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## MULTIDISCIPLINARY MANAGEMENT OF CORONARY ARTERY DISEASE COMPLICATING LUPUS NEPHROPATHY: A CASE STUDY

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### ABSTRACT:

**Background:** This case report presents a 30-year-old female patient with a history of systemic lupus erythematosus (SLE) leading to lupus nephropathy, currently managed with peritoneal dialysis and immunosuppressive therapy.

**Case Description:** The patient experienced worsening dyspnea and angina, prompting medical attention. Echocardiography revealed significant mitral and tricuspid regurgitation, with subsequent development of resting angina associated with autonomic nervous system dysfunction. Rheumatological assessment initially discounted active lupus. Cardiac catheterization confirmed intravascular coronary artery disease, including coronary dissection of intermediate and circumflex branches with associated coronary ectasia.

**Treatment and Outcome:** Cardiothoracic surgery considered tricuspid plasty, mitral valve replacement, and coronary artery bypass grafting following dissection of multiple coronary branches. Spontaneous coronary dissection, more common in women, manifested as unstable angina, with a rare incidence of 0.43% associated with SLE: extra-renal vasculitis, a rare complication of lupus nephropathy, further complicated management.

**Conclusion:** Individualized treatment strategies are essential, considering the extent of vessel involvement and hemodynamic status in patients with SLE-related coronary artery disease. Early recognition and multidisciplinary management are critical in optimizing outcomes for such complex cases.

**Keywords:** heart dissection, Ectasia Coronaria, Nephropathy due to lupus and Vasculitis.

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**INTRODUCTION:**

With a reported incidence of 0.1% to 1.1%, spontaneous coronary artery dissection (SCAD) is an uncommon diagnostic of exclusion in acute coronary syndromes. Most studies involve patients under 52 years old and women who are peripartum. It is not to be confused with atherosclerotic aetiology. It is described as a spontaneous separation of the coronary artery wall that is neither iatrogenic nor the result of trauma. There are now two accepted theories about pathogenesis: haemorrhaging of the medial layer due to injury to the vasa vasorum and rupture of the intima with subsequent formation of an intramural hematoma. Clinical onsets range from heart failure to sudden death, including all variations of acute coronary syndrome. Both a stressor that triggers the condition and an underlying vascular disease usually predispose people to it (Li et al., 2024).

In the first instance, the most common association, fibromuscular dysplasia, is discussed about idiopathic or hormonal treatment, connective tissue illnesses, systemic inflammatory diseases, and pregnancy. Predisposing stress factors include but are not limited to, extreme stress and physical activity, childbirth, severe Valsalva activities, and certain addictive substances. Less than 1% of instances in case reports have been linked to autoimmune inflammatory illnesses and SCAD, though it's likely that this number is underestimated. Chronic inflammation brought on by Vasculitis has been suggested as a risk factor. An early and precise diagnosis is essential since there are differences between SCAD with an atherosclerotic and non-atherosclerotic aetiology regarding presentation, development, and prognosis. These distinctions mainly relate to the treatment the latter requires (Chaaban & Kshatriya, 2022).

Although coronary angiography was the first-line procedure for diagnosing SCAD, it can now be performed during invasive angiography image acquisition due to restrictions in evaluating frames per second and the lack of improved structural clarity of the vessel in question. Optical coherence tomography (OCT) and intracoronary arteries employ ultrasound (IVUS, intravascular ultrasonography) to finish diagnosing and treating SCAD. Many case series have been used to propose therapeutic approaches.

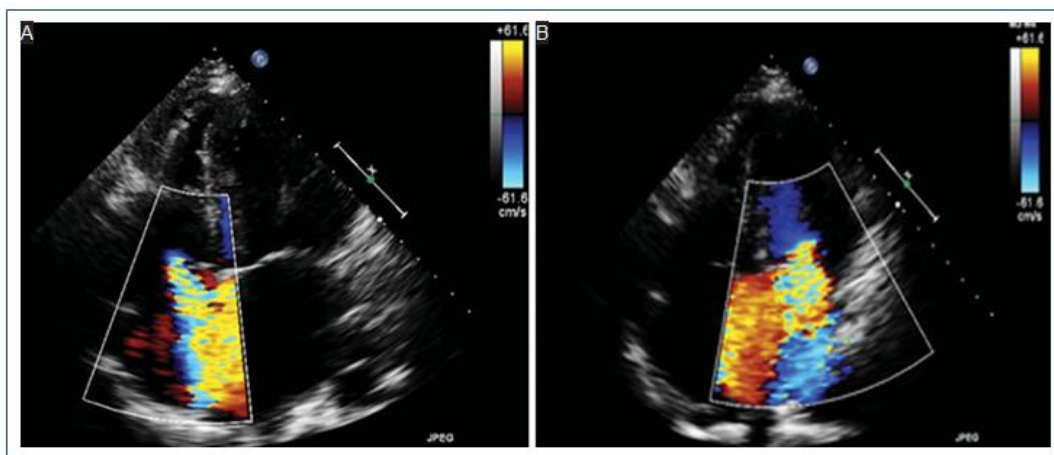
Reference	Summary
Li et al., 2024	Describes spontaneous coronary artery dissection (SCAD) as an uncommon diagnostic of exclusion in acute coronary syndromes, with a reported incidence of 0.1% to 1.1%. The study discusses pathogenesis theories, clinical manifestations, and predisposing factors, emphasizing the need for early and precise diagnosis.
Chaaban & Kshatriya, 2022	Explores the pathogenesis of SCAD, including theories involving injury to the vasa vasorum and rupture of the intima. Highlights associations with fibromuscular dysplasia, hormonal treatment, connective tissue illnesses, and pregnancy. The study also underscores the importance of distinguishing between atherosclerotic and non-atherosclerotic etiologies for appropriate treatment planning.
García-Sánchez et al., 2021	Reports a case of a young woman with systemic lupus erythematosus presenting with unstable angina and decompensated heart failure. Angiographic examination revealed multiple coronary artery dissections, prompting suspicion of lupus activity as a contributing factor.

Based on clinical and imaging findings, risk stratification should ideally facilitate customized treatment, ranging from conservative measures to coronary artery bypass graft surgery. We describe the case of a young woman with systemic lupus erythematosus who initially presented with unstable angina and decompensated heart failure. During her hospital stay, an angiographic examination revealed multiple coronary artery dissections, which led to the initial suspicion that the lupus activity was the cause of the decompensation (García-Sánchez et al., 2021).

**CASE PRESENTATION:** A 30-year-old female patient with a history of acute myopericarditis in 2020, lupus nephropathy on renal replacement treatment with peritoneal dialysis, and systemic lupus erythematosus on prednisone. Two and a half weeks before admission, the patient’s functional class deteriorated due to dyspnea and angina, changing from big to minor strains. The initial echocardiography showed maintained proper ventricular systolic function, significant mitral and tricuspid regurgitation, binaural dilation without thrombus, grade II diastolic dysfunction, a left ventricular ejection fraction of 36%, and widespread hypokinesia (Fig. 1). The rheumatology

service ruled out lupus at the time of admission (García-Sánchez et al., 2021; Melano-Carranza et al., 2023).

Her dyspnea worsened after a subsequent episode of angina at rest linked to adrenergic discharge that occurred while she was in the hospital. An ECG was taken during the incident, revealing sinus rhythm, a typical PR and QT interval, septal and left ventricular hypertrophy data, and symmetric positive sharp T waves in the precordial leads. The hemodynamic interventional cardiology service evaluated specific biomarkers that revealed a troponin I of 0.24 ng/ml, raising the possibility of an underlying ischemic disorder (Fig. 2). This evaluation supported the need for cardiac catheterization in patients with intravascular coronary disease, including those with Markis I coronary ectasia, coronary dissection of the intermediate branch, and circumflex branch distal to the first obtuse marginal (Fig. 3). After dissection of the intermediate, obtuse marginal, and posterolateral branches, the cardiothoracic surgery service was thought to be capable of performing tricuspid plasty, mitral valve replacement, and coronary artery bypass operations (Haddad et al., 2024).



*Figure 1. A: Apical four-chamber echocardiogram with a 10.2 mm vena contracta is present, along with significant tricuspid regurgitation. The right atrium has a surface size of 21 cm<sup>2</sup>. B: An apical four-chamber echocardiogram demonstrating severe mitral regurgitation with a 0.4 cm<sup>2</sup> regurgitant orifice area. There is 42 ml/m<sup>2</sup> of regurgitation in the left atrium and a volume of 125 ml.*

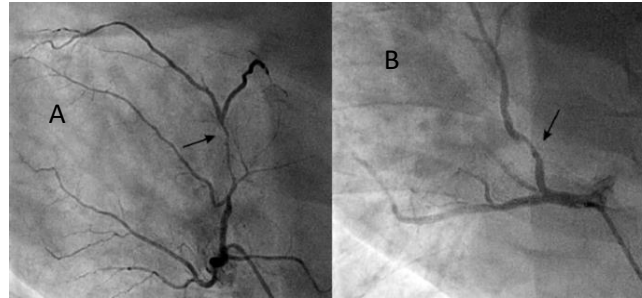


*Figure 2 depicts a 12-lead ECG that exhibits signs of subendocardial ischemia, including sinus rhythm, a 10° QRS axis, 140 ms P wave, 200 ms PR, 100 ms QRS, elevated precordial voltage, 25 mm Cornell, and 10 mm symmetrical acute T waves.*

#### **CASE DISCUSSION:**

There are currently no precise clinical guidelines for SCAD, even though the condition was first reported in 1931 during the autopsy of a 42-year-old woman. Since then, there have been many case reports and brief studies. In SCAD, a fake lumen is created by severing two of the artery wall's three layers, typically between the media and intima. Pregnancy accounts for 31 per cent of occurrences, and women are more likely to have it (69%). It might manifest as angina (95.6%) or even abrupt death (<0.5%). With an incidence of 0.43% and a risk three times higher than the general population, its relationship with systemic lupus erythematosus is highly uncommon. It is an unusual sign of extrarenal Vasculitis from the perspective of lupus nephropathy (Nor et al., 2023).

The aetiology of SCAD was previously thought to be postpartum or peripartum, atherosclerotic, and idiopathic; however, with the advent of new imaging techniques, associations with fibrodysplasia, the use of oral contraceptives or cocaine, Ehlers-Danlos and Marfan syndromes, and extreme exercise have been reported. To comprehend the cause, Saw et al.<sup>3</sup> suggest four categories: 1) linked to connective tissue disorders, 2) individuals suffering from Vasculitis or atherosclerosis, 3) postpartum, and 4) idiopathic (Khan & Elkammash, 2022).



*Figure 3. A: Coronary angiography, caudal left anterior oblique view; type B dissection of the circumflex artery's distal segment (thin arrow); NHLBI type B or Saw type 1 dissection of the middle branch's proximal segment (big arrow). B: Coronary angiography, cranial left anterior oblique view; proximal segment of the intermediate branch dissected according to NHLBI type B*

Intracoronary imaging techniques, like IVUS and OCT, have been proposed in the diagnosis and treatment algorithm due to limitations in lesion characterization and subsequent risk stratification. Nevertheless, invasive coronary angiography remains the first-line method for the initial diagnosis. Even though these studies are ideal, not all centres can access these invasive imaging techniques. Therefore, they are not necessary in clinical practice. Thus, the emphasis is on the earliest detection using coronary angiography. The National Heart, Lung, and Blood Institute (NHLBI) previously classified coronary artery dissections into six categories (A). Due to the patient's twofold lumen, divided by a radiolucent region and persisting following contrast washout, we classified their case as type B (Patel et al., 2023).

However, Saw et al. propose three categories in angiographic classification in their review due to the overlap of requirements with other conditions when dealing with types C to F. These are based on various characterizations employing intracoronary imaging. The first type of lesion is pathognomonic, found in 28% of cases. The second type of lesion is called a diffuse lesion, which is usually larger than 30 mm and has no visible dissection plane. It can cause complete occlusion of the vessel and is further divided into two subtypes: type B (lesions extending to the entire boat), which is the most common type (67.5%); the third type of lesion is called a focal or tubular lesion, which indicates stenosis due to atherosclerosis and usually requires intracoronary imaging studies to confirm the diagnosis. These lesions account for 3.4% of cases (MAN, 2021; Zafrullah et al., 2024).

The anterior descending artery is the most afflicted vessel in predominance (33–43%); it affects the mid-distal segments and, in less than 10% of instances, the proximal segments. In 9–18% of cases, multiple vascular dissection takes place. These latter percentages, however, are from assessments that cover all SCAD aetiologies. Numerous management algorithms exist. In this particular context, Saw et al., the leading authorities, discuss a step-by-step diagnosis that begins with coronary angiography. Based on the previously mentioned clinical presentation, it is advisable to search for the presence of type 1 SCAD objectively. If this is absent, we shall attempt to locate the fine, diffuse stenosis as type 2 SCAD describes. At this point, undergoing IVUS or OCT with intravenous nitroglycerin injection is advised to determine whether a re-evaluation in 4-6 weeks is feasible (Kim et al., 2021; Suresh et al., 2020).

If these features are unavailable, we will search for the type 3 appearance, mimicking atherosclerotic lesions; intracoronary imaging examinations will be necessary in the latter case. Regarding imaging modalities, OCT has the best resolution for identifying false lumens, intramural thrombi, and intimal rupture. However, because of its reduced penetration, it can be more challenging to determine the size of some intramural hematomas. Even with the clear benefits, there are dangers connected with coronary intervention that must be considered. These hazards include the possibility of thrombosis from the catheter's entry and the mechanical or hydraulic expansion of the dissection using the catheter and contrast medium. Because angiotomography cannot detect lesions in distal vessels, which are the most prevalent area of involvement, it is not used in decision-making (Ete et al., 2020).

Regarding the patient in the clinical instance, our understanding of the lesions may lead us to the typical clinical presentation of type 1, whereby the circumflex artery and the intermediate branch both exhibit double lumens. In addition, the patient presented with decompensated heart failure and unstable angina, two conditions that increase the risk of cardiovascular problems. Markis I coronary ectasia is another critical factor that significantly alters the possibility of percutaneous interventions and serves as a reminder of the many factors that need to be considered for both pharmacological treatment and hemodynamic flow applications in the revascularization of an ectatic vessel. Since no randomized clinical trials compare conservative management to revascularization strategies, the best course of treatment has not yet been determined (GÜZEL & AKTAN, 2022; Ogedegbe et al.).

Instead, the evidence comes from case series, where atherosclerosis is identified as the primary cause of SCAD. For this reason, individualizing care is necessary in our patient's case because, while an atherogenic background is not ruled out, the patient's clinical presentation points to different pathophysiological pathways. Because of the potential for increased hematoma and dissection, thrombolysis is recognized to carry a high risk of complications. In stable patients with minimal risk of complications and development, conservative therapy has been suggested based on reviews citing spontaneous remission in 4-6 weeks in non-complex cases. While surgical revascularization is still the primary choice for invasive treatment in up to 54% of instances, percutaneous coronary intervention is still the preferred method for most problems (Alvarez et al., 2023).

Similarly, more damaged vessels indicate a preference for surgical therapy. Medical intervention may be considered if sufficient flow is seen on coronary angiography following the acute situation. Saw et al. suggest a dichotomized therapy approach based on baseline risk (high risk: stable coronary artery disease, peripheral arterial disease, atrial fibrillation, acute or prior myocardial infarction, and cerebral vascular events) in response to this disparity. Without risk factors, medicinal therapy (such as renin-angiotensin axis blockers, antiplatelet medicines, and beta-blockers) is used for conservative management. If one or more of the following criteria are met, the case is deemed high risk: dissection of the coronary trunk, persistent angina or ischemia, ventricular fibrillation or tachycardia, cardiogenic shock; percutaneous coronary intervention is the treatment of choice for isolated trunk lesions; revascularization surgery is the treatment of choice when proximal lesions are added to other vessels (Agwuegbo et al., 2024; D'Amato et al., 2024). The latter is also taken into account in situations where percutaneous coronary intervention is not feasible due to hemodynamically stable ventricular fibrillation tachycardia or continuous angina or ischemia detected in more than two arterial areas. However, hemodynamic instability can happen in both cases, such as cardiogenic shock and ventricular arrhythmias. On the other hand, ventricular mechanical support or inotropes, among other measures, must improve the hemodynamic condition if hemodynamic instability arises, whether due to ventricular arrhythmias or cardiogenic shock. Given the patient's comorbidity and context, it would be appropriate to classify her as high-risk. She has a known decreased ejection fraction and did not experience ventricular arrhythmias or cardiogenic shock while in the hospital. The circumflex artery and the intermediate branch's proximal dissection can be determined using coronary angiography.



However, stent insertion through percutaneous coronary intervention can be challenging because of Markis I coronary ectasia. Due to these factors, the process made sense based on the revascularization procedure algorithm (Miranda et al., 2022).

**CONCLUSION:** This young lady had several ongoing issues related to an autoimmune condition. Acute coronary syndrome was suspected at the time of angina and decompensation of chronic heart failure based on the previously described signs, as well as on ECG and biomarker data. Coronary angiography was used to confirm the diagnosis. This is an example of retrospective analysis, where the chest pain follow-up protocol showed the angiographic finding and modest changes were interpreted using an electrocardiogram, a reproducible diagnostic tool that all cardiologists use. The identification of acute coronary syndrome and its failure to advance to infarction, as such, allowed for better adaptation by compensatory mechanisms.

Interestingly, acute changes in ischemia indicated in the EKG are linked with better prognosis in the postoperative phase. As previously indicated, the recommended course of care considered the patient's chronic comorbidities, the amount of vessels involved, and the discovered valvular pathology. It also took into account the high risk of post-surgical complications because immunosuppression was necessary to prevent the patient's systemic lupus erythematosus from getting worse. After 36 hours in the postoperative phase, extubation was accomplished. According to her most recent examination, the patient is on anticoagulant medication, without angina, and is in functional class II of the New York Heart Association. The multidisciplinary team follows up with her regularly.

Surgical surgery was thought to be the next course of action despite the significant risks of surgical complications, the lack of immediate improvement with conservative treatment, and the likely danger of the development of cardiogenic shock. In revascularization surgery, age and organic compensatory mechanisms most likely provided a good prognosis. When a young woman has acute coronary syndrome or heart failure, she should always be evaluated for SCAD. Clinical judgement should always direct therapeutic management, even in the absence of solid data, and this is especially true when treatment is individualized.

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