CATHALOGUE

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CASE REPORT

Mysteriously Mysterious MI: Recurrent ACS Secondary to Spontaneous Coronary Artery Dissection Associated with Rheumatoid Arthritis.

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Abstract

Background: Spontaneous coronary artery dissection (SCAD) is a rare but significant cause of myocardial infarction (MI), particularly in young females who often lack traditional cardiovascular risk factors. This condition is characterized by a tear in the coronary artery wall, which can lead to serious cardiac events.

Case Presentation: We present the case of a 36-year-old female who experienced recurrent episodes of myocardial infarction. Her clinical presentation included severe chest pain, ECG abnormalities, and angiographic findings consistent with coronary dissection. During her evaluation, she was also diagnosed with rheumatoid arthritis (RA), which was found to be an incidental but relevant finding in her medical history.

Results: Despite appropriate treatment, the patient continued to experience recurrent cardiac events, underscoring the chronic nature of her condition. The repeated episodes of MI were ultimately attributed to SCAD. The interplay of her chronic inflammatory state, autoimmune vasculitis, connective tissue abnormalities, and potential medication effects from RA were considered contributing factors to the development of SCAD.

Conclusion: This case underscores the challenges of managing spontaneous coronary artery dissection (SCAD) in a young woman with rheumatoid arthritis. It highlights the importance of thorough diagnostic evaluation and ongoing monitoring due to recurrent acute coronary syndrome episodes. Early identification and effective medical management, including dual antiplatelet therapy and anticoagulation, were crucial for favorable outcomes.

Keywords

Spontaneous Coronary Artery Dissection, Myocardial Infarction, Rheumatoid Arthritis, Coronary Angiography.

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Introduction

Acute myocardial infarction (MI) is typically linked to coronary artery disease (CAD). However, a growing number of younger female patients without traditional cardiovascular risk factors such as hypertension, diabetes, or hyperlipidemia may instead be experiencing spontaneous coronary artery dissection (SCAD). This condition involves a non-atherosclerotic disruption of the coronary artery's intima, caused by spontaneous weakening or tearing of the vessel wall. This leads to blood seeping into the arterial layers, creating a false lumen and potentially compressing the true lumen due to intramural hematoma, ultimately compromising myocardial blood supply.[1]

The exact etiology of SCAD remains largely unknown, but it is thought to involve several factors, including underlying connective tissue disorders like fibromuscular dysplasia, hormonal fluctuations associated with pregnancy or perimenopause, strenuous physical activity, and emotional stressors. Clinically, SCAD presents similarly to typical MI, featuring symptoms such as chest pain, dyspnea, and ECG abnormalities.

Case Presentation

A 36-year-old female presented to the emergency room with sudden onset of severe chest pain radiating to her left arm, accompanied by profuse sweating and multiple episodes of vomiting over the past four hours. Following her second episode of acute coronary syndrome (ACS), a detailed history revealed that she had been experiencing morning pain, redness, and swelling in the wrists and small joints of her hands for the past thirteen years. These symptoms were relieved by activity and were not associated with mouth ulcers, skin rashes, or photosensitivity.

On examination, she appeared mildly obese, with a puffy and plethoric face, hirsutism, and pink abdominal striae, while her vital signs remained normal. A systemic examination revealed wasting of the thenar and hypothenar muscles in her hands, along with swelling and tenderness in both wrists and the proximal interphalangeal joints, resulting in restricted movement.

The patient experienced two episodes of ACS, with the second occurring exactly five months after the initial presentation.

Diagnostic Assessment

During her initial presentation, an ECG and bedside echocardiogram revealed regional wall motion abnormalities (RWMA), prompting a coronary angiography that identified single-vessel coronary artery disease. A thrombus-laden left anterior descending artery (LAD) was found, causing critical obstruction in its mid-course. Further 2-D echocardiography showed an ejection fraction (EF) of 35%, indicating moderate left ventricular (LV) systolic dysfunction, along with Grade I diastolic dysfunction, mild mitral regurgitation (MR), and an organized apical clot.

Upon her second presentation with ACS, repeat angiography was conducted due to persistent ECG abnormalities and RWMA on echocardiogram. This revealed a subtotal occlusion in the proximal LAD with a substantial thrombus extending into the left main stem, resulting in Thrombolysis in Myocardial Infarction (TIMI) grade II distal perfusion. Notably, the previous angiographic findings in the mid LAD had completely healed with minimal residual disease, leading to an admission for anticoagulation treatment under the diagnosis of Unstable Angina Pain (USAP)/Non-ST Elevation Myocardial Infarction (NSTEMI) secondary to spontaneous coronary artery dissection (SCAD). The 2-D echocardiogram at this stage displayed an LV apical clot measuring 10x18 mm.

All baseline laboratory tests, including thyroid profile and C-reactive protein (CRP), were normal, except for an elevated erythrocyte sedimentation rate (ESR) of 75 mm/hr. Given her history of joint pain and swelling alongside the elevated ESR, a comprehensive immune profile for medium-sized vasculitides was conducted. The results were negative for antibodies associated with conditions such as systemic lupus erythematosus (SLE),

Sjögren's syndrome, and systemic sclerosis, but revealed a raised antinuclear antibody (ANA) titer of 1:160 (N: <1:80), a Rheumatoid Factor (RF) of 99.3 IU/ml (N <14 IU/ml), and an Anti-Cyclic Citrullinated Peptide (CCP) antibody level exceeding 500 U/ml (N: <17.0).

Given the established association between SCAD and fibromuscular dysplasia, Doppler studies of the carotid, renal, and lower limb arteries were performed, all returning normal results.2 Late Gadolinium Enhancement (LGE) imaging confirmed the presence of the LV apical clot and highlighted subendocardial to transmural myocardial enhancement. Although a further contrast angiography with optical coherence tomography (OCT) was considered to further elucidate the vascular anatomy, it was ultimately deemed unnecessary due to the patient's asymptomatic status and resolution of the previously identified lesion. A conventional coronary angiography, conducted after appropriate anticoagulation, showed a dramatic resolution of all previous abnormalities.

Ultimately, the diagnosis of spontaneous coronary artery dissection was confirmed, supported by her young age, complete healing of the mid LAD lesion during the second episode, involvement of the previously normal proximal LAD, and significant improvement observed in repeat angiography. A rheumatology consultation was obtained, resulting in a diagnosis of rheumatoid arthritis, correlating with her positive immune profile findings.

Therapeutic Intervention

During the initial event, the patient's chest pain resolved during angiography, and following ECG monitoring, she was started on tirofiban, an antiplatelet medication. Once recirculation was established, she was discharged on dual antiplatelet therapy (DAPT), rivaroxaban as an anticoagulant, and guideline-directed medical therapy (GDMT) including beta-blockers, mineralocorticoid receptor antagonists, angiotensin receptor neprilysin inhibitors, and sodium glucose cotransporter-2 inhibitors for four weeks.

After the second episode of ACS, the patient was initially anticoagulated during her hospital stay. Upon concluding the diagnosis of SCAD, she was discharged on DAPT, anticoagulants, GDMT, and immune-modulating drugs such as methotrexate with folic acid for her rheumatoid arthritis. Corticosteroids were discontinued due to their associated risks with dissection.

Follow-Up and Outcomes

At the two-month follow-up after the first episode, echocardiography revealed an improvement in ejection fraction (EF) to 50% and the dissolution of the left ventricular (LV) apical clot. However, following the second myocardial infarction (MI), the EF declined again to 35%, with a reappearance of the LV apical clot and signs of spontaneous coronary artery dissection (SCAD) evident on angiography. Remarkably, by the time of the third angiography, the patient's condition had improved significantly, showing resolution of the previous abnormalities. She was subsequently discharged with a continued regimen of medical therapy, emphasizing the importance of ongoing management in her recovery.

Discussion

Spontaneous coronary artery dissection (SCAD) is a rare yet critical cause of acute myocardial infarction (MI), particularly among young women. It primarily occurs through two mechanisms: the formation of intramural hematomas due to spontaneous rupture of the vasa vasorum or intimal tearing that creates a false lumen. These processes disrupt or impede myocardial blood flow, leading to ischemic injury.3 Unlike other forms of acute coronary syndrome (ACS), SCAD is not linked to atherosclerosis or traditional cardiovascular risk factors, and it is often seen in patients who are otherwise low-risk, such as females under age 50. Studies indicate that SCAD accounts for approximately 0.1% to 4% of ACS cases and up to 35% of MI cases,4,5 with it being the most common cause of pregnancy-associated MI.

Factors believed to contribute to SCAD include multifocal fibromuscular dysplasia, connective

tissue disorders, systemic inflammatory diseases, and hormonal fluctuations, all of which can weaken the arterial wall.2 Additionally, strong emotional and physical triggers such as repetitive Valsalva maneuvers and certain drug use may precipitate SCAD, although many cases lack identifiable triggers. The prognosis for SCAD varies; in this patient's case, severe symptoms improved with timely medical intervention, underscoring the importance of early diagnosis and appropriate management, along with continuous monitoring and follow-up.

Diagnostic strategies for SCAD include coronary angiography, which reveals the extent and location of dissection and informs management decisions. Advanced imaging techniques such as optical coherence tomography (OCT) and intravascular ultrasound (IVUS) provide detailed visualization of arterial wall structures, while echocardiography assesses cardiac function, detects complications such as left ventricular thrombus, and monitors therapy responses.6 Laboratory tests help identify underlying conditions contributing to SCAD by evaluating inflammatory markers, autoimmune profiles, and connective tissue disease indicators.

In this case, the clinical presentation primarily mimicked a myocardial infarction, but the recurrence of symptoms raised concerns for other differential diagnoses, including atherosclerotic coronary artery disease, Takotsubo cardiomyopathy, coronary artery embolism, Prinzmetal's angina, and myocarditis. These were ruled out due to the absence of typical atherosclerotic plaques and stenosis patterns on angiography, lack of characteristic apical ballooning on echocardiography, and the evolving pattern of dissection and healing observed on repeat angiographies. Furthermore, no embolic source was identified, and there was an absence of transient vasospasm and viral prodrome, alongside normal inflammatory markers except for an elevated erythrocyte sedimentation rate (ESR).3,7

It is plausible that this patient's chronic inflammatory state, autoimmune vasculitis, connective tissue abnormalities, and the potential effects of her rheumatoid arthritis treatment contributed to the development of SCAD.

Management of SCAD typically involves a combination of conservative medical therapy, lifestyle modifications, and occasionally percutaneous intervention (PCI). Initial treatment generally includes dual antiplatelet therapy (DAPT), anticoagulants, and guideline-directed medical therapy (GDMT) aimed at reducing myocardial oxygen demand, preventing thrombus formation, and improving heart failure outcomes, especially in cases without flow-limiting dissections.8 According to the American Heart Association's 2021 Revascularization Recommendations, PCI or coronary artery bypass grafting (CABG) may be considered for patients who remain symptomatic despite conservative management, particularly those with persistent ischemia and hemodynamic instability. However, PCI is often associated with poor outcomes due to significant risks, including the potential for iatrogenic dissection, particularly in the context of increased vascular fragility. Additionally, there is a risk of exacerbating intramural hematomas, which can propagate along the length of stents and lead to malapposition. This is particularly concerning given that spontaneous coronary artery dissection (SCAD) frequently occurs in distal coronary segments that may be too small for effective intervention.3,9

For this patient, methotrexate was initiated as a first-line treatment for rheumatoid arthritis, in accordance with American College of Rheumatology guidelines. Folic acid was also prescribed to mitigate the potential toxicity associated with methotrexate.10

Conclusion

This case study emphasizes the significance of recognizing spontaneous coronary artery dissection (SCAD) as a potential cause of acute myocardial infarction, especially in younger women with underlying autoimmune conditions such as rheumatoid arthritis. The complexities surrounding SCAD necessitate a multidisciplinary approach, integrating careful diagnostic strategies and individualized therapeutic interventions. Continuous monitoring and proactive management are crucial in mitigating complications and improving patient outcomes.

Learning points

- Recognition of SCAD: Understanding that spontaneous coronary artery dissection can occur in young, seemingly healthy individuals, particularly women, without traditional cardiovascular risk factors.
- Autoimmune Association: Awareness of the potential link between autoimmune disorders, such as rheumatoid arthritis, and the development of SCAD, highlighting the need for comprehensive patient evaluations.
- Diagnostic Strategies: Emphasizing the importance of advanced imaging techniques, such as coronary angiography, optical coherence tomography (OCT), and intravascular ultrasound (IVUS), for accurate diagnosis and assessment of SCAD.
- Management Approaches: Recognizing the role of conservative management, including dual antiplatelet therapy (DAPT) and anticoagulants, as the first-line treatment, reserving interventional procedures for cases with ongoing ischemia or hemodynamic instability.

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Figure/Video

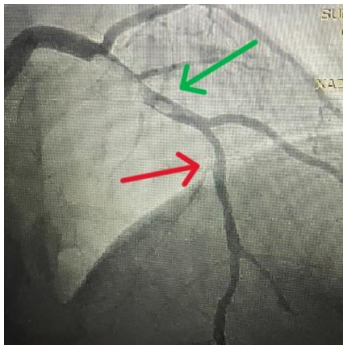

Figure 1: Angiogram (Green arrow showing Grade 4 thrombus in proximal LAD, Red arrow showing healed previous dissection in mid LAD).

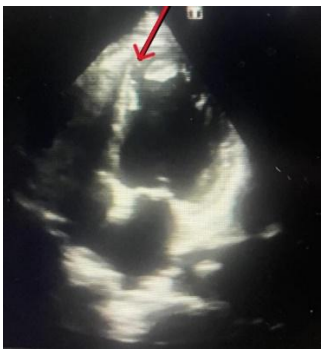

Figure 1: 2D Echo showing Left Ventricular apical clot.

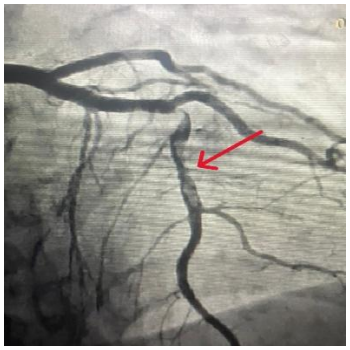

Figure 3: Angiogram Showing SCAD

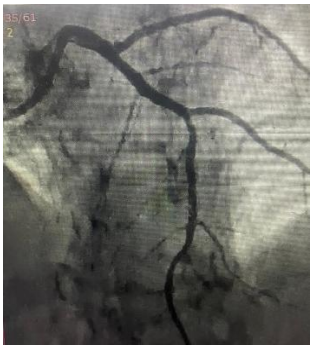

Figure 4: Angiogram after anticoagulation follow up showing no clot or residual disease.