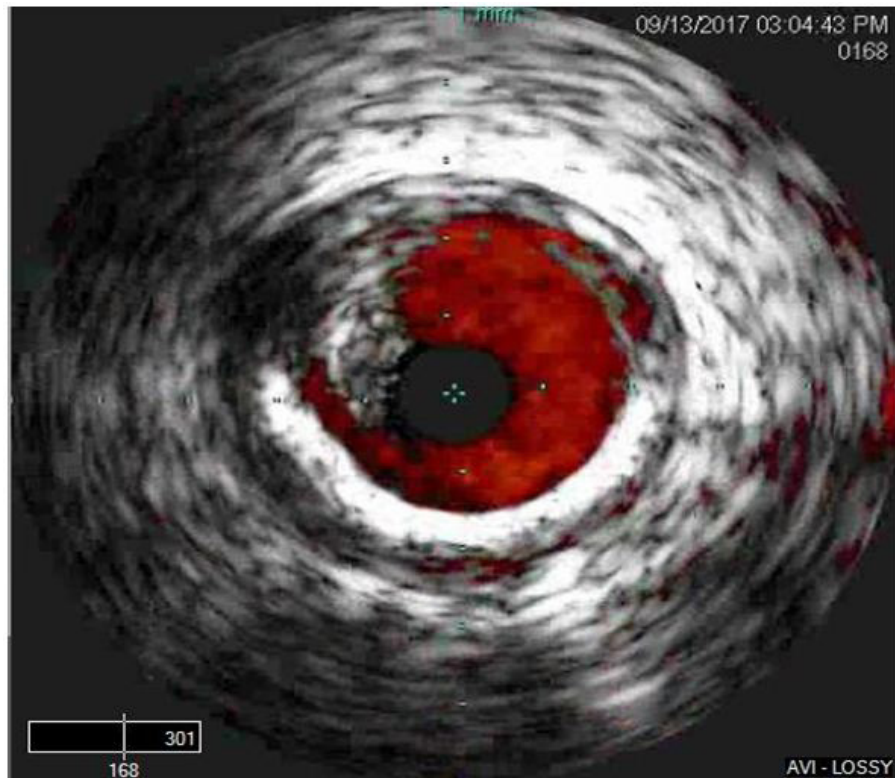


Figure 3: IVUS demonstrating artery dissection seen on coronary angiography

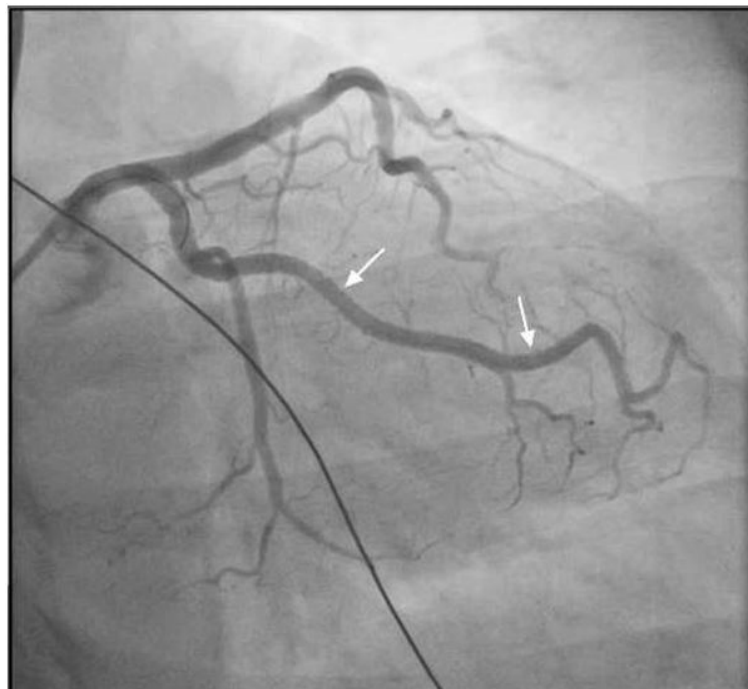


Figure 4: Good restoration of blood flow after insertion of drug eluting stents

We subsequently learned that patient's mother at the age of 38 years (1986) had presented to the emergency department with excruciating chest pain radiating to left shoulder associated with diaphoresis. We obtained her medical records from an outside hospital. Her past medical history was reported as seizures due to Addison's disease for which she was on steroids. Her examination was remarkable for hypotension. EKG demonstrated normal sinus rhythm with poor R wave progression across precordial leads. Her pain resolved with vasodilator therapy. She was admitted for further workup and next day she had a recurrence of similar chest pain. Repeat EKG was consistent with anterolateral wall MI with the elevation of cardiac biomarkers. Coronary angiography did not have any evidence of atherosclerotic disease &/or spasm. Left ventriculography revealed enlarged left ventricle with a decreased ejection fraction, akinetic anterior wall. Her anterior wall MI was attributed to probable spasm. A stress test was performed during

hospitalization for further risk stratification, and it did not reveal ischemia. She was discharged on Diltiazem and Aspirin with the resolution of her chest pain and later regained normal left ventricular function in the months after her presentation.

Discussion

Our patient presented with suspected MI at a young age despite a low coronary risk profile. Her presentation, angiographic findings, and family history raise the possibility of a familial/hereditary coronary artery dissection and autoimmune association. Her mother's diagnosis of "vasospastic angina" was made in an era when SCAD was an underappreciated entity due to lack of intravascular imaging. Similar to our patient, her mother did not have traditional risk factors for atherosclerotic cardiovascular disease, and we presume that her symptoms may have been due to dissection that was not recognized due to her presentation in the setting of emotional stress.

There is a lack of data regarding familial SCAD. We found single case series reporting five families having SCAD. Both recessive and dominant pattern of inheritance with incomplete penetrance and variable expression were found [1]. Female gender, fibromuscular dysplasia, hormonal influences and connective tissue disorders have been implicated as potential predisposing causes [1-3].

SCAD occurs predominantly in middle-aged women from 44 to 55 years of age with no or minimal cardiovascular risk factors [4,5]. Recent series indicate an estimated prevalence of 1.7% to 4% [6]. Patients usually have a precipitating stressor with underlying vulnerability such as intense exercise, emotional stress, ingestion of certain drugs such as cocaine, labor, and delivery, intense hormonal therapy such as beta-HCG, corticosteroid therapy, etc. Most patients present with chest pain and elevation of cardiac enzymes [1-3,7]. However; a wide spectrum of presentations have been reported including Ventricular arrhythmia (3 to 10%), cardiogenic shock (<3%) and sudden cardiac death (<1%) [4,8-10]. Less frequent presenting symptoms include pain in the arms or neck, nausea or vomiting, diaphoresis, dyspnea, and back pain.

In a young patient with personal or family history of fibromuscular dysplasia, connective tissue diseases, presenting with chest pain following emotional stress and physical exertion, SCAD should be considered [4,11].

Initial EKG may be normal or may have ST-segment elevations (24-87%) [2,4]. Coronary angiography with selective use of advanced intracoronary imaging techniques, including optical coherence tomography(OCT) and intravascular ultrasonography(IVUS) is the gold standard for diagnosis of SCAD [12]. There are three pathognomonic appearances on coronary imaging of which diffuse smooth stenosis is the most common angiographic manifestation [9,13]. Management options include conservative therapy or revascularization measures. Conservative management has generally been associated with favorable outcomes as SCAD heals spontaneously [8,14,15]. Beta blockers have a protective effect, and along with Aspirin are routinely used for acute and long-term management. Clopidogrel is often administered for 1 to 12 weeks and then discontinued if no further pain or angiographic healing is demonstrated [7]. ACE inhibitors are used for cases having evidence of left ventricular dysfunction. Statins are recommended if hyperlipidemia is present [1,4]. Adequate blood pressure is recommended since hypertension has been associated with recurrent SCAD [16]. Cases managed conservatively should be monitored in hospital for 3 to 5 days due to the risk of early progression of dissection which may need coronary intervention [4]. Repeat angiography to confirm healing is not advised as vessels in these patients are more prone to rupture because of fragility unless strong clinical indication is present. The prevalence of extra-coronary vasculopathy, such as fibromuscular dysplasia, aneurysms, and dissections, is high in patients with SCAD, screening of the brain, neck, and visceral vessels with computed tomographic angiography or magnetic resonance angiography are advised [4,7,12].

Percutaneous interventions with stenting yield markedly lower success rates in SCAD as compared to atherosclerotic ACS [14,15]. It should be considered in cases that have ongoing ischemia or chest pain, hemodynamic instability, ventricular arrhythmia and the left main dissection. Emergent CABG should be considered for patients with left main dissections, extensive dissections involving proximal arteries, or in patients in whom PCI failed or who are not anatomically suitable for PC such as patients having multi-vessel atherosclerotic coronary artery disease.

Cardiac rehabilitation should be recommended to all SCAD patients. A dedicated program including exercise rehabilitation, psychosocial counseling, and peer group is beneficial for SCAD patients who often have substantial anxiety, depression, and ongoing physical symptoms [1,7,17]. SCAD, patients are often advised to avoid weightlifting, bodybuilding, competitive racing, or athletic pursuits at high-intensity levels and instead are encouraged to pursue moderate daily aerobic activity and resistance training with low weight and more repetitions [1,15].

Conclusion

SCAD is an infrequent but fatal condition presenting most commonly with chest pain similar to an acute coronary syndrome. A high index of suspicion is vital for accurate diagnosis. The typical patient is middle-aged female without traditional or minimal cardiovascular risk factors. Family history, probable genetic predisposition, and presence of autoimmune/ connective diseases facilitate better risk stratification for accurate diagnosis & treatment of MI or sudden death of unclear etiology in patients with

SCAD. Conservative therapy is the cornerstone, as most lesions are known to heal spontaneously. However, recurrences may still occur in up to 10-20% of cases. Revascularization remains particularly challenging and may be associated with suboptimal results due to acute complications, and poor long-term outcome. Patients with SCAD may also benefit from formal medical genetics consultation for familial assessment and genetic panel testing since it has been associated with genetic connective tissue diseases [1,10]. The effectiveness of integrated rehabilitative services involving exercise rehabilitation, psychosocial counseling, and peer groups is emerging.

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