CASE REPORT

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Fulminant Multisystem Thrombosis in a Middle-Aged Male: A Case Report

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Abstract

Fulminant multisystem thrombosis is a life-threatening condition that requires rapid diagnosis and aggressive management due to its high morbidity and mortality. We describe a 44-year-old male with no prior autoimmune or thrombotic history who presented with dyspnoea, abdominal pain, haematuria, and chest discomfort. Imaging revealed widespread simultaneous arterial and venous thromboses involving the pulmonary arteries, inferior mesenteric artery, renal vasculature, and deep veins of the right lower limb. Antiphospholipid antibody testing demonstrated a positive lupus anticoagulant and elevated β2-glycoprotein I IgM, raising strong suspicion for catastrophic antiphospholipid syndrome (CAPS). Despite prompt initiation of anticoagulation, immunomodulatory therapy, and supportive care, the patient developed anterior spinal artery thrombosis causing paraparesis, followed by a fatal acute myocardial infarction due to presumed coronary thrombosis. This case underscores the fulminant nature of multisystem thrombosis, highlights the diagnostic challenges of de novo CAPS, and emphasizes the need for early recognition and aggressive multimodal treatment to improve outcomes, while also noting that although CAPS is more common in females, it can rarely occur in males as demonstrated in this case.

Introduction

Antiphospholipid syndrome (APS) is an acquired autoimmune thrombophilia defined by recurrent arterial or venous thrombosis and pregnancy morbidity associated with persistent antiphospholipid antibodies (aPLs), including lupus anticoagulant (LA), anticardiolipin (aCL), and anti-β2-glycoprotein I antibodies (aβ2GPI)[1]. APS may occur as a primary condition or secondary to systemic autoimmune diseases, most commonly systemic lupus erythematosus. Catastrophic antiphospholipid syndrome (CAPS), first described by Asherson in 1992[2], is an uncommon but severe APS variant that occurs in fewer than 1% of APS patients[3]. It is characterised by rapidly progressive thrombosis involving multiple organ systems over days to weeks. Despite advances in anticoagulation and immunomodulatory therapy – including corticosteroids, intravenous immunoglobulin (IVIG), and plasma exchange – mortality remains between 30% and 50%[4]. Although CAPS typically affects middle-aged women with known APS or autoimmune disease, it can arise *de novo* without identifiable triggers, and atypical presentations pose significant diagnostic challenges. It may affect men rarely, as in our case.

Case Presentation

A 44-year-old male with a history notable only for hypertension and no previous thrombotic, autoimmune, or inflammatory illness presented with 5 - 6 days of progressive shortness of breath, mild haemoptysis, atypical chest pain, diffuse abdominal pain accompanied by nausea and vomiting, and macroscopic haematuria. On arrival, he appeared acutely ill and dyspnoeic, with tachycardia, tachypnoea, normotension, and an afebrile status. Physical examination revealed decreased breath sounds in the left hemithorax and diffuse abdominal tenderness without guarding or rebound. Initial cardiovascular and neurological evaluations were unremarkable. Laboratory investigations (Table I) demonstrated neutrophilic leucocytosis (20 x 10^3/μL), thrombocytosis (556 x 10^3/μL), elevated alkaline phosphatase (176 U/L), hypoalbuminaemia (2.1 g/dL), and preserved renal function (blood urea 35.6 mg/dL, serum creatinine 1.2 mg/dL, uric acid 7.1 mg/dL). Coagulation profile was normal, with a Prothrombin Time (PT) of 12.5 seconds, an Activated Partial Thromboplastin Time (aPTT) of 30.0 seconds, and an International Normalised Ratio (INR) of 1.0. Additionally, fibrinogen levels were measured at 300 mg/dL, and the peripheral smear showed no schistocytes, making disseminated intravascular coagulation (DIC) unlikely.

Different imaging studies revealed extensive multiorgan thrombosis. Doppler ultrasound of both lower limbs identified acute thrombosis of the right femoral, popliteal,

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and peroneal veins (Fig. 1). CT pulmonary angiography demonstrated thrombi in the right lower lobe segmental pulmonary arteries and a large embolus in the left main pulmonary artery (Fig. II). Contrast-enhanced CT of the abdomen revealed multiple renal infarcts, and CT angiography identified thrombosis of the inferior mesenteric artery. Given the presence of multiple simultaneous arterial and venous thromboses affecting several organ systems within a short interval, a systemic hypercoagulable disorder was strongly suspected, and evaluation for APS was pursued. The patient had no history of thrombosis, surgery, trauma, or infection, and no evidence of disseminated intravascular coagulation, which increased the suspicion of catastrophic antiphospholipid syndrome.

Antiphospholipid testing revealed a positive β2-glycoprotein I IgM level (>20) and a positive lupus anticoagulant, with negative anticardiolipin antibodies, supporting an antiphospholipid-mediated thrombotic process. On hospital day 4, he experienced sudden onset sensorimotor paraparesis. MRI of the whole spine demonstrated acute anterior spinal artery thrombosis corresponding to his neurological deficits. The rapid development of widespread arterial and venous thromboses affecting the pulmonary, renal, mesenteric, venous, and spinal circulations within one week fulfilled the diagnostic criteria for CAPS.

Table I: Investigations reports.

Investigation	Result	Normal Range/Reference
Complete Blood Count (CBC)	Neutrophilic leucocytosis: $20 \times 10^3/\mu L$ Thrombocytosis: $556 \times 10^3/\mu L$	WBC: $4.0 - 11.0 \times 10^3/\mu L$ Platelets: $150 - 400 \times 10^3/\mu L$
Liver Function Tests (LFTs)	Elevated alkaline phosphatase: 176 U/L	Alkaline Phosphatase: 30 - 120 U/L
Renal Function Tests (RFTs)	Blood urea: 35.6 mg/dL Serum creatinine: 1.2 mg/dL Uric acid: 7.1 mg/dL	Urea: 10 - 20 mg/dL Creatinine: 0.6 - 1.2 mg/dL Uric acid: 3.5 - 7.2 mg/dL
Coagulation Profile	Prothrombin Time (PT): 12.5 seconds Activated Partial Thromboplastin Time (aPTT): 30.0 sec International Normalised Ratio (INR): 1.0 Fibrinogen: 300 mg/dL	PT: 11.0 - 14.0 seconds aPTT: 25.0 - 35.0 seconds INR: 0.9 - 1.1 Fibrinogen: 200 - 400 mg/dL
Peripheral Blood Smear	No schistocytes	No schistocytes
ESR	ESR- 12 mm/hr	ESR- 0 - 15 mm/hr
CRP	CRP- 2 mg/L	CRP- less than 3mg/L

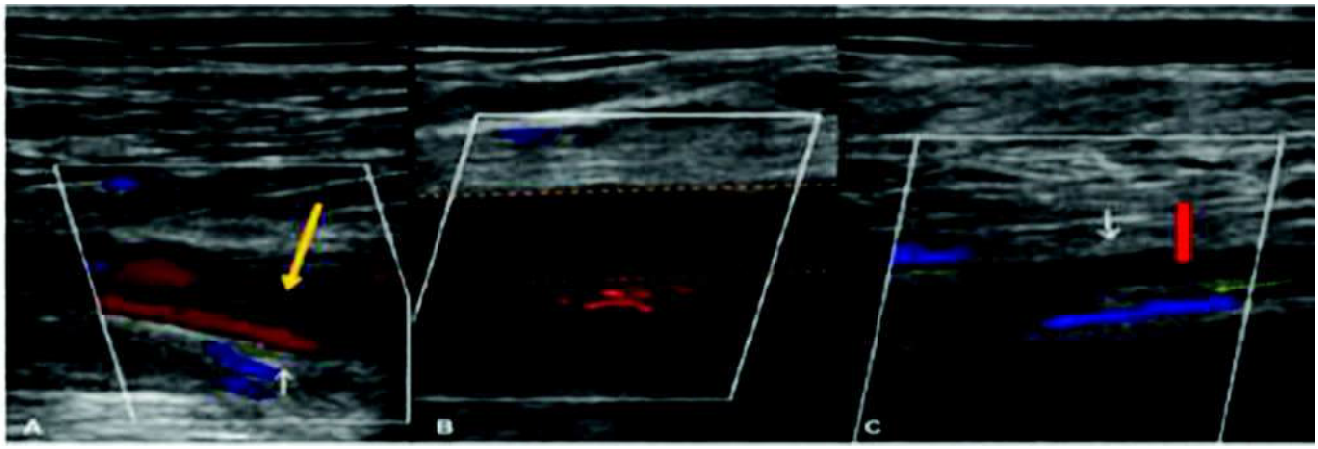

Fig. 1: *Doppler venous ultrasound demonstrating thromboses in the right (A) femoral, (B) popliteal, and (C) peroneal veins.*

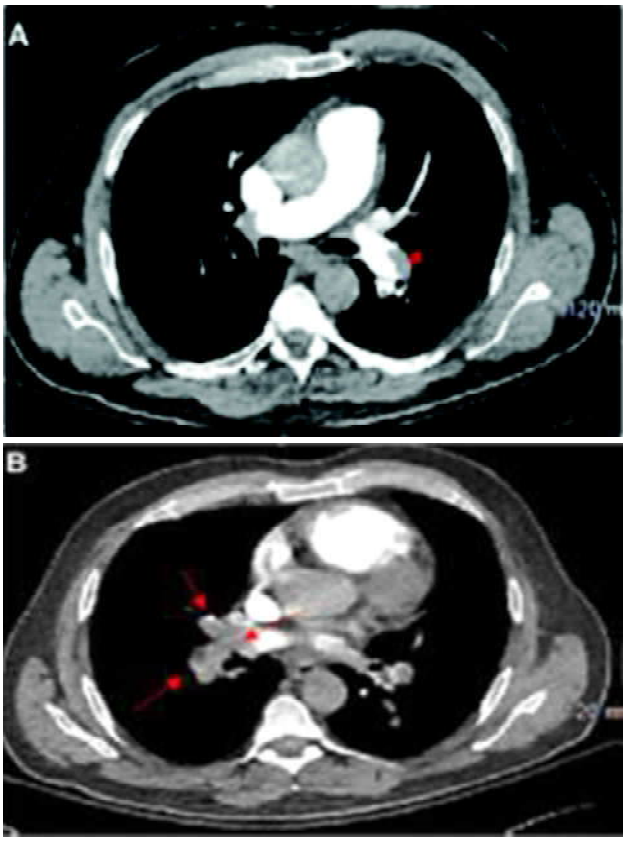

Fig. 2: *CT chest angiogram demonstrating pulmonary emboli in (A) the left main pulmonary artery, (B) the right main pulmonary artery and right lobar branches.*

Differential Diagnosis

The patient's abrupt and widespread arterial and venous thromboses prompted consideration of several critical differential diagnoses (Table II). Sepsis-induced coagulopathy was initially considered, as severe infection may trigger widespread endothelial injury and thrombosis. However, the patient exhibited no fever, haemodynamic instability, leucocytosis typical of infection, or positive cultures, making this diagnosis unlikely. Disseminated intravascular coagulation was also evaluated given the multiorgan ischaemia, but normal fibrinogen levels, preserved platelet count, and the absence of schistocytes argued against consumptive coagulopathy. Primary inherited thrombophilias such as factor V Leiden mutation, prothrombin gene mutation, or deficiencies of antithrombin III, protein C, or protein S were considered but deemed unlikely because they rarely cause simultaneous arterial and venous thrombosis in multiple territories and typically do not present with such explosive progression. Thrombotic microangiopathies, including thrombotic thrombocytopenic purpura and atypical Haemolytic uraemic syndrome, were considered but were inconsistent with the absence of haemolysis, normal platelet count, preserved renal function at presentation, and initially normal neurological findings. Vasculitis disorders such as ANCA-associated vasculitis and polyarteritis nodosa were also evaluated, but there was no evidence of systemic inflammation, characteristic vascular abnormalities, or supportive serologies. Ultimately, the simultaneous involvement of large and small vessels across multiple organ systems over a short time frame, combined with positive antiphospholipid antibodies, was most consistent with catastrophic antiphospholipid syndrome.

Table II: Differential diagnosis.

Differential Diagnosis	Points in Favor	Points Against
Sepsis-induced coagulopathy	– Presence of multiorgan thrombosis. – Elevated white blood cell count (neutrophilic leucocytosis).	– No fever or haemodynamic instability. – No positive cultures or evidence of infection – No systemic inflammation, which is usually seen in sepsis.
Disseminated Intravascular Coagulation (DIC)	– Multiorgan thromboses could indicate disseminated thrombosis. – The presence of thrombosis in multiple organs suggest DIC.	– Normal fibrinogen levels (usually decreased in DIC). – Normal platelet count, no schistocytes seen in peripheral smear. – Absence of haemolysis or significant anaemia.
Inherited Thrombophilias	– No prior history of thrombosis or autoimmune disease to suggest APS. – The abrupt nature of thrombosis affecting multiple vascular territories could suggest hypercoagulability	– Rarely causes both arterial and venous thrombosis simultaneously. – The explosive progression of thrombosis seen here is not typical of inherited thrombophilias – Inherited thrombophilias do not generally involve the spinal arteries, as in this case.
Thrombotic Microangiopathies (TTP or aHUS)	– Multiorgan involvement, including renal ischaemia, could suggest a microvascular pathology.	– No evidence of haemolysis, which is typical in TTP or aHUS. – Normal platelet count at presentation. – Preserved renal function and lack of neurological findings initially.

Vasculitis Disorders (ANCA-associated vasculitis, Polyarteritis nodosa)	– Systemic vascular involvement and thrombosis could mimic vasculitis. – Thrombosis in multiple vascular beds is common in vasculitis	– No evidence of systemic inflammation (e.g., elevated CRP or ESR). – No characteristic vascular abnormalities on imaging
Catastrophic Antiphospholipid Syndrome (CAPS)	– Widespread arterial and venous thrombosis across multiple organ systems. – Positive antiphospholipid antibodies (lupus anticoagulant, a2-GPI IgM).	– Initially, no prior history of autoimmune disease or thrombotic events – Diagnosis is rare, and initial symptoms overlap with common conditions. – It is more common in females and rarely seen in males
Myeloproliferative Neoplasm with neutrophilic leukocytosis	– Marked leukocytosis and thrombocytosis – Known association with thrombosis (arterial and venous) – Can cause hypercoagulable state	– No splenomegaly reported – No prior history suggestive of chronic MPN – Acute fulminant presentation atypical – No confirmatory tests (e.g., JAK2 mutation)
Hyperhomocysteinaemia	– Can predispose to arterial and venous thrombosis – May explain multisite thrombosis	– Usually causes chronic/recurrent thrombosis, not fulminant – No homocysteine levels reported – Does not typically cause such rapid progression
Occult systemic malignancy (paraneoplastic thrombosis)	– Malignancy is a known cause of hypercoagulability (Trousseau syndrome) – Can present with unexplained thrombosis	– No weight loss or constitutional symptoms – No mass lesions on imaging – Abrupt catastrophic course less typical – No prior cancer history

Management

Management focused on stabilizing the patient's respiratory status, controlling the thrombotic process, and addressing potential triggers. He was provided supplemental oxygen and immediately initiated on therapeutic low molecular weight heparin to prevent further thrombus formation. Because infection is a common precipitant of CAPS and could not initially be excluded, empirical broad-spectrum antibiotics were administered. IVIG was initiated early due to strong suspicion for CAPS, and antiplatelet therapy was added to help mitigate thrombus propagation. Despite these measures, the patient's clinical condition deteriorated, and the development of acute paraparesis prompted urgent imaging, confirming anterior spinal artery thrombosis and signalling ongoing catastrophic vascular involvement.

Advanced immunosuppressive and complement-targeted therapies, including cyclophosphamide (often used in SLE-associated CAPS) and eculizumab (which has demonstrated benefit in complement-mediated or refractory CAPS)[5], were considered. However, the rapid pace of his clinical decline precluded timely initiation.

Despite aggressive anticoagulation and initiation of immunomodulatory therapy, the patient's condition continued to deteriorate. On hospital day 6, he developed new-onset central chest pressure radiating to the left arm, accompanied by diaphoresis, nausea, and worsening dyspnoea. His clinical status declined rapidly, progressing to hypotension and tachyarrhythmia. A 12-lead ECG demonstrated acute ST-segment elevations in the anterior precordial leads, consistent with an extensive anterior wall ST-elevation myocardial infarction, a known but rare manifestation of CAPS-related coronary thrombosis. Concurrent bedside cardiac ultrasound revealed new severe left ventricular systolic dysfunction with regional wall motion abnormalities. Despite immediate initiation of advanced cardiac life support, including vasopressor support and antithrombotic escalation, he suffered recurrent ventricular arrhythmias culminating in pulseless ventricular tachycardia. Prolonged resuscitative efforts were unsuccessful, and the patient was pronounced deceased. The abrupt onset of myocardial ischaemia in the setting of active, uncontrolled systemic thrombosis underscores the fulminant nature of CAPS and highlights the challenge of preventing fatal macrovascular events even with timely and appropriate therapy.

Discussion

Catastrophic antiphospholipid syndrome represents the most severe form of APS, characterised by rapidly evolving, widespread thrombosis affecting multiple vascular beds within days. The underlying pathogenesis involves a self-amplifying cycle of endothelial activation, cytokine release, platelet aggregation, and complement activation, leading to uncontrolled microvascular and macrovascular thrombosis[5]. Although APS itself is relatively uncommon, CAPS occurs in fewer than 1% of APS patients[3] and carries a mortality rate of 30 - 50% despite early recognition and treatment[4]. Importantly, nearly one-third of CAPS cases occur *de novo*, without prior APS diagnosis or underlying autoimmune disease[6-8], contributing to diagnostic delays.

This patient's initial symptoms – including dyspnoea,

abdominal pain, and haematuria – were nonspecific and overlapped with far more common conditions such as pulmonary embolism, nephrolithiasis, or mesenteric ischemia from atherosclerotic disease. Such nonspecific presentations often obscure the diagnosis until multiorgan involvement becomes evident. The rare combination of deep vein thrombosis, pulmonary embolism, renal infarction, mesenteric artery thrombosis, and spinal artery thrombosis arising within days is highly unusual and strongly characteristic of CAPS.

Management of CAPS is centered around early initiation of triple therapy consisting of therapeutic anticoagulation, high-dose corticosteroids, and either IVIG or plasma exchange[9]. This multimodal approach aims to attenuate immune-mediated vascular injury, reduce circulating pathogenic antibodies, and halt propagation of thrombosis. In recent years, complement inhibitors such as eculizumab have emerged as promising adjunctive therapies in refractory cases[5]. However, the aggressiveness of CAPS in some patients, as demonstrated in this case, limits the opportunity to introduce additional therapies.

Epidemiologically, APS is more prevalent in women than men, and male patients presenting with fulminant or early-onset APS may represent a distinct phenotypic subgroup with potentially different risk profiles or disease trajectories[6-8]. The absence of identifiable triggers, the extraordinary tempo of disease progression, and the involvement of multiple major arterial and venous systems contributed to the poor prognosis in this case. Spinal arterial thrombosis, a rare manifestation even within APS, underscores the severity of vascular compromise occurring in catastrophic presentations.

In CAPS, myocardial infarction (MI) can occur due to coronary artery thrombosis, a result of the pro-thrombotic effects of antiphospholipid antibodies (aPLs). These antibodies promote endothelial dysfunction, platelet aggregation, and clot formation, not only in small vessels but also in larger arteries like the coronary vessels. While MI is less common in APS compared to venous thromboembolism, its occurrence in CAPS is significant, as it can rapidly lead to severe myocardial ischaemia and cardiac arrest. The hypercoagulable state in CAPS, coupled with complement activation, increases the risk of arterial thrombosis, including in the coronary circulation. This underscores the need for early detection and aggressive management, as coronary involvement can contribute to high mortality in these patients[10].

This case is exceptional due to the combination of male sex, absence of prior autoimmune disease, absence of identifiable precipitating factors, and the rapidly progressive nature of multisystem arterial and venous thrombosis that developed within a matter of days. The sudden onset of anterior spinal artery thrombosis, along with the subsequent fatal myocardial infarction, underscores the fulminant nature of CAPS and its capacity to rapidly compromise multiple vascular territories. These unique and severe manifestations highlight the diagnostic challenges in recognizing CAPS early, particularly when presented without a prior history of autoimmune disease or typical risk factors. Furthermore, this case emphasizes the clinical variability of CAPS and the therapeutic difficulties in managing such an aggressive and rapidly progressing condition, contributing valuable insights into the complexity of diagnosing and treating this rare but devastating syndrome.

Conclusion

Catastrophic antiphospholipid syndrome is a rare but devastating thrombotic emergency that requires high clinical suspicion, particularly when patients present with rapidly evolving thromboses across multiple vascular beds. This case highlights the importance of early recognition of CAPS even in patients without pre-existing autoimmune disease, conventional risk factors, or identifiable triggers. The patient's fulminant presentation despite timely anticoagulation and immunomodulatory therapy illustrates the limitations of current treatment strategies and the need for improved diagnostic tools and more effective targeted therapies.

References

1. Miyakis S, Lockshin MD, Atsumi T *et al*. International consensus statement on an update of the classification criteria for APS. *J Thromb Haemost* 2006; 4: 295-306.
2. Asherson RA. The catastrophic antiphospholipid syndrome. *J Rheumatol* 1992; 19 (4): 508-12.
3. Cervera R, Bucciarelli S, Plasín MA *et al*. Catastrophic APS (CAPS): Descriptive analysis of 250 cases. *Ann Rheum Dis* 2009; 68: 446-52.
4. Cervera R, Rodríguez-Pintó I, Colafrancesco S *et al*. 14th International CAPS Registry analysis. *Autoimmun Rev* 2015; 14 (2): 105-09.
5. Bitsadze V, Yakubova F, Khizroeva J *et al*. Catastrophic APS: mechanisms and management. *Int J Mol Sci* 2024; 25: 668.
6. Duarte-García A, Pham MM, Crowson CS *et al*. Epidemiology of APS: population-based study. *Arthritis Rheumatol* 2019; 71: 1545-52.
7. Khellaf M *et al*. Incidence and prevalence of antiphospholipid syndrome: a systematic review. *BMJ Open* 2024; 14: e084563.
8. Dabit JY *et al*. Epidemiology of antiphospholipid syndrome in the general population. *Arthritis Rheumatol* 2022; 74: 1883-90.
9. Asherson RA, Cervera R, de Groot PG *et al*. Catastrophic APS: updated recommendations for treatment (CAPS Task Force). *Lupus* 2003; 12 (7): 530-4.
10. Bicocca M *et al*. Catastrophic antiphospholipid syndrome and myocardial infarction: Pathophysiology and management challenges. *J Thromb Haemost* 2020; 18 (1): 34-42.