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Case Report

Spontaneous coronary artery dissection and valvular disease revealing an antiphospholipid syndrome in a young woman: Case report ☆

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ABSTRACT

Antiphospholipid syndrome (APS) is characterized by hypercoagulability, manifesting as venous or arterial thrombosis, and pregnancy-related complications, such as fetal loss, preeclampsia, and eclampsia. These conditions occur in the presence of antiphospholipid antibodies (aPL), including anticardiolipin antibodies (aCL) and/or a positive lupus anticoagulant (LA) test.

Antiphospholipid syndrome (APS) can manifest as a primary, isolated condition or be associated with connective tissue diseases, such as systemic lupus erythematosus (SLE). The cardiac system is one of the main target organs affected by APS.

This report describes a rare clinical case involving a young woman with a history of recurrent miscarriages, who presented with cardiac complications, including severe mitral regurgitation and Spontaneous coronary artery dissection. Etiological assessment identified antiphospholipid antibody syndrome as the underlying cause.

The multidisciplinary management plan included initiating therapeutic-dose anticoagulation, followed by referring the patient for potential mitral valve replacement surgery.

Introduction

Antiphospholipid antibodies are autoantibodies that target proteins bound to phospholipids. Antiphospholipid syndrome is a complex, multisystem autoimmune disorder [1], itś a prothrombotic disorder defined by the recurrence of venous thrombotic events, such as deep venous thrombosis and pulmonary embolism, as well as arterial thrombosis. Additionally, APS is associated with recurrent pregnancy loss due to placen-

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tal thrombosis and is marked by the presence of circulating antiphospholipid antibodies [2].

Antiphospholipid syndrome can impinge upon a multitude of organs and tissues, including the cardiovascular system.

Patients with antiphospholipid syndrome often experience substantial cardiovascular involvement. The most common manifestations are coronary artery disease and valvular abnormalities, which together account for more than two-thirds of cases [3].

In this clinical case, the presentation of cardiac manifestations prompted an etiological investigation, leading to the diagnosis of antiphospholipid syndrome.

Case report

This case involves a 24-year-old female patient with a medical history of 4 unexplained fetal demises of morphologically normal fetuses at a gestational age of at least 10 weeks, confirmed by normal ultrasound findings. She initially presented for evaluation and management of New York Heart Association (NYHA) class II dyspnea, which she had been experiencing for the past 4 months.

The patient was eupneic at rest with a heart rate of 60 beats per minute, a systolic blood pressure of 140 mmHg, an oral temperature of 36.5°C, and a saturation level of 98%.

Cardiovascular examination revealed a 5/6 holosystolic murmur, most pronounced at the mitral area, without clinical signs of heart failure. Pleuro-pulmonary examination findings were normal, with symmetric auscultation and no audible rales. Both neurological and dermatological examinations yielded normal results.

The electrocardiogram revealed sinus rhythm with a heart rate of 60 beats per minute, and a left axis deviation. The PR interval was normal at 160 ms, with no evidence of ventricular hypertrophy or repolarization disorder (Fig. 1).

Initial blood tests revealed a hemoglobin level of 11.4 g/dL and a platelet count of 112 × 103/μL. Serum liver enzymes and renal function were within normal limits, with CRP at 0.6 g/dL

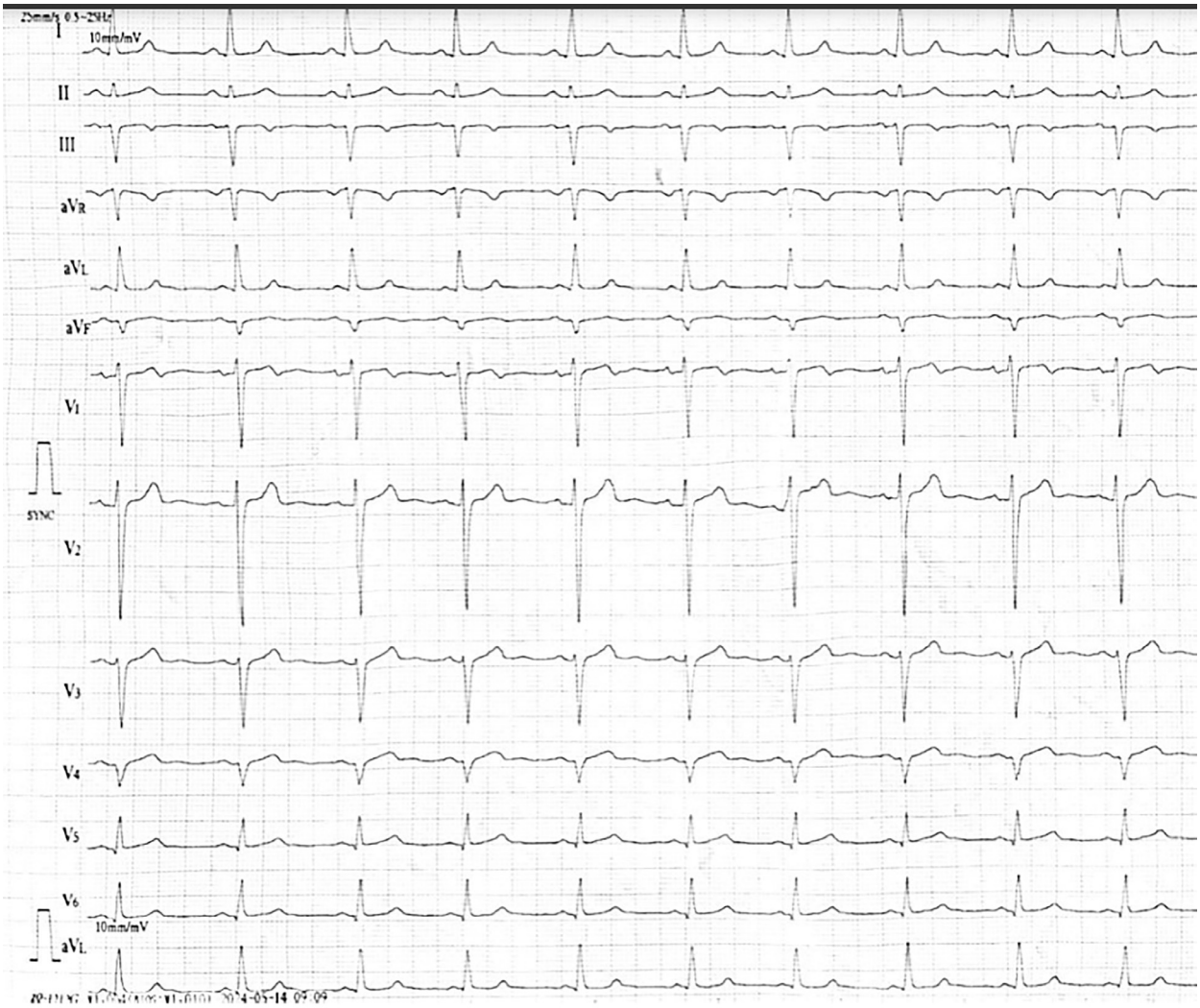

Fig. 1 – Electrocardiogram of the patient.

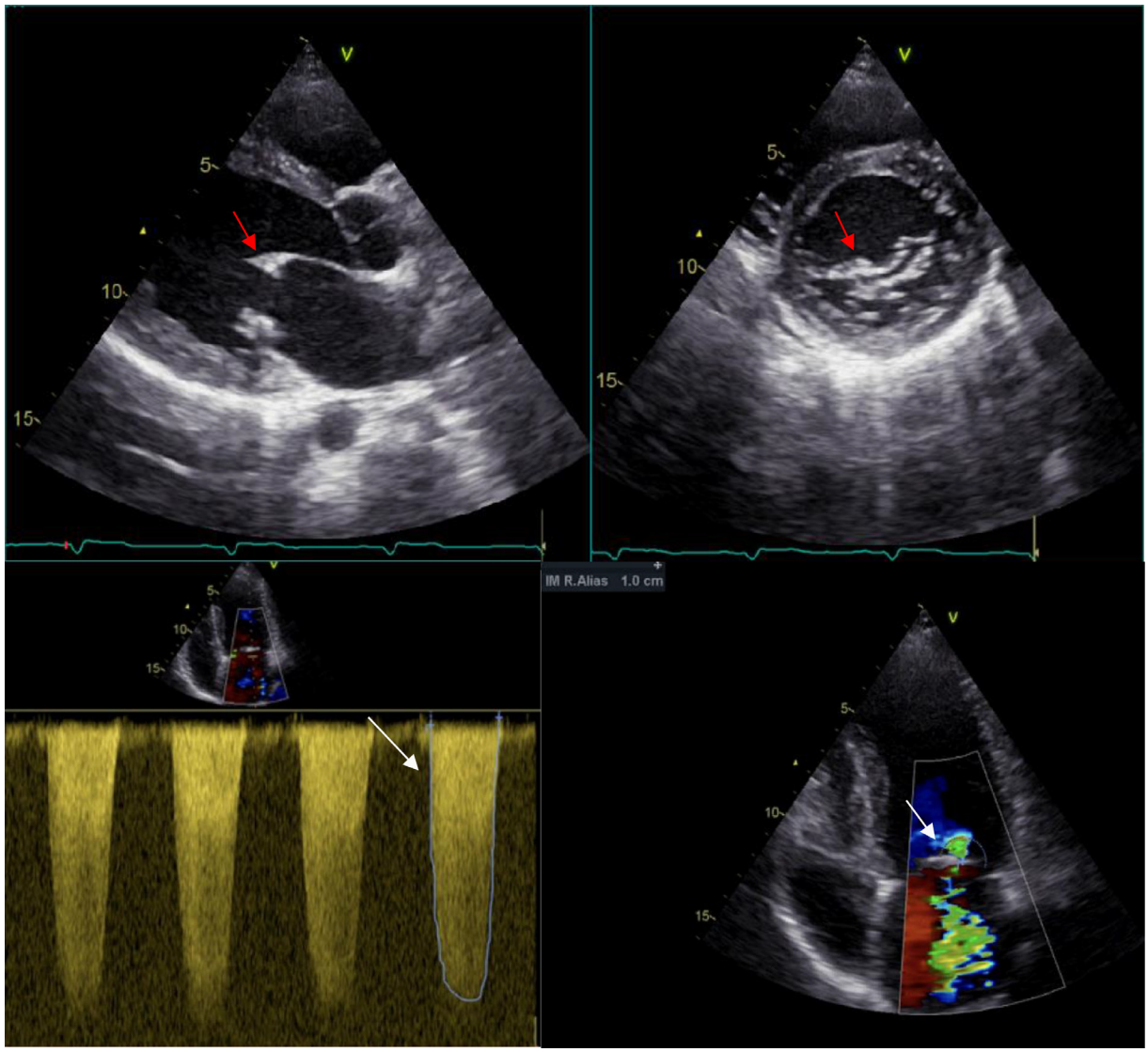

Fig. 2 – Echocardiogram images showing thickened leaflets (red arrows) and severe mitral regurgitation (white arrows).

and a PT-INR of 1.15. ProBNP levels were elevated, peaking at 2251 mg/L, beta HCG level was negative.

Given the symptomatology presented by the patient, an echocardiogram was performed, revealing a dilated left ventricle, with segmental wall motion abnormalities as follows: akinesia of the mid-segment of the anteroseptal wall, akinesia of the apex extending to adjacent wall segments. The left ventricular ejection fraction (LVEF) was 45% by Simpson's biplane method (SBP). The mitral valve exhibited thickened leaflets and a severe mitral regurgitation (MR) (PISA: 1.0 cm, EROA: 0.43 cm^2, RV: 76 mL). The right ventricle was nondilated with good function (Fig. 2).

The patient also underwent a cardiac MRI, which revealed necrosis sequelae in the left anterior descending (LAD) artery territory affecting the apical segments of the left ventricle, which were identified as nonviable (Fig. 4).

Consequently, coronary angiography was conducted, revealing a smooth coronary network with findings suggestive of dissection at the distal segment of the left anterior descending (LAD) artery, with a TIMI-III flow (Fig. 3).

Given the context of recurrent miscarriages and cardiac involvement, an immunological workup was requested, revealing positive anti-beta-2 glycoprotein antibodies (IgM and IgG > 100 GPL, and positive anti-cardiolipin antibodies (IgG > 120 GPL, IgM > 80 GPL). Upon retesting after 3 months, anti-cardiolipin antibodies were always high.

Given this set of clinical and biological criteria, the diagnosis of antiphospholipid antibody syndrome has been made with subsequent cardiac involvement.

Therapeutically, the patient was placed on curative anticoagulation, ACE inhibitors and beta blockers and she was referred to the cardiothoracic surgery department for a potential mitral valve replacement.

In the next follow-up for our patient, we plan to adjust the treatments for heart failure and introduce SGLT2 inhibitors to improve her left ventricular ejection fraction.

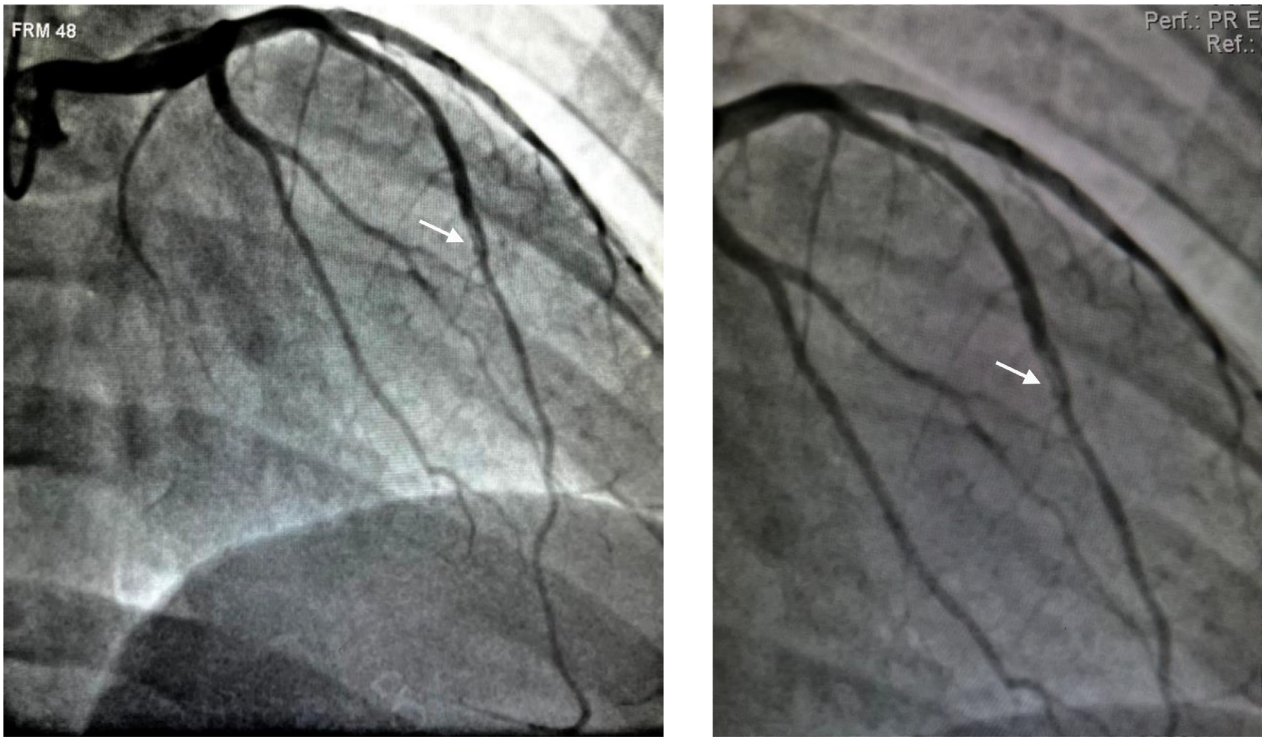

Fig. 3 – Coronary angiogram image suggestive of a dissection at the distal LAD with a TIMI-III flow (white arrow).

Discussion

The revised Sapporo Classification Criteria for antiphospholipid syndrome establish that the diagnosis is substantiated when a patient satisfies a minimum of 1 clinical criterion in conjunction with at least 1 laboratory criterion [4]. This was the case for our patient. Antiphospholipid antibody syndrome is characterized by significant clinical heterogeneity, presenting in a wide array of manifestations.

In antiphospholipid syndrome, cardiac manifestations include valvular disease, coronary artery disease, intracardiac thrombus formation, pulmonary hypertension, and dilated cardiomyopathy [5,6].

Cardiac valvular pathology in antiphospholipid syndrome is characterized by irregular thickening of the valve leaflets, resulting from the deposition of immune complexes. This pathological process can lead to the formation of vegetations and subsequent valve dysfunction. Such lesions are common and may constitute a significant risk factor for stroke [5].

The mitral valve is the most frequently affected, followed by the aortic and tricuspid valves. This predilection is attributed to the increased vulnerability of the left-sided valves to micro injuries, which result from stress, jet effect, and turbulence [6,7].

Studies report a wide range in the prevalence of heart valve disease in primary antiphospholipid syndrome patients, spanning from 10% to more than 60% [7].

While the majority of patients develop only mild valvular regurgitation, about 4-6% progress to severe regurgitation, ultimately necessitating surgical valve replacement [3,8]. Our patient is part of this latter category.

Furthermore, our patient presented a spontaneous coronary artery dissection (SCAD). It is a nonatherosclerotic cause of coronary artery dissection. It manifests either as a classical dissection with a visible false lumen or as an intramural hematoma, which may be accompanied by an intimal tear [8]. Myocardial infarction develops as a consequence of coronary obstruction due to luminal compression, which can arise from either a dissection or the propagation of an intramural hematoma.

Although SCAD can affect any coronary artery, the left anterior descending (LAD) artery and its diagonal branches are most commonly involved [9,10]. While most SCAD lesions can be identified through angiography alone, differentiating potential SCAD from other causes of coronary artery stenosis remains challenging.

The precise mechanism of SCAD remains unclear. A definitive connection has not been established. Registry data show a low incidence of systemic inflammatory disorders among SCAD patients (less than 5% in most cohorts), though these conditions may be underdiagnosed [11].

In terms of the therapeutic strategy for mitral insufficiency, given the relatively young age of patients and the frequent requirement for long-term anticoagulation in antiphospholipid syndrome, mechanical valves often emerge as the preferred option for replacement. However, the risk of thromboembolic complications associated with mechanical valves poses a significant threat to their functionality [12]. In the other hand the primary advantage of a bioprosthetic valve

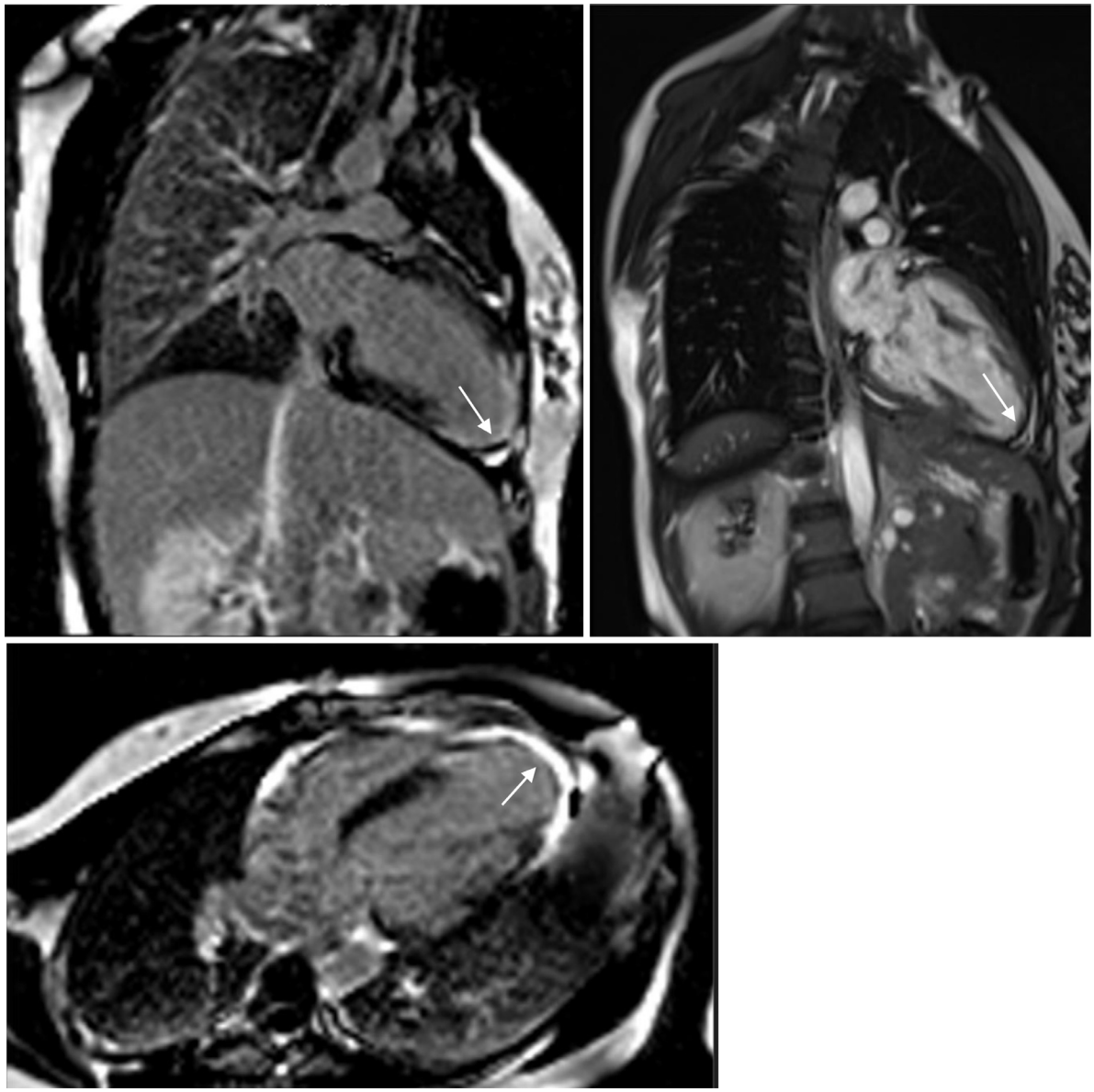

Fig. 4 – MRI images showing the presence of necrosis sequelae in the left anterior descending artery territory affecting the apical segments of the left ventricle (white arrows).

is its independence from oral anticoagulation. Nonetheless, valve failure due to excessive pannus formation and subsequent stenosis often necessitates replacement after a certain period [13]. In our case, we opted for a potential valve replacement with a mechanical valve, accompanied by effective anticoagulation.

In the other hand SCAD lesions managed medically often show angiographic improvement, evidenced by restored blood flow and decreased stenosis severity. Consequently, over 80% of patients can achieve favorable outcomes with conservative treatment [11]. Although SCAD can occur in other coronary arteries besides the initial site of dissection, revascularization has not been shown to prevent recurrent SCAD or myocardial infarction [11] in our case, we opted for a conservative strategy.

Given that the patient experienced sequelae of myocardial infarction and heart failure, treatment with ACE inhibitors and beta blockers was initiated in accordance with clinical guidelines. Beta blockers may provide additional benefits in mitigating the recurrence of SCAD, demonstrating up to a 64% reduction in the incidence of recurrent SCAD over a median follow-up period of 3.1 years [8].

Patient consent

Written informed consent for the publication of this case report was obtained from the patient.

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