Case Report

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Diagnosis and management of spontaneous coronary artery dissection: Two cases and a review of the literature

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Introduction and importance: Spontaneous coronary artery dissection (SCAD) is a rare but potentially fatal condition, often underdiagnosed despite its significance in acute coronary syndrome (ACS). The true prevalence remains uncertain due to diagnostic challenges. Identifying SCAD cases is crucial for reducing mortality and morbidity, especially considering the recurrence risk. The authors present two cases highlighting the importance of multimodality imaging in diagnosing and managing SCAD.

Case presentation: Case 1: A 53-year-old man with a history of brain aneurysm presented with chest pain and shortness of breath. Despite negative EKGs and stress tests, coronary computed tomography angiography (CCTA) revealed non-obstructive dissection flaps. Medical management improved his condition. Case 2: A 55-year-old woman with no significant medical history experienced recurrent chest pain. Initial tests were negative, but CCTA revealed SCAD. Further screening uncovered undiagnosed fibromuscular dysplasia.

Clinical discussion: SCAD poses diagnostic challenges, often mimicking other cardiac conditions. Traditional tests may yield negative results, necessitating advanced imaging techniques like CCTA. Recognizing SCAD's association with connective tissue disorders (CTD) is vital for comprehensive patient care. The authors' cases emphasize the importance of a systematic approach to diagnosing chest pain, including noninvasive modalities and considering underlying etiologies.

Conclusion: SCAD diagnosis requires a high index of suspicion, especially when traditional cardiac tests are inconclusive. Beyond treatment, patients should undergo further evaluation for CTDs, particularly in those with minimal risk factors for atherosclerosis. Increased awareness and a multimodal diagnostic approach are crucial for timely intervention and improved outcomes in SCAD patients.

Learning objectives: The authors aim to increase awareness regarding different clinical presentations of SCAD to decrease the risk of missed or late diagnosis. The authors' case series also signifies the multimodal imaging approach's role in evaluating chest pain. Upon diagnosis of SCAD, it is imperative to go beyond treatment and implement a reverse algorithmic strategy to discover any underlying causes and risk factors for SCAD predisposition.

Keywords: acute coronary syndrome (ACS), case reports, chest pain, connective tissue disorders (CTD), coronary computed tomography angiography (CCTA), multimodality imaging, spontaneous coronary artery dissection (SCAD)

Introduction

Spontaneous coronary artery dissection is an acute non-iatrogenic tear in the coronary artery wall compressing the coronary lumen and possibly causing myocardial infarction or, in some cases, sudden death. It can present with different clinical presentations and may have various risk factors associated with it. The true prevalence of SCAD remains uncertain, primarily because it is an underdiagnosed condition. Missed diagnoses are driven by a low suspicion, even in the presence of classic presenting symptoms, limitations of current coronary angiographic

HIGHLIGHTS

- Acute coronary syndrome (ACS) should be considered in the differential diagnosis for patients with connective tissue disease, particularly when conventional diagnostic methods yield negative results.
- Multimodal imaging, like CCTA, is crucial for diagnosis, especially when traditional tests are negative.
- Management involves medical therapy and investigation for underlying conditions like CTD.
- A reverse algorithmic approach post-diagnosis aids in uncovering predisposing factors for SCAD.

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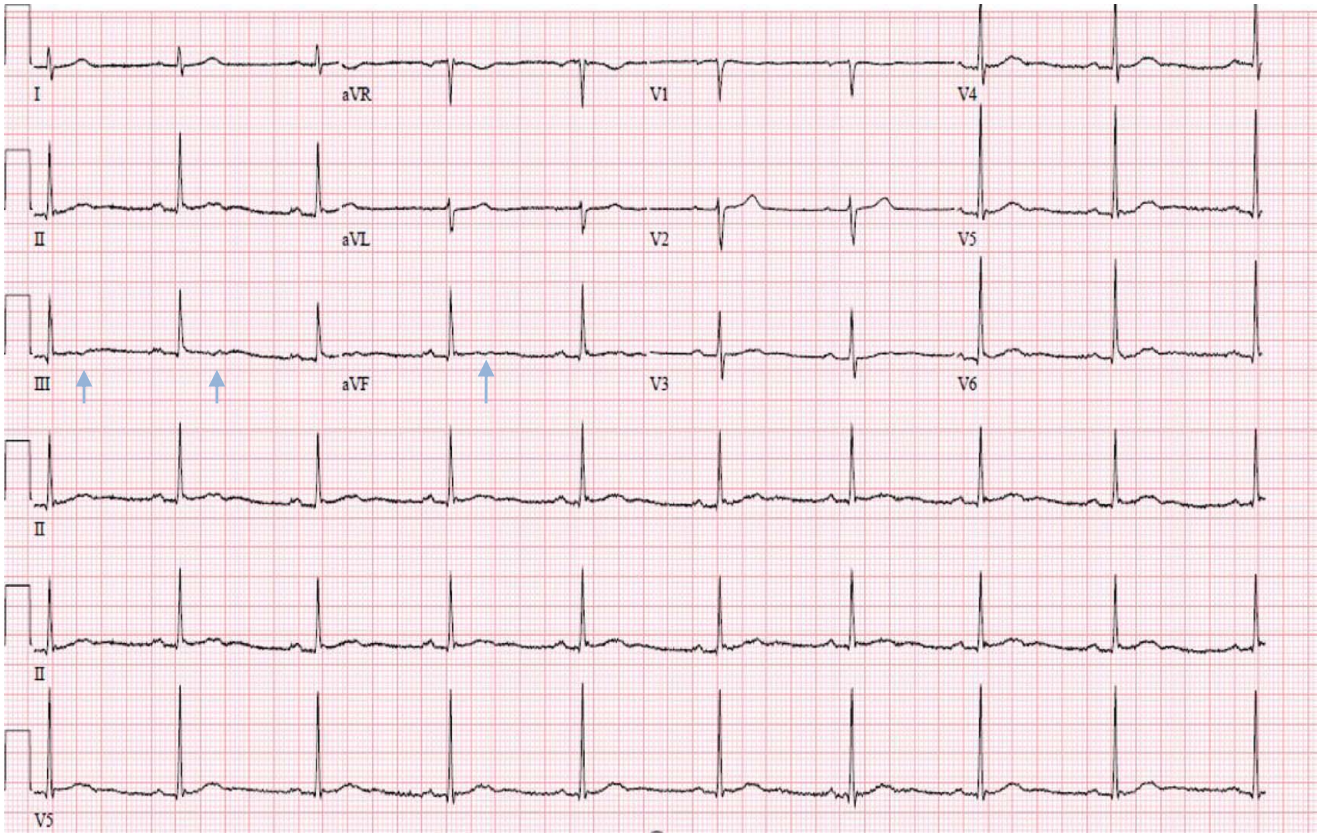

Figure 1. EKG demonstrating sinus bradycardia and nonspecific T-wave abnormalities.

techniques, and lack of clinician familiarity with the condition. SCAD most commonly occurs in patients with few or no traditional cardiovascular risk factors. Studies suggest that SCAD accounts for 1–4% of cases of ACS overall[1]. Recent data showing a high prevalence of extra coronary arterial abnormalities in SCAD patients suggest that SCAD may be an organ-specific manifestation of a systemic vascular disorder or arteriopathy[2]. Coronary computed tomography angiography (CCTA) is becoming more commonly used as a 'triple rule-out computed tomography (CT) protocol for evaluating acute aortic, coronary, and pulmonary syndromes. It could be a valuable, noninvasive technique for assessing patients with SCAD, as traditional coronary angiography has a higher risk of propagating the dissection.

Presentation: Case 1

History

A 53-year-old male with a history of type two diabetes mellitus and remote history of epilepsy s/p brain aneurysm excision presented in the clinic for evaluation for chest pressure and shortness of breath. He described his chest pressure as retrosternal, progressive over three months. His pain was exertional, non-radiating, non-positional, and non-respirophasic, relieved by rest. He noticed occasional worsening with emotional stress. He denied any smoking or alcohol use. Physical examination was unremarkable.

Investigation

He had an exercise tolerance test, which was negative for ischemia, followed by a pharmacologic stress test, which was negative as well. Initial laboratory tests were unremarkable. ECG showed sinus bradycardia and nonspecific T-wave abnormality—Figure 1.

Trans-thoracic echocardiogram showed mild concentric left ventricular hypertrophy with normal Left ventricular systolic function and ejection fraction of 62%. There were no segmental left ventricular wall motion abnormalities noted. A repeat pharmacological stress test showed no acute ischemic changes. Due to the typical nature of the chest pain, he underwent computed tomography angiography of coronary arteries that showed non-obstructive dissection flaps within the left and right coronary arteries—Figure 2.

However, no pieces of evidence of acute obstructive features were seen. There was only mild narrowing of the left anterior descending due to an intramural hematoma; thus, he was diagnosed with Spontaneous coronary artery dissection.

Treatment

The decision was made to treat the patient medically. He was started on Aspirin 81 mg daily and Metoprolol 50 mg daily. Non-pharmacologic interventions included educating the patient to avoid strenuous activities. The patient was doing well at the 1-month follow-up in the cardiology clinic. Given the patient history of cerebral aneurysm and coronary artery dissections on the

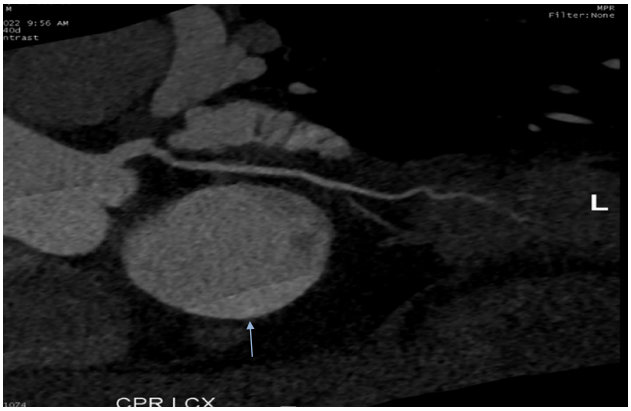

Figure 2. Computed tomography angiography of the coronary arteries revealing non-obstructive dissection flap

CT scan, he underwent scanning of the neck, abdomen, and pelvis vessels to evaluate for possible underlying CTD. CT angiogram (CTA) showed splenic artery aneurysms with subsequent workup with rheumatology and was diagnosed with fibromuscular dysplasia.

Presentation: Case 2

A 55-year-old former smoker female with no significant past cardiac history presented in the office after an episode of sudden onset of chest pain. She started to experience substernal chest heaviness, which was exertional, relieved on rest with radiation to her back associated with shortness of breath for 1 month. She had initial testing, including an exercise stress test, EKG, and troponins, which were unremarkable. Eventually, the pain started evolving with pain at rest for extended periods, and she was referred to the outpatient cardiology clinic for further investigation. She denied any family history of premature coronary disease or sudden deaths.

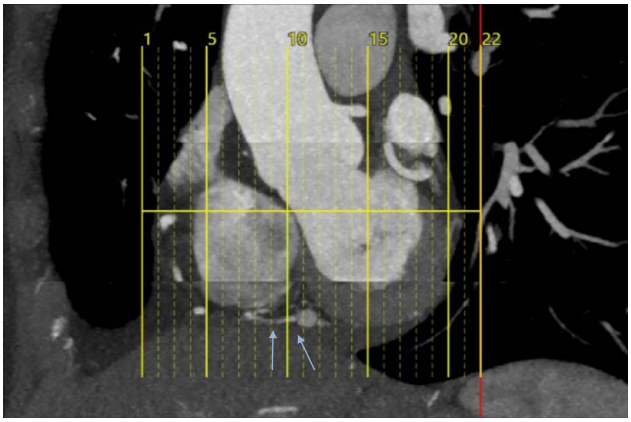

Figure 3. Coronary CTA demonstrating spontaneous coronary artery dissection (SCAD)

Physical examination was within normal limits

Investigation

Repeat exercise stress test and EKG were unremarkable. An echocardiogram was done, which showed normal left ventricular size and normal systolic function, with an estimated LVEF of 60%. Coronary CTA was done that showed SCAD as shown in Figure 3.

Treatment

The patient was started on Aspirin 81 mg and Metoprolol succinate 50 mg daily. She continued to have episodes of recurrent chest pain, and given she could not tolerate Isosorbide Mononitrate, she was started on Ranolazine 1000 mg twice daily. On 6-month follow-up, coronary CTA was repeated, and the changes remained stable. Further workup revealed a fusiform celiac artery aneurysm, bilateral iliac ectasia, and bilateral renal artery dissections, and the patient is currently being worked up for underlying CTD given the extensive nature of vascular pathologies.

Discussion

Our case series patients presented with symptoms indicative of ACS, yet their troponin, EKG, and stress test outcomes yielded negative results. Due to recurrent chest pain and a possibility of high suspicion angiography was indicated. CCTA was ordered over traditional angiography because it is less invasive and provides detailed, high-resolution images of the coronary arteries and surrounding structures. The results showed the presence of SCAD.

The most common causes of chest pain in the primary care population include chest wall pain (20%); reflux esophagitis (13%); costochondritis (13%); and other non-cardiac causes[2]. Even though Cardiac disease remains the leading cause of death in the United States, only 1.5% of patients presenting with chest pain to a primary care office will have unstable angina or an acute myocardial infarction (MI)[3]. SCAD has emerged as an important cause of ACS, comprising 1–4% of all ACS cases, although the incidence of SCAD detected in coronary angiograms ranges from 0.1 to 1.1%[4]. These numbers highlight the rarity of prevalence and the challenges of its timely diagnosis.

SCAD is more common in women and is associated with emotional stress, fibromuscular dysplasia (FMD), inherited arteriopathy, chronic kidney disease (CKD), pregnancy, childbirth, use of oral contraceptives, and carotid artery disease[5]. The pathophysiology could be more precise, and clinical presentation usually demonstrates great variability. Three types of spontaneous coronary artery dissection have been recognized: atherosclerotic, puerperal, and idiopathic, which require careful screening. Idiopathic causes comprise a considerable number in etiology[6].

Given the rarity of this diagnosis and less prevalence in males, it can be overlooked if not associated with acute coronary syndrome. Additionally, if it presents in the outpatient setting, it can be missed, mainly when stress testing is negative, as it was in both of our cases. Our case series proposes systematic approaches to the diagnosis and workup of the patients presenting with chest pain using different noninvasive modalities in a step-wise fashion even after the initial testing is negative when the patient continues to be symptomatic. Despite that, the traditional approach

through stress testing showed no abnormality; using coronary CTA proved vital to detect SCAD[7]. Coronary angiograms and intravascular ultrasounds can also provide helpful diagnostic information; however, CTA is a noninvasive technique that should be adopted first while exploring the atypical cases of chest pain before invasive strategies. Overall, there is a lack of consensus on investigating algorithms and treatment guidelines, highlighting the need for further research to increase the diagnosis rate and prevent adverse outcomes.

The author also stresses that the investigation should continue beyond the diagnosis of SCAD. Instead, patients should also be worked up for the presence of CTDs. Fibromuscular dysplasia in one of the most common causes of SCAD found in literature. In patients with suspected acute coronary syndrome (ACS) and inconclusive diagnostic tests, a rheumatology-centered physical examination is crucial. Since connective tissue diaseases (CTD) are often rheumatologic, this exam can reveal insight missed by standard care. Cardiologist should look for signs like joint swelling, skin changes, Raynaud's phenomenon, sclerodactly, and muscle weakness. Recognizing these features can guide a broader differential diagnosis.

Only a minority of patients with SCAD who undergo genetic evaluation have a likely pathogenic mutation identified on gene panel testing for CTD—even fewer exhibit clinical features of connective tissue disorder. In a study done spanning the year 1984–2014 for patients of SCAD, Of the 116 patients, 59 patients underwent genetic testing, of whom 3 (5.1%) received a diagnosis of connective tissue disorder[8]. Given its small number, there is a significant trend of managing the disease alone, leading to a high risk of adverse outcomes and an increase in the prevalence of underdiagnosed cases of SCAD secondary to CTDs. Our cases emphasize a multimodal approach in diagnosing chest pain in the ambulatory setting and adopting a reverse algorithmic methodology for investigating the etiology rather than taking it as an isolated diagnosis for timely prevention of adverse outcomes.

Management of Spontaneous Coronary Artery Dissection (SCAD) primarily involves medical therapy for clinically stable patients, emphasizing antiplatelet therapy with aspirin and possible P2Y12 inhibitors. Revascularization (PCI or CABG) is reserved for high-risk cases like left main coronary artery dissection, ongoing ischemia, hemodynamic instability, or refractory arrhythmia. Beta-blockers are used to reduce vessel wall stress, and thrombolysis is contraindicated due to the risk of exacerbating dissection. Mechanical circulatory support may be considered cautiously in SCAD complicated by cardiogenic shock[9].

Method

This case has been reported in Line with CARE guidelines for case reports.

Conclusions

SCAD is a relatively rare diagnosis and tends to be overlooked, particularly if not associated with ACS or has negative traditional cardiac testing (EKG, stress test, troponins). Physicians should consider this etiology of chest pain, especially when the presentation is unusual. Once diagnosed and treated, patients should undergo further testing for connective tissue disorders, especially in middle-aged people with minimal risk factors for atherosclerosis.

Ethical approval

NA as we are not submitting study and our case report does not deal with any original human data or animal data.

Consent

Informed consent was obtained from the patients discussed in this case series. They have received a copy of the informed consent form and signed it.

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Author contribution

A.A.: research, manuscript development, data collection, interpretation, conception and design. K.A.: research, manuscript development, data collection, interpretation, conception and design. F.A.: research, manuscript development, data collection, interpretation. R.L.: research, manuscript development, data collection, interpretation. M.A.M.: manuscript development, data collection, interpretation. M.W.K.: research, manuscript development, data collection, interpretation.

Conflicts of interest disclosure

None of the authors have a conflict of interest to disclose concerning our manuscript.

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Guarantor

Khurram Arshad is the name of the guarantor who accepts full responsibility for the work and/or the conduct of the study, had access to the data, and controlled the decision to publish.

Data availability statement

Not applicable.

Provenance and peer review

None.

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