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CASE REPORT
*Heart failure*

# A case report of acute heart failure and cardiogenic shock caused by catastrophic antiphospholipid syndrome and lupus myocarditis

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**Background** Catastrophic antiphospholipid syndrome and lupus myocarditis are two rare life-threatening conditions.

**Case summary** We present a case of a 47-year-old woman admitted in profound cardiogenic shock due to catastrophic antiphospholipid syndrome and lupus myocarditis requiring advanced heart failure therapies, including early mechanical circulatory support. She improved with steroids, immunoglobulins, mycophenolate, and eculizumab.

**Discussion** This case highlights the importance of early identification of cardiogenic shock secondary to catastrophic antiphospholipid syndrome and lupus myocarditis, the arrhythmogenic complications of myocarditis, and the subsequent management of the disease progression with mechanical and medical support.

**Keywords** Antiphospholipid syndrome • Systemic lupus erythematosus • Cardiogenic shock • Mechanical circulatory support • Myocarditis • Torsades de Pointes • Case report

**ESC Curriculum** 6.4 Acute heart failure • 6.1 Symptoms and signs of heart failure • 7.4 Percutaneous cardiovascular post-procedure • 7.3 Critically ill cardiac patient • 7.1 Haemodynamic instability

## Learning points

- Catastrophic antiphospholipid syndrome and lupus myocarditis can cause cardiogenic shock. Endomyocardial and skin biopsy should be used to support the diagnosis.
- Early mechanical and pharmacologic support is crucial in myocarditis-induced cardiogenic shock. Providers should be vigilant as life-threatening arrhythmias can occur during the acute phase of severe myocarditis, even after the initiation of steroids and immunosuppressants.

## Introduction

Catastrophic antiphospholipid syndrome (CAPS) and lupus myocarditis are two rare and life-threatening conditions. The diagnosis of CAPS requires at least three affected organ systems within a 7-day time frame, biopsy evidence of microthrombi, and laboratory evidence of antiphospholipid antibodies.[1] The estimated incidence of CAPS among patients with antiphospholipid syndrome (APLS) was reported as <1% over a 7-year time frame in one study.[2] Among patients with systemic lupus erythematosus (SLE), the prevalence of lupus myocarditis has been reported to be between 8% and 25%.[3]

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There have been only a few of case reports describing concurrent CAPS and lupus myocarditis with cardiogenic shock.[4,5] These cases occurred post-partum or with withdrawal of immunosuppressives.[4] The treatment of CAPS and lupus myocarditis focuses on immunosuppression, anticoagulation, and supportive therapy. Despite these treatments, CAPS still carries a high mortality rate. Early recognition with prompt involvement of a multidisciplinary team and early aggressive supportive care is essential in the care of these patients. In this case report, the successful management of a patient in cardiogenic shock due to catastrophic antiphospholipid syndrome and lupus myocarditis is presented.

## Timeline

| Day | Event |
|---|---|
| Two weeks before admission to our hospital | A left ureteral stent is placed for nephrolithiasis. |
| | A large perinephric haematoma develops; home warfarin is held; vitamin K and FFP are given. |
| | Renal artery angiogram is performed and a right femoral pseudoaneurysm is injected with thrombin. |
| Three days before admission to our hospital | Suffers a cardiac arrest, requiring epinephrine, and intubation. A new left bundle branch block is seen on electrocardiogram. |
| A day before admission to our hospital | Non-obstructive coronary artery disease is seen on catheterization and a ventriculogram demonstrates an ejection fraction of 15%. |
| | An intra-aortic balloon pump (IABP) is placed initially and later upgraded to an Impella. |
| | Methylprednisolone is started; endomyocardial biopsy is performed, and the patient is transferred to our hospital. |
| Day 1 | Arrives on dobutamine, epinephrine, and vasopressin with an Impella. The Impella is dislodged and switched to an IABP. |
| Day 2 | Skin biopsy with intravascular fibrin thrombi is consistent with catastrophic antiphospholipid syndrome. |
| | Endomyocardial biopsy with mononuclear infiltrates without thrombi or vasculitis is consistent with lupus myocarditis. |
| | Heparin, high-dose hydrocortisone, intravenous immunoglobulin and continuous veno-venous haemofiltration are continued. |
| Day 5 | IABP is weaned, and the patient is extubated. |
| Day 11 | Suffers a premature ventricular contraction-triggered Torsades de Pointes that self-resolved. |
| Day 13 | Eculizumab and mycophenolate mycortif are started. |
| Day 14 | Vasopressors are weaned off. |
| Day 25 | A long-term haemodialysis catheter is placed. |
| Day 30 | Patient is discharged on warfarin, eculizumab, mycophenolate, midodrine, and intermittent haemodialysis with good cardiac recovery. |

## Case presentation

A 47-year-old woman with a history of SLE, APLS, recent bariatric surgery with rapid weight loss, and chronic kidney disease Stage III was transferred to our hospital for cardiogenic shock.

Six months prior to the index hospitalization, she underwent bariatric surgery with an ensuing 150-pound weight loss. A transthoracic echocardiogram (TTE) prior to surgery reported a left ventricular ejection fraction (EF) of 50% without significant valvular disease. She was on stable doses of hydroxychloroquine, mycophenolate mycortif, and warfarin for her known APLS.

Two weeks prior to admission, she developed nephrolithiasis, requiring left ureteral stent placement. The procedure was complicated by a large perinephric haematoma. Her home warfarin was held while vitamin K, fresh frozen plasma, and packed red blood cells were given. Repeat CT scan showed enlargement of the haematoma. An ensuing renal arterial angiogram did not demonstrate active haemorrhage and a right femoral pseudoaneurysm was treated with thrombin injection during the angiogram.

She remained in the hospital for pain control; however, 11 days later, she developed sudden onset chest pain and dyspnoea, followed by a cardiac arrest. She achieved return of circulation after one round of compressions, one ampule of epinephrine, and was intubated. Electrocardiogram (ECG) post-arrest demonstrated a new left bundle branch block (LBBB). Her emergent cardiac catheterization revealed non-obstructive coronary artery disease but a new globally depressed EF of 15% on ventriculogram (*Figure 1*).

An intra-aortic balloon pump (IABP) was placed and later upgraded to an Impella 5.0 device. Repeat TTE revealed an EF of 15–20%, mild mitral regurgitation, and moderate aortic regurgitation. The patient was started on continuous norepinephrine, epinephrine, and vasopressin. Blood work demonstrated evidence of multiorgan dysfunction with elevated liver enzymes, rising creatinine, and troponin I of >40 ng/mL. Given worsening renal function, the patient was started on continuous veno-venous haemofiltration (CVVH). Methylprednisolone 1000 mg was started for 3 days as recommended by rheumatology. She underwent an endomyocardial biopsy on the day prior to transfer to our facility for evaluation of durable mechanical support.

On arrival to our hospital, she was intubated and sedated on dobutamine 2.5 mcg/kg/h, epinephrine 5 mcg/min, and vasopressin 1.8 units/h. ECG demonstrated sinus rhythm with a LBBB (*Figure 2*). Repeat TTE revealed that the Impella was inserted deep in the left ventricular cavity. Unfortunately, the device was dislodged during an attempted repositioning at bedside and could not be replaced due to a congenital vascular abnormality; the device was removed and replaced with an IABP.

Her physical examination was notable for diffuse nonblanching purpuric lesions with haemorrhagic bullae over the flank and abdomen

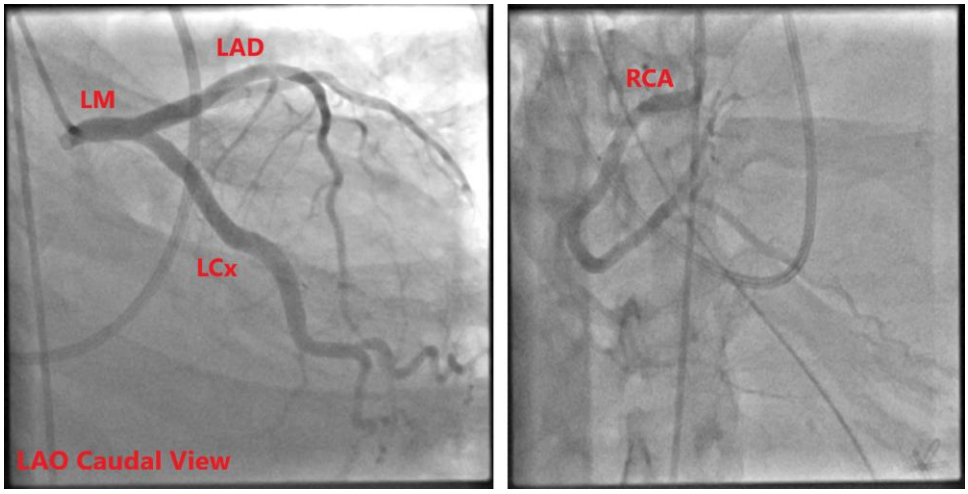


**Figure 1** Normal coronaries on left heart catheterization. LM, left main coronary artery; LAD, left anterior descending coronary artery; LCx, left circumflex coronary artery; LAO, left anterior oblique; RCA, right coronary artery.

with negative Nikolsky sign (*Figure 3*). Given the vasculitic appearance, dermatology performed a skin biopsy. The dermatopathology demonstrated coagulopathy with intravascular fibrin thrombi while the endomyocardial biopsy demonstrated mononuclear inflammatory infiltrates associated with myocardial damage. No vascular thrombi, vasculitis, giant cells or eosinophilic granulocytes were identified on the endomyocardial biopsy. Given the skin pathology and endomyocardial biopsy, the patient was continued on high-dose hydrocortisone for suspected CAPS and lupus myocarditis. She was also kept on unfractionated heparin with monitoring of factor Xa activity given elevated activated partial thrombin time in the context of APLS. Other laboratory work was notable for hypocomplementemia, positive ANA, and positive anti-SSA antibody.

The patient completed 5 days of intravenous immunoglobulin (IVIG) and began a slow steroid taper over 6 weeks. The IABP was removed on hospital day 5 and she was extubated the same day. Given the suspicion that her ongoing renal disease may be secondary to residual effects of CAPS, she was treated with eculizumab. She was also started on mycophenolate mycortif for her SLE.

On Day 11 of hospitalization, the patient suffered a premature ventricular contraction (PVC)-triggered Torsades de Pointes (*Figure 4*). Her arrhythmia self-resolved without intervention and no further episodes of sustained ventricular arrhythmia occurred during her hospitalization.

Over the following hospital days, she was weaned off of intravenous vasopressor medications though still required midodrine for blood

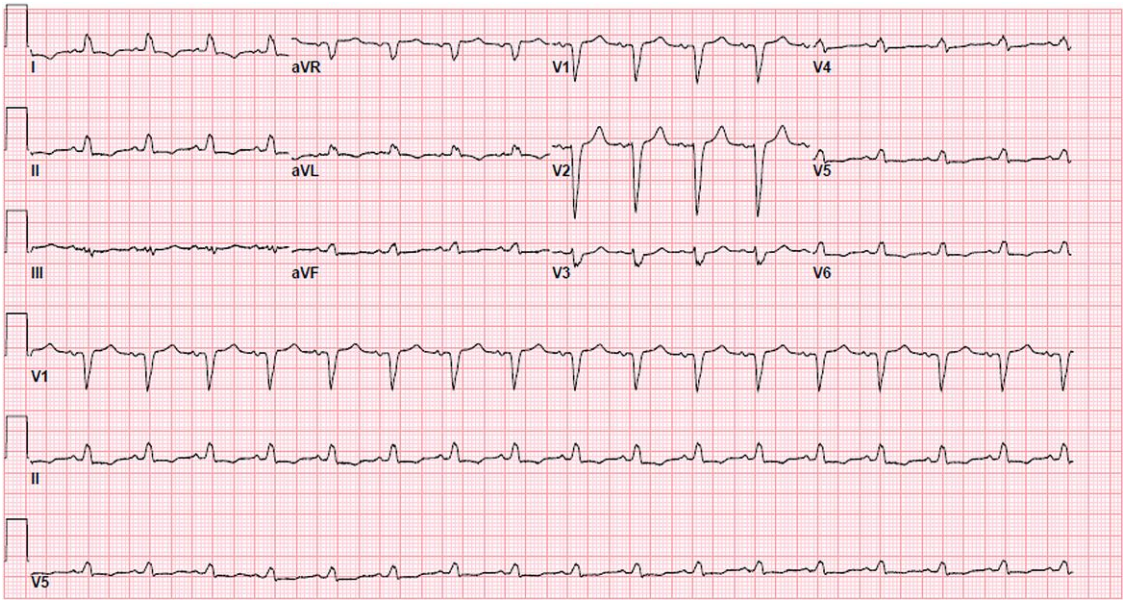


**Figure 2** Electrocardiogram on admission to our hospital demonstrating sinus rhythm with a left bundle branch block morphology.

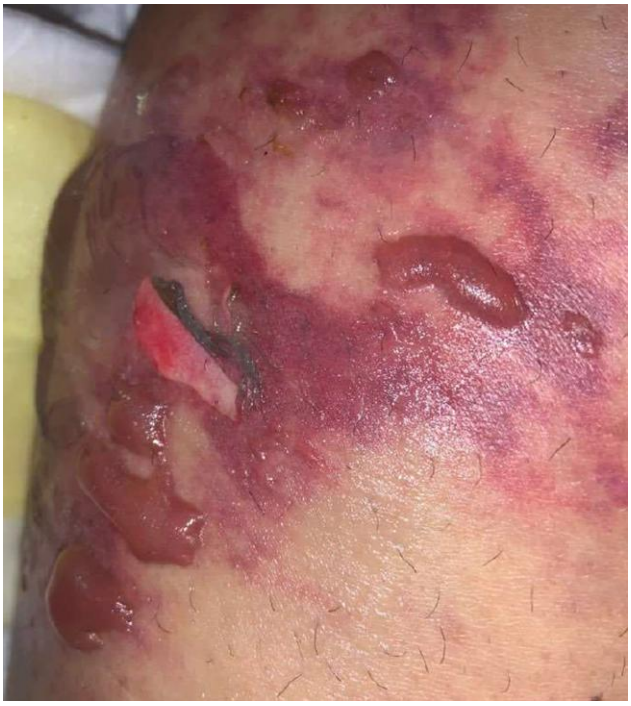

**Figure 3** Purpuric lesions with bullae on physical examination.

pressure support. She was discharged to an acute rehabilitation facility on haemodialysis with good recovery of left ventricular function (EF 47%) with normal right ventricular function without guideline-directed medical therapy. She was maintained on warfarin. Infusions of eculizumab and mycophenolate mycortif were given in hopes of promoting renal recovery.

## Discussion

This is a rare case of combined CAPS and lupus myocarditis leading to cardiogenic shock and renal dysfunction. The possible trigger in this case was the combination of anticoagulation withdrawal for an active bleed and the local injection of thrombin for a pseudoaneurysm.

Our differential diagnosis included CAPS, lupus myocarditis, hydroxychloroquine toxicity,[6] and wet beriberi in the setting of recent bariatric surgery. CAPS appeared to be the unifying diagnosis given the recent changes to her anticoagulation, history of thrombin injection, rapid decline, multiorgan failure, and skin findings. Interestingly, the microthrombotic disease on skin biopsy was consistent with CAPS, while the inflammatory infiltrate without evidence of thrombi on endomyocardial biopsy was more suggestive of lupus myocarditis. Thus, both CAPS and lupus myocarditis were active processes in our case.

While awaiting histological diagnosis, the use of early empiric high-dose steroids and IVIG likely helped modify the disease processes. The cornerstone treatment for CAPS traditionally includes IVIG, plasmapheresis, and anticoagulation.[7] Of note, due to significant

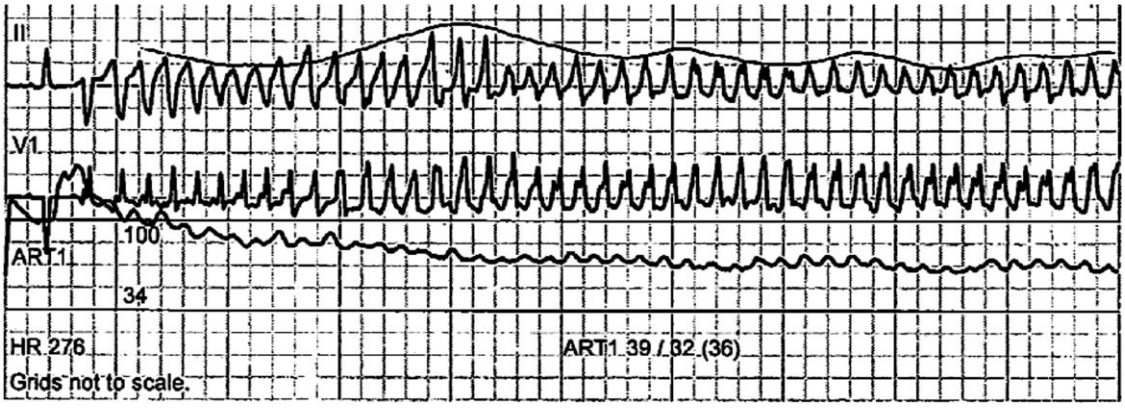


**Figure 4** PVC-triggered Torsades de Pointes.

vasopressor requirements, plasmapheresis was withheld given the concern that plasmapheresis could remove the vasoactive agents. It was presumed that the underlying myocarditis led to the Torsades de Pointes and that the continuation of steroids and immunosuppressants would resolve the ventricular ectopy.

Despite improvement in cardiac function, she still required haemodialysis on discharge. The decision to utilize eculizumab for her renal dysfunction is based on a few cases series in patients with CAPS.[8] The anti-C5 monoclonal antibody presumably aborts the thrombotic process and prevents further progression of renal injury. In the end, we were pleased that the patient had almost complete cardiac recovery from such a catastrophic event as the mortality rates for CAPS approaches 50%.[9]

## Conclusions

APLS and SLE are both rare entities that can have devastating complications. This case highlights the importance of early disease recognition, empiric high-dose steroid administration, eculizumab for renal dysfunction, and aggressive early pharmacological and mechanical circulatory support while waiting for a histological diagnosis. This case further emphasizes vigilance for life-threatening arrhythmias in patients with severe myocarditis.

## Lead author biography

Ashton Lai is a cardiovascular disease fellow at the Mount Sinai Hospital in New York City. He graduated from the Yale School of Medicine with an MD/PhD and subsequently completed his internal medicine residency at the Mount Sinai Hospital.

## Specialties involved other than cardiology

- Rheumatology
- Nephrology
- Haematology
- Critical Care
- Dermatology

## 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 images and associated text has been obtained from the patient in line with COPE guidance. IRB approval is not required for this case report.


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