JEIMP

The Journal of European Internal Medicine Professionals

Letter to Editor

A Shocking Cause of Kidney Failure: Antiphospholipid Antibody Syndrome

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DOI:10.5281/zenodo.18462557

Submitted at: 18.11.2025, **Accepted at:** 26.01.2026, **Published at:** 01.02.2026

Dear Editor,

Acute kidney injury (AKI), often originating from vasculitis or thromboembolism, rarely results from antiphospholipid syndrome (APS) manifesting as thrombotic microangiopathy. Although catastrophic APS occurs in <1% of cases, it remains a frequently overlooked cause of AKI.

A 64-year-old male smoker with chronic kidney disease presented with a one-week history of diarrhea, dyspnea, and weakness. Physical examination revealed hypertension (150/104 mmHg), tachycardia (142 bpm), icterus, and basal rales. Laboratory results (Table 1) showed acute kidney injury (creatinine 5.59 mg/dL, eGFR 10 mL/min), hyperkalemia (6.2 mmol/L), thrombocytopenia (48,000/µL), and liver dysfunction (AST 656 U/L, ALT 826 U/L, total bilirubin 6.62 mg/dL). Peripheral smear identified schistocytes (3-4/HPF). CT imaging suggested a main pulmonary artery thrombus, massive pleural effusion, and pulmonary infarcts.

On Day 1, the patient was started on enoxaparin and hemodialysis (HD). Due to suspected thrombotic microangiopathy, plasmapheresis was initiated immediately post-HD. On Day 2, high-dose pulse steroid therapy was added. On Day 5, livedo racemosa developed (Figure 1). Skin biopsy results on Day 7 revealed fibrinoid thrombi in superficial and mid-dermis vessels, confirming systemic microvascular involvement.

The clinical presentation, along with negative ADAMTS13 activity and positive antiphospholipid antibodies (lupus anticoagulant, anti-beta2-glycoprotein IgA), fulfilled the criteria for 'probable Catastrophic Antiphospholipid Syndrome (CAPS)'. Despite the immediate initiation of comprehensive 'triple therapy' (comprising therapeutic anticoagulation, high-dose glucocorticoids, and plasma exchange/intravenous immunoglobulin), the patient's condition deteriorated due to widespread thromboembolic events. The patient succumbed to cardiopulmonary arrest in the intensive care unit.

APS is an autoimmune disorder leading to a prothrombotic state (thrombophilia) (1).In women of childbearing age, it frequently presents with clinical manifestations such as recurrent miscarriages, early pregnancy loss, and preeclampsia (2).

APS also constitutes a significant portion of thromboembolic diseases. The prevalence of antiphospholipid antibodies was found to be 10% among patients with deep vein thrombosis (DVT) and 14% in individuals who had experienced a stroke. In cases of obstetric morbidity, the prevalence of these antibodies ranged from 6% to 9% (3).

The commonly observed clinical features of APS include thrombocytopenia, cardiac valve disease, transient ischemic attack, and livedo racemosa (4). A rare, life-threatening manifestation of APS is CAPS, which presents with widespread thrombosis and multiorgan failure (5).CAPS is an autoimmune disease characterized by symptoms developing in less than one week, involving three or more organs, and histologically confirmed small vessel occlusion in at least one organ. It is also defined by the presence of aPL antibodies, documented as positive at least twice, with a minimum of 12 weeks between tests (6).In our case, a second antibody test after 12 weeks could not be performed. Consequently, this case is classified as 'probable CAPS' based on the preliminary positive aPL titers and the severity of the clinical manifestation. This highlights a significant diagnostic pitfall in clinical practice: the high

early mortality rate often prevents patients from meeting formal criteria, necessitating rapid clinical judgment and aggressive empirical therapy before definitive classification can be achieved.

The clinical presentation of thrombotic microangiopathy (TMA) in this case necessitated a rigorous differential diagnosis. Thrombotic thrombocytopenic purpura (TTP) was excluded due to ADAMTS13 activity levels being within the normal range (>10%). Although the patient presented with severe acute kidney injury, the rapid multi-organ involvement and the presence of high-titer antiphospholipid antibodies shifted the diagnosis away from atypical hemolytic uremic syndrome (aHUS). Furthermore, while the patient exhibited systemic inflammatory features, the absence of overt consumption coagulopathy (normal fibrinogen levels and absence of significant PT/aPTT prolongation) helped differentiate this condition from primary sepsis-induced disseminated intravascular coagulation (DIC). The hallmark finding of livedo racemosa, combined with multi-visceral thrombosis, strongly pointed toward CAPS as the primary etiology of the TMA.

In addition to conditions like stroke and kidney infarction, CAPS can also lead to macrovascular involvement and multi-organ thrombosis, distinguishing it from APS. The disease process can be triggered by complement activation, which in turn can be stimulated by factors such as infection, inflammation, surgery, or pregnancy (7, 8). The standard approach, often referred to as "triple therapy," includes anticoagulation, glucocorticoids, and either therapeutic plasma exchange (TPE) or intravenous immunoglobulin (IVIG). As demonstrated in our case management, despite applying the triple therapy approach, the progressive nature of the disease necessitated considering rituximab, a treatment option for refractory cases. Rituximab may be the preferred choice in select patients with severe thrombosis and thrombocytopenia (9). Given the pivotal role of complement activation in the pathogenesis, eculizumab may be preferred over rituximab as an adjunctive therapeutic option to the core treatment regimen.

In conclusion, CAPS is an autoimmune disease with high mortality that can lead to progressive multiorgan failure, requiring careful and experienced management. Livedo racemosa is a critical cutaneous marker that should alert clinicians to underlying systemic microvascular thrombosis and the potential onset of CAPS. In fulminant cases, early mortality may preclude the 12-week confirmatory testing for antiphospholipid antibodies. In such scenarios, the diagnosis of "probable CAPS" should be sufficient to initiate aggressive multimodal therapy. Severe acute kidney injury in the setting of multisystem failure and thrombotic microangiopathy requires immediate differentiation from TTP and aHUS to avoid delays in starting life-saving plasmapheresis and anticoagulation. A high index of suspicion and early intervention with the triple therapy is the cornerstone of managing a "thrombotic storm," even when all definitive criteria are not yet met. Despite its rarity, the fatal outcome for this patient underscores the critical importance of effective and rapid treatment initiation based on a solid understanding of the diagnostic process.

DECLARATIONS

Ethics committee approval: None,
Consent was obtained per the Declaration of Helsinki.
Financial Disclosure: The author declare that they received no financial support for the research, authorship, and/or publication of this article.

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