

# Acute papillary muscle infarction and rupture in the puerperium complicating Libman–Sacks endocarditis in a patient with systemic lupus erythematosus and antiphospholipid syndrome: a case report

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## Background

Acute heart failure caused by severe mitral regurgitation (MR) due to papillary muscle rupture has been described in the puerperium by case reports; however, the majority of cases of papillary muscle rupture are caused by myocardial infarction. We describe papillary muscle rupture occurring in the postpartum period in a patient with systemic lupus erythematosus (SLE), antiphospholipid syndrome (APLS), and chronic Libman–Sacks endocarditis and explore the multifactorial nature of the papillary muscle infarction and rupture in the setting of postpartum fluid shifts, chronic myocardial injury from Libman–Sacks, and high thrombotic risk.

## Case summary

A 29-year-old woman presented with acute heart failure 2 weeks' postpartum and was found to have acute MR due to a flail leaflet caused by papillary muscle rupture. She proceeded to emergency surgery with mitral valve (MV) replacement and the histology revealed evidence of chronic Libman–Sacks endocarditis and papillary muscle infarction with thrombi in the intramyocardial arteries.

## Discussion

This is the second case report of papillary muscle rupture in the puerperium in a patient with SLE in the literature, the other case was caused by catastrophic APLS. However, in this case, the cause of the rupture is likely to be multifactorial; as a consequence of thrombosis in the microvasculature causing isolated papillary muscle ischaemia, and fibrosis of the muscle due to chronic Libman–Sacks endocarditis resulting in limited pliability which caused rupture of the papillary muscle when faced with the added stress of increased volume that occurs in the puerperium.

## Keywords

Papillary muscle rupture • Systemic lupus erythematosus • Puerperium • Case report

## Learning points

- A rare cause of papillary muscle infarction and rupture due to both thrombosis and tissue abnormalities due to chronic Libman–Sacks endocarditis in the setting of complicated systemic lupus erythematosus and the puerperium.
- Subclinical chronic Libman–Sacks may be missed by transthoracic echocardiogram.

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## Introduction

Papillary muscle rupture most commonly occurs in the setting of myocardial infarction (MI), it has also been seen in the setting of myocarditis, infective endocarditis, and other disorders.<sup>1</sup> It has been described in only four cases in the puerperium: three cases were due to infarction of the papillary muscle and a single case was attributed to Ehler–Danlos syndrome.<sup>1–3</sup> It causes severe mitral regurgitation (MR) which has high mortality without surgical intervention.<sup>4</sup> In most cases, the rupture of the papillary muscle is due to ischaemia of varying mechanisms and rarely due to tissue abnormality—as in the case of a patient with Ehler–Danlos. In this case, we attribute the papillary muscle rupture to isolated infarction associated with the high thrombotic risk state coupled with pre-existing fibrosis of the muscle due to chronic Libman–Sacks endocarditis, as well as the volume changes which occur in the puerperium, all increasing the stress on the papillary muscle and resulting in rupture with almost catastrophic consequences.

## Timeline

Age 12 years	Presentation with idiopathic thrombocytopenic purpura, diagnosed with systemic lupus erythematosus.
Age 19 years	Investigated for Libman–Sacks endocarditis with normal transthoracic echo.
Age 20 years	Spontaneous deep vein thrombosis.
Age 21 years	Class IIIa lupus nephritis.
Age 23 years	Class IV lupus nephritis.
Age 28 years	Planned pregnancy.
Age 28 years	Premature delivery for intrauterine growth retardation (IUGR) at 29 weeks' gestation.
Age 28 years	Presentation with severe chest pain and acute heart failure 2 weeks' postpartum, acute mitral valve replacement for flail mitral regurgitation due to papillary muscle rupture.
Age 28 years	Histology 2 weeks after presentation showed chronic Libman–Sacks endocarditis and thrombus within the intramyocardial arteries.
Age 28 years	2.5 months after presentation with chest pain despite a stormy post-operative course had significant improvement and discharged home.

## Case presentation

A 29-year-old woman with systemic lupus erythematosus (SLE) diagnosed at age 12 years with severe idiopathic thrombocytopenic purpura (ITP), presented acutely while visiting her baby in the Neonatal Intensive Care Unit with sudden onset chest pain and shortness of breath. Her SLE was complicated by spontaneous deep vein thrombosis, triple positive antiphospholipid syndrome (APLS), and Class IV lupus nephritis. This triggered an emergency call to the neonatal

ward, with examination notable for tachycardia, but normotensive and normal oxygen saturation initially. Her chest was clear with dual heart sounds and no oedema. Her medications at presentation were enoxaparin 100 mg and aspirin 100 mg daily, transition to warfarin was planned at 2 weeks' postpartum. Her immunosuppression regime was hydroxychloroquine 400 mg daily, azathioprine 150 mg daily, and prednisone 5 mg daily, other medications included calcitriol 0.25 µg daily and calcium carbonate 1.25 g daily.

Electrocardiogram (ECG) showed sinus tachycardia with no ischaemic changes. Initial blood results shown in [Table 1](#). Chest X-ray showed cardiomegaly with increased interstitial markings ([Figure 1](#)). She deteriorated rapidly over the next hour with climbing oxygen requirements and hypotension and was investigated for pulmonary embolism (PE) and treated with morphine and enoxaparin. She was escorted to radiology from the neonatal ward for the CT pulmonary angiography (CTPA) which showed pulmonary oedema and no PE and was admitted to cardiovascular intensive care (CVICU). On arrival, she was intubated for respiratory distress and, as no transthoracic echocardiogram had been performed, she had a transoesophageal echocardiogram. This demonstrated the posterolateral papillary muscle rupture and flail anterior leaflet with torrential MR and preserved left ventricular (LV) function (see [Supplementary material](#)).

She proceeded to emergency mitral valve (MV) replacement, on transfer to the operating table she had a pulseless electrical activity (PEA) arrest, resuscitated with adrenaline and CPR before return of circulation and the start of operative intervention. The posterolateral papillary muscle was pale and sheared directly off the ventricular wall. The MV was replaced by a 29-mm St Jude's Mechanical valve. At the end of the procedure, VA ECMO was instituted due to persistent severe pulmonary oedema and biventricular failure despite maximal inotropic support.

The histology of the valve revealed preserved architecture with scattered fibrinous deposits and a focal larger fibrinous vegetation with occasional inflammatory cells, with no organisms, consistent with chronic Libman–Sacks endocarditis ([Figure 2](#)). The papillary muscle showed extensive central necrosis with a surrounding rim of acute inflammation consistent with infarction several days old, thrombi were seen within the intramyocardial arteries ([Figure 3](#)). The endocardial surface had mixed inflammatory infiltrates and focally fibrinous material with granuloma-like appearance, in keeping with Libman–Sacks.

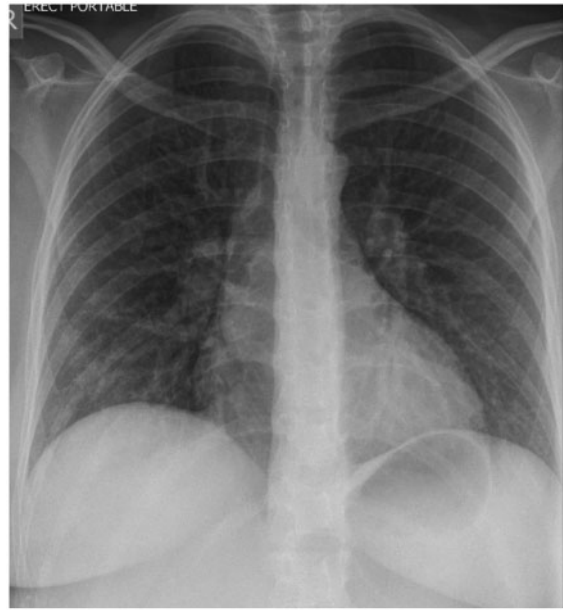
ECMO was required for 2 days, on decannulation and desedation, she had some abnormal movements so magnetic resonance imaging (MRI) brain was performed which showed widespread hypoxic injury. She had paroxysmal atrial fibrillation and mild LV impairment so was commenced on metoprolol CR 166.25mg and cilazapril 1 mg daily and transitioned from heparin back to warfarin and aspirin 100 mg daily. She underwent inpatient rehabilitation after 4 weeks in CVICU before discharge. At this point, her neurological function was normal.

She continued the same immunosuppression regime from admission on discharge as well as the medications mentioned above. She was planned to continue both aspirin and warfarin given the recurrent thrombosis previously.

**Table 1** Initial investigations

Haemoglobin	124 g/L (115–130 g/L)
Platelets	$290 \times 10^9/L$ ( $150\text{--}400 \times 10^9/L$ )
WBC	$13.67 \times 10^9/L$ ( $4\text{--}11 \times 10^9/L$ )
Neutrophils	$11.16 \times 10^9/L$ ( $1.9\text{--}7.5 \times 10^9/L$ )
Troponin T	410 (<15) ng/L

WBC, white blood cell.

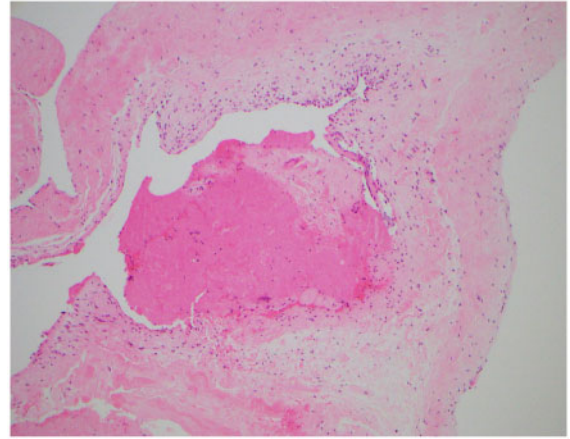
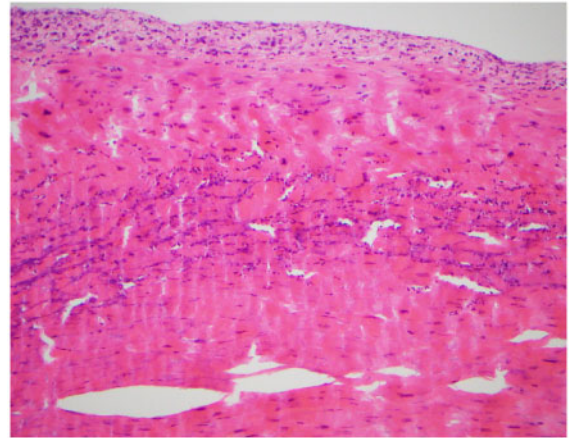
**Figure 1** Chest X-ray: cardiomegaly with increased interstitial markings, patchy opacity at the right base.

## Discussion

This case of acute heart failure caused by papillary muscle rupture in a patient with SLE and APLS. The likely cause is multifactorial; thrombosis, endothelial injury due to SLE/APLS, the postpartum volume changes, and chronic injury and fibrosis of the papillary muscle caused by chronic Libman–Sacks endocarditis.

Papillary muscle histology showed thrombosis within the microcirculation, supporting the role of thrombosis in the infarction. She had multiple risk factors for thrombosis: the autoimmune condition, personal history of thrombosis and postpartum state.

Systemic lupus erythematosus and APLS have high risk of arterial and venous thrombotic complications. The tendency to thrombosis is due to several factors; endothelial injury and platelet activation due to the antibodies produced and inflammation in itself.<sup>5</sup> This patient also had other risk factors for thrombosis—the volume loaded postpartum state causes increased shear stress on the endothelium and an increase in procoagulant factors and reduction in anticoagulant

**Figure 2** Vegetation on mitral valve 10× composed of fibrin with minimal inflammatory cells.**Figure 3** Infarcted papillary muscle 200× necrotic papillary muscle below with viable muscle above with overlying endocardium.

factors and fibrinolytic factors which persists until 6 weeks' postpartum.<sup>6</sup>

The chronic Libman–Sacks endocarditis also contributed to the vulnerability of the papillary muscle. Libman–Sacks is a common complication of SLE with necropsy studies revealing up to 75% of patients having evidence of this at postmortem, while echocardiographic studies have demonstrated its presence in 11% of patients, indicating that those with minor disease may be missed on transthoracic echocardiogram.<sup>7–10</sup> The combination of SLE and APLS together increases the thrombotic risk as well as the prevalence of Libman–Sacks endocarditis.<sup>10</sup> The presentation of valvular involvement ranges from asymptomatic, progressive valvular dysfunction to embolic events. Rarely, it can cause acute valve failure in the setting of valvular perforation.<sup>11</sup> This chronic inflammatory process would have enhanced the

thrombotic risk as well as contributing to shortening of the papillary muscle by fibrosis which would have rendered it less flexible and at greater risk of rupture due to postpartum volume shifts.<sup>12</sup>

Another possibility is that this was part of an early presentation of microvasculature thrombosis as a complication of APLS. She did not have other organ involvement to suggest catastrophic APLS, nor did the blood film show evidence of microangiopathic haemolysis, which is present in 33% of catastrophic APLS.<sup>13</sup> It is possible the brain injury reflected a more widespread coagulopathy but the MRI pattern was more in keeping with hypotension hypoxaemia with involvement of watershed areas. Likewise, it is possible that some of the treatments she received particularly anticoagulation abrogated the full development of catastrophic APLS.

Another possibility is embolic infarction due to Libman–Sacks endocarditis, but this is unlikely given the focal zone of infarction and no evidence of infarction in a coronary artery territory based on ECG and echocardiographic findings. It is also possible that she did have an acute MI, given her history of smoking and inflammatory disease she is likely to have an accelerated atherosclerotic process, but usually, the papillary muscle rupture occurs some days after the initial MI and there was no evidence of this.

This case demonstrates an exceedingly rare, near fatal complication of SLE in combination with APLS, chronic Libman–Sacks endocarditis, and the unique stressors of puerperium.

## Lead author biography



Elizabeth Curtis is a general medicine and cardiology trainee based in Auckland, New Zealand, having attained her medical degree (MBChB) at the University of Otago, Dunedin in 2012. She is interested in general cardiology, heart failure, and imaging and is currently working at North Shore Hospital in the Cardiology department as the cardiac catheterization registrar.

## Supplementary material

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

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

**Consent:** The author/s 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.

**Conflict of interest:** none declared.

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