SAGE Open Medical Case Reports
Volume 11: 1–4

DOI: 10.1177/2050313X231220803
journals.sagepub.com/home/sco
Sage

# Cardiac arrest in the setting of probable catastrophic antiphospholipid syndrome in young patient with a history of COVID infection and polyglandular disorder—Case report

**Seyed M Nahidi[1], Yash Garg[1], Devi S Mahadeo[2], Manjari Sharma[1], Cesar Acosta[1], Karthik Seetharam[1] and Mukosolu F Obi[1]**

## Abstract

Antiphospholipid syndrome is an autoimmune disorder characterized by arterial and venous thrombosis and recurrent spontaneous abortions due to the persistent presence of antiphospholipid antibodies. Probable Catastrophic antiphospholipid (Catastrophic antiphospholipid-like syndrome) is a life-threatening presentation of antiphospholipid syndrome which manifests as intravascular thrombosis, leading to rapid onset of symptoms and involvement of multiple organ systems. We present a case of a 28-year-old woman with a history of polyglandular autoimmune syndrome, systemic lupus erythematosus, provoked bilateral deep vein thrombosis in the setting of Severe Acute Respiratory Syndrome Coronavirus 2 (SARS-CoV-2) infection 2 years prior, and hypothyroidism who presents with a cardiac arrest in the setting of an acute ST-elevation myocardial infarction with thromboembolic occlusion of two coronary arteries simultaneously in the setting of noncompliance with anticoagulation for the past 1 week. Her presentation was further complicated by acute hypoxic respiratory failure due to diffuse alveolar hemorrhage during the hospital course with progressive multiorgan failure and eventual death. Catastrophic antiphospholipid is associated with high morbidity and mortality, thus a timely diagnosis and multidisciplinary approach to management is needed for evaluation and management.

## Keywords

Cardiac arrest, myocardial infarction, antiphospholipid syndrome, catastrophic antiphospholipid syndrome

Date received: 30 May 2023; accepted: 20 November 2023

## Introduction

Antiphospholipid syndrome (APS), an autoimmune disorder, is characterized by hypercoagulation (arterial/venous thrombosis), recurrent spontaneous abortion, and antiphospholipid antibodies (aPL).[1] APS can be classified as primary or secondary where only the latter is associated with concurrent autoimmune disorders.[2] A rare and life-threatening presentation of APS is Catastrophic Antiphospholipid Syndrome (CAPS), which manifests as diffuse microthrombosis leading to multiorgan failure.[3] CAPS is diagnosed if thromboses simultaneously develop in >3 organs within 1 week, along with histopathological evidence of multiple microthrombosis in the presence of high aPL titres.[3] CAPS can appear as the initial presentation of APS and can be precipitated by risk factors, including concomitant autoimmune diseases, abrupt cessation of anticoagulation therapy in known cases of APS and infection.[3] Considering its high rate of morbidity and mortality, timely diagnosis and intervention can improve prognosis. The advent of triple therapy, a combination of anticoagulation,

[1]Internal Medicine Resident at Wyckoff Heights Medical Center, Brooklyn, NY, USA
[2]St George's University, Grenada, West Indies

**Corresponding Author:**
Yash Garg, Wyckoff Heights Medical Center, 374 Stockholm Street, Brooklyn, NY 11237, USA.
Email: yashgargmd@gmail.com

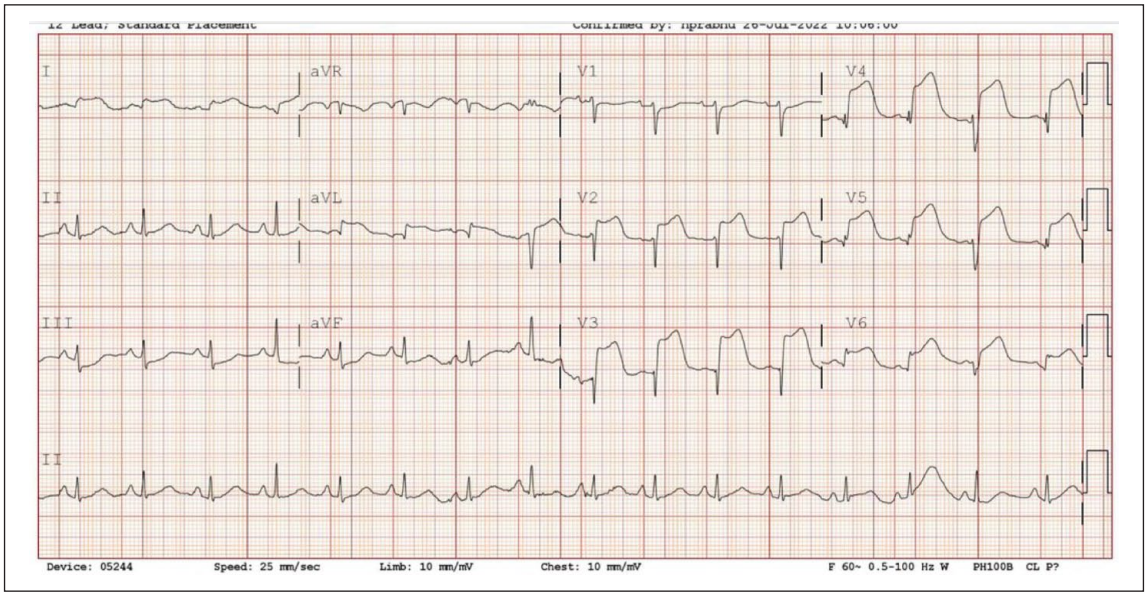


**Figure 1.** 12-Lead EKG at the time of presentation to the emergency department conveying ST-segment elevations in the anterior and lateral leads.

corticosteroids and intravenous immunoglobulin (IVIG)/plasma exchange, has contributed to an increased survival rate.

## Case presentation

A 28-year-old female presented to our emergency department (ED) after being found with pulseless electrical activity that converted to ventricular fibrillation, for which she received defibrillation. Patient achieved return of spontaneous circulation after 21 minutes and was intubated. Patient was started on a norepinephrine drip after fluid resuscitation due to hypotension. Initial Electrocardiogram (EKG), conveyed sinus rhythm with ST-segment elevations in anterior and anterolateral precordial leads (Figure 1). Patient was immediately taken for cardiac catheterization.

Coronary angiography showed acute total occlusion of the left anterior descending artery (LAD) and RAMUS intermedius with Thrombolysis in myocardial infarction (TIMI) and zero flow. TIMI flow is a well graded system which explains the coronary reperfusion where TIMI three flow means flow to distal vessel by third cardiac cycle and TIMI zero means a total occlusion. Patient had percutaneous transluminal coronary angioplasty of mid to distal LAD and percutaneous coronary intervention with one drug eluting stent of Ramus intermedius. TIMI three flow was never fully restored in both vessels due to the thrombus that completely occluded the distal LAD at the apex (Figure 2). The patient required intracoronary nitrates/nipride/and thrombus aspiration. During the cardiac catheterization, the patient developed severe hypoxemia and hypotension secondary to pulmonary hemorrhage which led to an emergent bronchoscopy where greater than 1 L of blood aspirated. Patient developed hypovolemic shock and had a ventricular fibrillation rhythm for which patient underwent defibrillation. In the cardiac catheterization unit, the patient received packed red blood cells via rapid transfusion and was transferred to the ICU for the second bronchoscopy while on vasopressors. About 4 hours after presentation, family members were identified who revealed the patient's past medical history included Systemic Lupus Erythematosus (SLE), bilateral lower extremity Deep Venous Thrombosis (DVT) (initially due to SARS-CoV-2 in 2020) on enoxaparin, Hypothyroidism, and polyglandular autoimmune disease (on

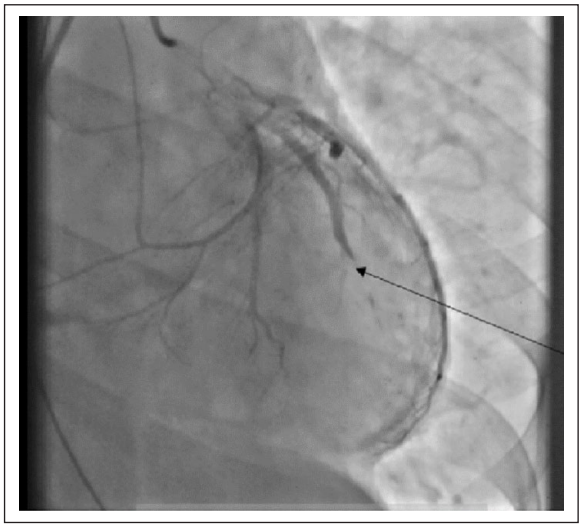

**Figure 2.** Cardiac catheterization of left coronary artery demonstrating LAD occlusion TIMI zero flow (By the arrow).

estrogen therapy), and also informed them that the patient ran out of her enoxaparin about a week prior to admission.

On hospital day 2, echocardiogram conveyed a left ventricular ejection fraction of 20%–25% and a possible left ventricular thrombus. Due to hemodynamic instability, transfer to tertiary center was not possible during this admission.

On hospital day 3, the patient became hypoxic, for which she underwent a bronchoscopy where 900 ml of blood was suctioned. On hospital day 4, the patient did not improve. Given the patient's history, rheumatology was consulted due to suspicion of CAPS and was started on IVIG, high-dose steroids and antiphospholipid antibody was collected.

On day 5, the patient's condition worsened and eventually led to cardiac arrest and death. Postmortem aPL was found in high titres (Cardiolipin IgM Antibodies- 48 MPL U/mL, beta-2 glycoprotein IgM antibodies- 54.2 U/mL and phosphatidylserine IgM antibodies- 19 U/mL).

## Discussion

APS has an estimated incidence of 2.1 cases per 100,000 and a prevalence of 50 per 100,000.[1] Secondary APS is associated with underlying disorders such as SLE.[2] An unusual manifestation of APS is CAPS. This can be the initial presentation of APS in nearly 50% of patients while the remaining subset has a known history of APS, appearing in approximately 1% of these patients.[3] Albeit rare, it is a life-threatening condition with a mortality rate of 36.9%.[4] APS and CAPS can manifest as a spectrum of clinical disorders. CAPS is diagnosed if thromboses simultaneously develop in more than three organs within 1 week, along with histopathological evidence of multiple microthrombosis in the presence of high aPL titres.[5] A definitive diagnosis of CAPS can be made once all four criteria have been met. If, however, only a combination of these criteria is present, the patient can be diagnosed with a CAPS-like disease.[3] This patient developed multiple thromboses, including deep vein thrombosis and myocardial infarction. There was also diffuse alveolar hemorrhage evident by the large volume of blood aspirated on bronchoscopy which may have occurred secondary to microthrombosis in the lungs. While this is a reasonable deduction, the pulmonary microthrombosis was not confirmed with imaging due to the patient's hemodynamic instability and constant high risk of mortality. The Task Force on CAPS performed an analysis on 547 CAPS patients and found that the kidneys (74%), lungs (55%), brain (56%), heart (53%), and skin (45%) were most commonly affected, whereas peripheral vessels (37%) were less commonly affected.[6]

The serological hallmark of CAPS are aPLs, which induce a prothrombotic and proinflammatory state due to the activation of endothelial cells, platelets, and immune cells. Additionally, they have been shown to impair fibrinolysis and activate complement.[3] SARS-CoV-2 has also been shown to induce a hypercoagulable state. According to studies, not only is the virus associated with higher titres of aPL, but it has also been theorized to potentiate the effect of aPL, leading to an increased widespread microvascular injury and coagulopathy by activating endothelial cells, platelets, and complement.[7,8] There is limited evidence that directly supports the association between CAPS and COVID-19. However, since infections are a known trigger, it is conceivable that SARS-CoV-2 can induce CAPS. In this case, the patient with a history of SLE and APS contracted the SARS-CoV-2 infection in 2020 and later, within the same year, developed bilateral DVT. It is possible that the SARS-CoV-2 virus increased this patient's risk of developing bilateral DVT and, later, CAPS.

Apart from the past medical history of this patient, another significant aspect of this case is the patient's abrupt cessation of anticoagulation therapy. The prospect of long-term anticoagulation therapy in patients with APS is reduction in the risk of recurrent thrombotic events and the progression to CAPS.[9] A retrospective series demonstrated a high rate of recurrent thromboembolism in patients with APS that stopped anticoagulation, approximating 30% per year.[10] In congruence with the aforementioned study, the patient in this case also exhibited detrimental recurrent thrombosis with progression to CAPS and a lethal outcome after abruptly discontinuing enoxaparin.

Patient education and periodic follow-up is crucial which may help to improve survival rate. A combination of anticoagulation therapy (either heparin or warfarin), corticosteroid, and IVIG/plasma exchange; a triple therapy has shown a higher survival rate among patients with CAPS. A study was conducted to assess the efficacy of triple therapy on 471 patients in the CAPS registry. Triple therapy was prescribed in 189 instances (40.1%), 270 (57.3%) with other combinations and 12 (2.5%) with none of these treatment options; the mortality rate of these three groups was 28.6%, 41.1%, and 75%, respectively (adjusted odds ratio = 9.7, 95% CI: 2.3, 40.6).[11] The high morbidity and mortality of CAPS warrants multidisciplinary collaboration which can greatly improve prognosis.

## Conclusion

Although rare, APS has a potential to progress to CAPS, particularly in the context of abrupt cessation of long-term anticoagulation therapy. CAPS can also be the initial presentation of APS in a subset of patients. In addition to APS and autoimmune conditions such as SLE, the risk of developing CAPS is greater if a patient has been infected with SARS-CoV-2. CAPS can manifest as a spectrum of clinical disorders, including myocardial infarction, stroke, and pulmonary embolism. Despite its high mortality, an increased survival rate has been shown with multidisciplinary approach and the administration of triple therapy, including anticoagulation, corticosteroid, and IVIG/plasma exchange in the management of CAPS. In a setting where some treatment options are not available, quick stabilization and transfer of the patient to a tertiary center is warranted, however, it is not always

possible. As an example, our patient had recurrent hemorrhages and had unstable vitals throughout her stay.

## Acknowledgements

Not applicable.

## Authors' contribution

The author's below have all read and consented to the submission of this article. S.M.N. and Y.G. contributed in data collection, review, and write-up; D.S.M., M.S., C.A., K.S., and M.F.O. contributed to write-up.

## Declaration of conflicting interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

## Funding

The author(s) received no financial support for the research, authorship, and/or publication of this article.

## Ethics approval

Our institution does not require ethical approval for reporting individual cases or case series.

## Informed consent

Informed consent was obtained from the legally authorized representative of the deceased subject for the publication of the case report.

## ORCID iDs

Seyed M Nahidi https://orcid.org/0000-0002-5468-8838

Yash Garg https://orcid.org/0000-0002-3167-2657

## References

1. Duarte-García A, Pham MM, Crowson CS, et al. The epidemiology of antiphospholipid syndrome: a population-based study. *Arthritis Rheumatol (Hoboken, N.J.)* 2019; 71(9): 1545–1552.
2. Gezer S. Antiphospholipid syndrome. *Disease-a-Month* 2003; 49(12): 696–741.
3. Nayer A and Ortega LM. Catastrophic antiphospholipid syndrome: a clinical review. *J Nephropathol* 2014; 3(1): 9–17.
4. Tinti MG, Carnevale V, Inglese M, et al. Eculizumab in refractory catastrophic antiphospholipid syndrome: a case report and systematic review of the literature. *Clin Exp Med* 2019; 19(3): 281–288.
5. Carmi O, Berla M, Shoenfeld Y, et al. Diagnosis and management of catastrophic antiphospholipid syndrome. *Exp Rev Hematol* 2017; 10(4): 365–374.
6. Cervera R, Rodríguez-Pintó I, Legault K, et al. 16th International Congress on Antiphospholipid Antibodies Task Force report on catastrophic antiphospholipid syndrome. *Lupus* 2020; 29(12): 1594–1600.
7. Chidharla A, Syed SB, Chatterjee T, et al. A case report of COVID-associated catastrophic antiphospholipid syndrome successfully treated with eculizumab. *J Blood Med* 2021; 12: 929–933.
8. El Hasbani G, Taher AT, Jawad A, et al. Covid-19, antiphospholipid antibodies, and catastrophic antiphospholipid syndrome: a possible association? *Clinical Medicine Insights: Arthritis and Musculoskeletal Disorders* 2020; 13: 117954412097866.
9. Rahman A. Management of antiphospholipid syndrome. *Clin Rheumatol* 2020; 39(7): 2111–2114.
10. Khamashta M, Taraborelli M, Sciascia S, et al. Antiphospholipid syndrome. *Best Pract Res Clin Rheumatol* 2016; 30(1): 133–148.
11. Ramírez E, Romero-Garrido JA, López-Granados E, et al. Symptomatic thromboembolic events in patients treated with intravenous-immunoglobulins: results from a retrospective cohort study. *Thrombosis Res* 2014; 133(6): 1045–1051.