

Myocardial rupture and systemic lupus erythematosus: a case report

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Background

Systemic erythematosus lupus (SLE) is an autoimmune disease associated with significant cardiovascular morbidity and mortality, even in young patients.

Case summary

A young female with recently diagnosed SLE under corticotherapy developed pulmonary oedema and respiratory failure. The diagnostic workup revealed mildly elevated cardiac troponin, significantly elevated NT-proB-type natriuretic peptide (NT-proBNP) and mild pericardial effusion without other echocardiographic abnormalities. Systemic erythematosus lupus-associated myocarditis was presumed, and her clinical status improved after corticotherapy intensification. However, transthoracic echocardiogram repeated days later revealed a large pericardial effusion with findings suggestive of a contained myocardial rupture originating in the inferolateral basal left ventricular (LV) segment, which was confirmed by computed tomography scan. Cardiac catheterization exhibited normal coronary arteries. The patient was submitted to cardiac surgery and the LV pseudoaneurysm was successfully repaired.

Discussion

Myocardial rupture with LV pseudoaneurysm formation usually occurs in the setting of acute myocardial infarction, but also in other rare contexts. Cardiac rupture is associated with an extremely high mortality unless early diagnosis and urgent surgical intervention are provided.

Keywords

Heart rupture • Lupus erythematosus • Systemic • Myocardium • Case report

Learning points

- Systemic lupus erythematosus is an autoimmune disease associated with significant cardiovascular morbidity and mortality, even in young patients.
- Multimodality imaging is valuable for the correct diagnosis and characterization of left ventricular pseudoaneurysm.
- A left ventricular pseudoaneurysm carries a high risk of expansion and rupture, and thus requires urgent surgical repair.

Introduction

Cardiovascular (CV) involvement in systemic erythematosus lupus (SLE) is frequent; pericardial involvement is the most common but

coronary artery disease, myocarditis, and endocarditis are other major conditions associated with SLE.¹

Myocardial rupture with left ventricular (LV) pseudoaneurysm formation usually occurs in the setting of acute myocardial infarction

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(AMI), but can also be seen in the context of cardiac trauma, cardiac infection, cardiac tumours, or aortic dissection. It can also be iatrogenic, and more recently it has been reported in stress cardiomyopathy.

To our knowledge, there are no reports of SLE-associated myocardial rupture, except for cases of overt AMI with associated coronary artery disease and an identifiable culprit artery.²

Timeline

Day 0 (initial presentation)	Constitutional syndrome for 3 weeks. Initial investigation: bicytopenia, elevated inflammatory markers, proteinuria, inflammatory myopathy, and polyserositis [bilateral pleural effusion; transthoracic echocardiogram (TTE) with mild circumferential pericardial effusion (PE) and no other abnormalities]
Day 3	Mild hypoxemic respiratory insufficiency
Day 14	Systemic lupus erythematosus (SLE) diagnosis was presumed and corticotherapy (CCT) was initiated
Day 16	Progressive respiratory failure due to pulmonary oedema, mechanical ventilation for 3 days Cardiology evaluation: mild increase in hs-TroponinI and a significant rise of NT-proBNP; electrocardiogram with a slight superior concave elevation in leads V5-6, DI, DII, and aVL; TTE with similar findings; assumed as a secondary myocardial injury/LES-associated myocarditis
Day 19	Corticotherapy was intensified and the clinical status improved
Day 34	New progressive respiratory insufficiency and pulmonary congestion
Day 35	Transthoracic echocardiogram: medium-large hyperechogenic PE, presence of a 30 × 30 mm sac with a narrow neck adjacent to the inferolateral wall with systolic contrast filling, suggesting the diagnosis of a pseudoaneurysm; no clinical or echocardiographic signs of cardiac tamponade Cardiac computed tomography confirmed the presence of a pseudoaneurysm and a loculated large volume haemopericardium Cardiac catheterization showed normal coronary arteries
Day 36	Transferred to the referral surgical centre
Day 40	Submitted to cardiac surgery—left ventricular pseudoaneurysm was successfully repaired with a pericardium patch
Day 55	Discharged
3 months after discharge	Cardiac magnetic resonance imaging: preserved biventricular systolic function with segmental wall motion abnormalities and late contrast

Continued

enhancement pattern compatible with previous inferolateral myocardial infarction; thickening of the pericardium adjacent to those segments with diffuse contrast uptake

Case presentation

A 30-year-old female with no relevant past medical history was admitted to the emergency department due to a constitutional syndrome over the last 3 weeks. She complained of persistent fever, myalgias, weight loss, marked asthenia, and anorexia. Initial investigation revealed painful joints during mobilization (wrists, elbows, and knees bilaterally), but without inflammatory signs, bicytopenia (anaemia—10.6 g/dL—and thrombocytopenia—140,000/ μ L), elevated inflammatory markers (PCR 15 mg/L, normal range <7 mg/L; ESR 57 mm/h, normal range <20 mm/h), proteinuria, inflammatory myopathy, and polyserositis (bilateral pleural effusion, mild circumferential pericardial effusion). Besides mild pericardial effusion, her initial transthoracic echocardiogram (TTE) was normal. After an exhaustive workup, she was diagnosed with SLE and treated with corticotherapy (CCT). She developed progressive respiratory insufficiency due to pulmonary oedema.

At that point, she was evaluated by the cardiology team. There was a mild increase in troponin I (1.6 ng/mL maximum; normal range <0.05 ng/mL) and a significant rise of NT-proBNP (3970 pg/mL; normal range <150 pg/mL). Electrocardiogram presented sinus rhythm and a slight superior concave elevation in leads V5-6, DI, DII, and aVL. Transthoracic echocardiogram was similar to the initial one, with a small pericardial effusion, normal left ventricular (LV) function, and no wall motion abnormalities (WMA). She had no chest pain. The diagnosis of SLE-associated myocarditis or secondary myocardial injury was assumed. The patient was then admitted to the intensive care unit due to respiratory failure and needed mechanical ventilation for 3 days. After CCT intensification, her clinical status improved.

However, 15 days later she developed new progressive hypoxic respiratory insufficiency and pulmonary congestion. Patient-related mild dyspnoea at rest with no other complaints. At physical examination, she was slightly tachypnoeic with FiO₂ 0.32, normotensive and normocardic, with no fever. She had bilateral rales at pulmonary auscultation and no other relevant findings. Transthoracic echocardiogram was repeated and showed a medium-large translucent pericardial effusion, predominantly adjacent to the posterior and lateral LV walls. The basal segments of these walls were thinned and there was evidence of bidirectional flow into pericardial space using Doppler colour flow. The injection of contrast material during TTE further unveiled the presence of a 30 × 30 mm sac with a narrow neck adjacent to the inferolateral wall, with systolic contrast filling, suggesting the diagnosis of a pseudoaneurysm (*Figure 1*). There were no clinical or echocardiographic signs of cardiac tamponade.

Further evaluation with cardiac computed tomography scan (*Figure 2*) confirmed the presence of a massive pseudoaneurysm originating in the inferolateral basal segment of the LV (42 × 34 × 32 mm) and a loculated large volume haemopericardium adjacent to the

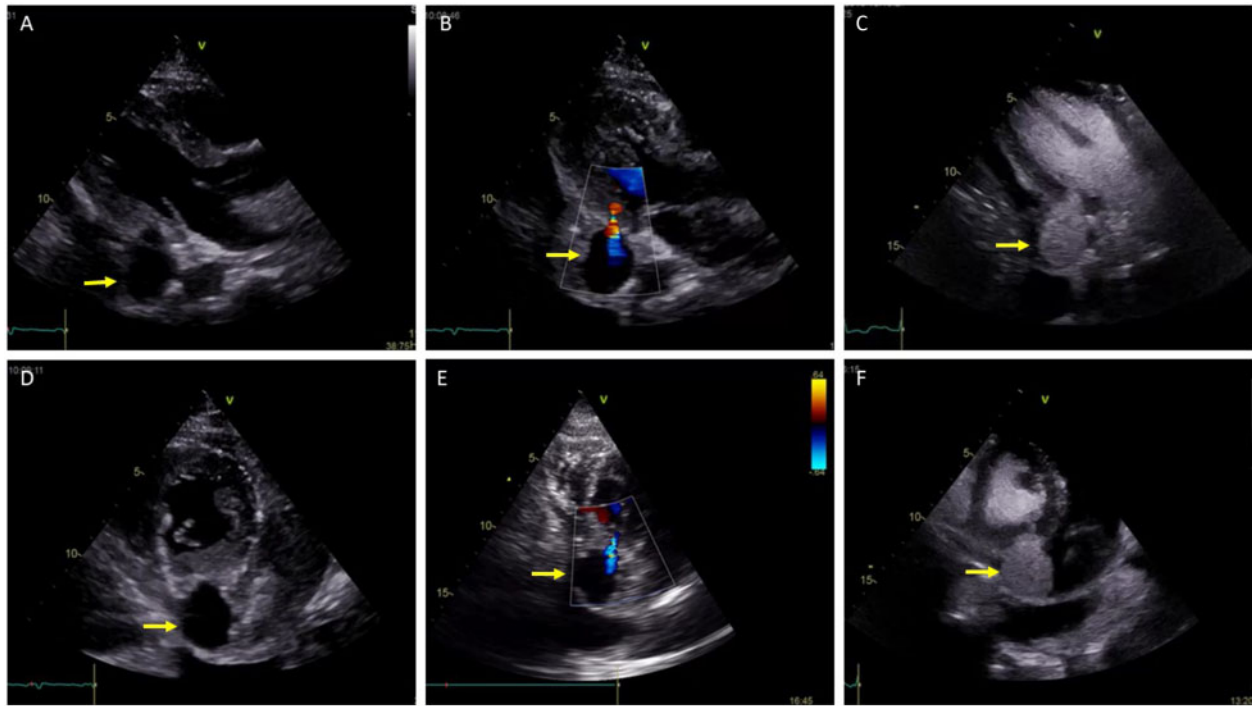


Figure 1 Pseudoaneurysm (30×30 mm; arrow) at the basal segment of the inferolateral left ventricular wall on transthoracic echocardiogram (A–C: parasternal long-axis views, D–F: modified parasternal short-axis views), with passage of contrast through a narrow neck (C, F).

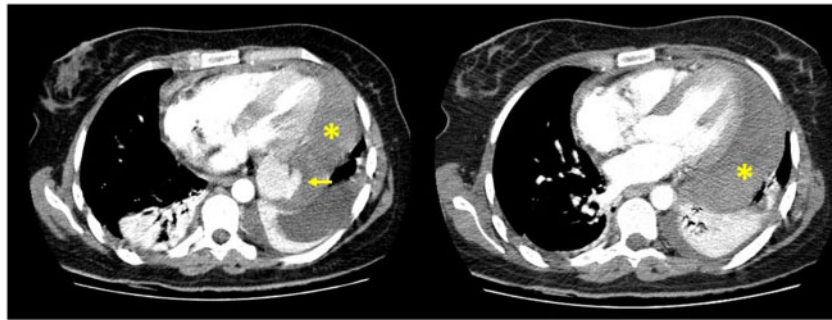


Figure 2 Pseudoaneurysm originating in the inferolateral basal segment of the left ventricular ($42 \times 34 \times 32$ mm; arrow) and loculated large volume haemopericardium adjacent to the left ventricular lateral wall (asterisk), on chest computed tomography scan.

lateral wall of the LV (maximum thickness: 36 mm). Cardiac catheterization exhibited angiographically normal coronary arteries.

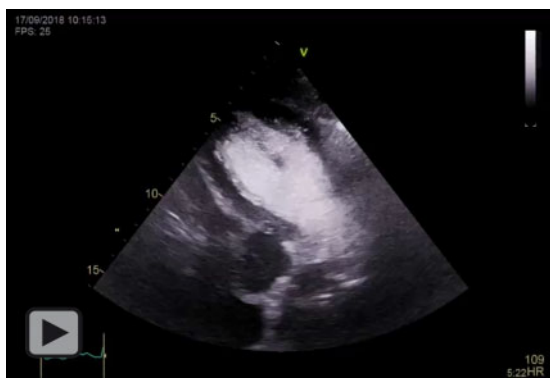
The patient was then transferred to the referral surgical centre and submitted to cardiac surgery. Intraoperative findings were a large haematic pericardial effusion and myocardial rupture with LV pseudoaneurysm that was successfully repaired with a pericardium patch.

Cardiac magnetic resonance imaging performed 3 months later showed preserved biventricular systolic function with segmental WMA and late contrast enhancement pattern compatible with

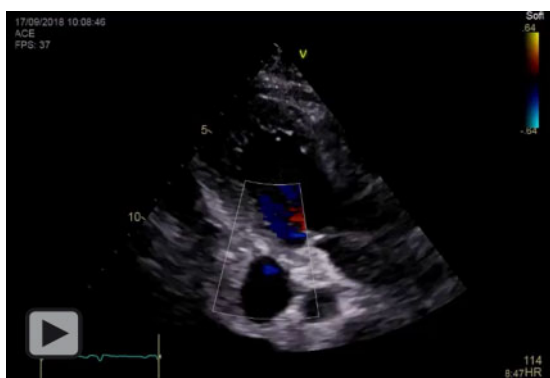
previous inferolateral myocardial infarction. Thickening of the pericardium adjacent to those segments with diffuse contrast uptake was also evident. One year later, the patient remains free of cardiac symptoms or events.

Discussion

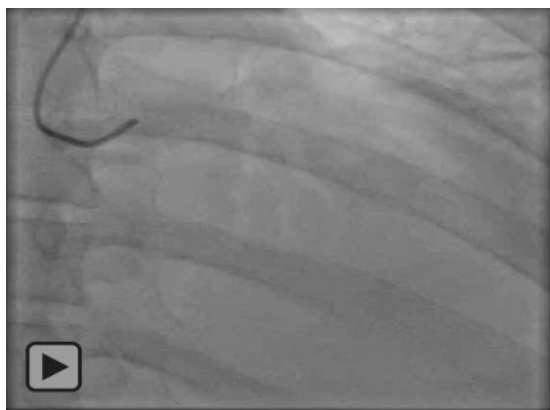
The relationship between SLE and coronary artery disease is well established; however, our patient had angiographically normal



Video 1 The injection of contrast material during TTE further unveiled the presence of a 30 × 30 mm sac with a narrow neck adjacent to the inferolateral wall, with systolic contrast filling, suggesting the diagnosis of a pseudoaneurysm.



Video 2 Medium-large translucent pericardial effusion, predominantly adjacent to the posterior and lateral LV walls. The basal segments of these walls were thinned and there was evidence of bidirectional flow into pericardial space using Doppler colour flow. There were no clinical or echocardiographic signs of cardiac tamponade.



Video 3 Cardiac catheterization exhibited angiographically normal coronary arteries.

coronary arteries, with no signs of coronary atherosclerosis or arteritis.

On the other hand, there are reported cases of transmural myocardial infarction in SLE patients with angiographically normal coronary arteries. Farooq *et al.*,³ who reported one of such cases, hypothesized that coronary microcirculation vasculitis could have been the aetiology of the AMI. Microcirculation thrombosis, demonstrated on histology, was also assumed as a possible cause of myocardial infarction in a young SLE patient in the case described by Curtis *et al.*,⁴ in which the clinical presentation was an acute papillary muscle rupture. In fact, it is well-known that SLE patients have a high risk for arterial and venous thrombosis, which is multifactorial and involves altered blood coagulation pathways mediated by inflammation and endothelial damage associated with glucocorticoids.⁵

Regarding myocarditis-associated cardiac rupture, there are only a few reported cases. In a case described by Fragkouli *et al.*,⁶ histological examination at the rupture site revealed interstitial oedema, myocyte necrosis, and a marked inflammatory infiltrate.

Moreover, in a review of cardiac rupture in Takotsubo cardiomyopathy,⁷ pathology findings at the rupture site included myocyte necrosis and inflammatory infiltrates, implying a prolonged and irreversible process leading to necrosis and cardiac rupture, possible due to demand ischaemia driven by high adrenergic state.

Although the aetiology of the cardiac rupture in our case is not clear, possible explanations could have been a myocardial infarction with normal coronary arteries, due to SLE-associated microcirculation thrombosis or vasculitis or an embolic phenomenon, with subsequent rupture. Although less likely, cardiac rupture in the setting of SLE-associated myocarditis is another hypothesis.

Cardiac rupture is associated with an extremely high mortality unless early diagnosis and urgent surgical intervention are provided. This report describes a unique case of a contained myocardial rupture in a young woman with active SLE and angiographically normal coronary arteries, who underwent cardiac surgical repair with an excellent outcome.

Lead author biography



Dr Ana Neto is a Portuguese cardiology fellow, born in Porto in 1992. She had studied Medicine at University of Porto and is currently in training, at the Centro Hospitalar Tâmega e Sousa (Penafiel, Porto). Moreover, she is a book & cinema lover and someone who is always in the mood to get to know different places in the world.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

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Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as [Supplementary data](#).

Consent: The authors confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

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