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Case Report

A Case of Catastrophic Antiphospholipid Syndrome Presenting as Pulmonary Embolism and Renal Thrombosis: A Case Report and Literature Review

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Abstract

Catastrophic antiphospholipid syndrome (CAPS), also known as Asherson's syndrome, is a rare and life-threatening condition that represents the most severe clinical manifestation of antiphospholipid syndrome. Due to its rapid and aggressive course, CAPS is characterized by the development of widespread micro- and macrothrombosis, which results in multi-organ ischemia and failure. Antiphospholipid syndrome, an autoimmune disorder, is characterized by thrombotic and/or obstetric events, accompanied by persistent antiphospholipid antibodies. The case of a 36-year-old woman was described, who initially presented with low-effort dyspnea and abdominal pain, with subsequent imaging revealing pulmonary embolism and aortic thrombosis, alongside acute ischemic cerebellar infarcts. The eventual confirmation of a positive lupus anticoagulant solidified the diagnosis of CAPS. This case underscores the diagnostic challenge posed by CAPS, particularly when presenting with prominent pulmonary embolism, and highlights the critical need for prompt recognition and multidisciplinary management. Furthermore, the patient's positive outcome demonstrates the potential for recovery even in such severe presentations.

Keywords Case report, catastrophic antiphospholipid syndrome, infarct, multidisciplinary, thrombosis

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Introduction

Antiphospholipid syndrome (APS) is a systemic autoimmune disorder that manifests through arterial and/or venous thrombosis and pregnancy loss in individuals who test positive for antiphospholipid antibodies (aPL), typically lupus anticoagulant (LA), anticardiolipin (aCL), and anti-beta-2 glycoprotein (anti-β2GPI) autoantibodies.[1,2]

Catastrophic antiphospholipid syndrome (CAPS), also known as Asherson's syndrome, is the rare, accelerated, and potentially fatal extreme of APS. It is marked by the rapid development of thrombosis that can compromise the arterial, venous, or microvascular systems.[1,2] Thrombotic microangiopathy is a prominent feature in CAPS; however, it can also occur in classical APS, highlighting that this feature is not exclusive to the catastrophic form but is part of the broader APS spectrum. Catastrophic antiphospholipid syndrome can be the initial presentation of APS or occur in those with a pre-existing diagnosis. Fortunately, it affects less than 1% of the APS population; however, mortality remains high at 30%-50% despite treatment.[1-4] Its diagnosis is based on Asherson's preliminary criteria, which include: (a) involvement of at least 3 organs, (b) immediate or rapid progression (less than a week), (c) histopathological evidence of small vessel occlusion, and (d) laboratory confirmation of aPL.[5] Given its high mortality, early recognition and aggressive management are paramount for improving patient outcomes.

Case Presentation

A 36-year-old woman presented to the emergency department due to progressively worsening dyspnea over 48 hours. She also described severe abdominal pain in the mesogastric area radiating to the left iliac fossa and a painful headache radiating to her forehead, nose, and chin, which intensified upon movement. She was diagnosed with hypertension 5 years prior and was treated with enalapril. She experienced mild preeclampsia during her first pregnancy, as well as an episode of deep vein thrombosis in her right lower extremity during her second pregnancy. There was no pertinent medical family history.

Upon physical examination, she was afebrile with a pulse of 90 beats per minute, blood pressure of 150/100 mmHg, and SpO2 of 88% on 36% FiO2 with evident signs of respiratory distress. She was conscious and oriented. Laboratory values upon presentation demonstrated positive LA and elevated aCL IgG antibody, 65 GPL U/mL (reference range < 20 GPL U/mL) and anti-β2GPI IgG 22 U/mL (normal < 20 U/mL). The early assessment of antiphospholipid antibodies, including LA and aCL, reflected the clinical suspicion for underlying APS given the patient's prior history of DVT and preeclampsia.

As part of the diagnostic assessment, a pulmonary and renal computed tomography (CT) was conducted, revealing substantial infarction in the left kidney, along with partial thrombosis of the celiac trunk (Figure 1) and findings consistent with bilateral pulmonary embolism (Figure 2).

Based on the clinical presentation, known medical history, and imaging characteristics, the patient was diagnosed with APS, and suspicion arose regarding CAPS once differential diagnoses were ruled out. Given the multi-organ involvement, other thrombotic microangiopathies such as TTP and HUS were considered. However, the absence of thrombocytopenia (platelet count of 252 × 109/L) and hemolytic anemia (normal LDH and bilirubin levels) made TTP and HUS less likely. DIC was less probable as initial coagulation studies revealed a normal coagulation profile, which did not suggest the consumptive coagulopathy typically seen in DIC. The lack of prior exposure to heparin made HIT unlikely, further supported by the stable platelet count.

Though therapeutic plasma exchange (TPE) was considered a potential intervention, particularly considering the confirmed renal involvement, the institution lacked the resources to conduct this procedure. Consultation with nephrology and vascular surgery led to the initiation of dual immunomodulatory therapy, consisting of intravenous methylprednisolone 80 mg/day (equivalent to 100 mg/day of prednisone) and intravenous immunoglobulin 0.4 g/kg/day for 5 days. On the same day of her admission, the patient underwent a radical open nephrectomy on her left kidney. This decision was made due to the finding of complete renal necrosis, and importantly, to remove a potential source of infection in the setting of extensive tissue infarction, which could further complicate her critical condition.

Notably, the post-operative period was complicated by the development of neurological symptoms. She reported a brief episode of amaurosis. This was followed by a cranial nerve evaluation revealing findings consistent with right homonymous hemianopsia. Neuroimaging with cerebral CT then demonstrated an ischemic cerebrovascular event involving the left occipital lobe (Figure 3), anatomically correlating with the visual deficit described. Additionally, hypodensities were noted in the territory of the right cerebellum, suggesting multiple vascular territories affected. These conclusive findings, in conjunction with the pre-operative elevated aPL, strongly supported the diagnosis of CAPS.

Given this diagnosis and the patient's rapidly deteriorating clinical condition, she was admitted to the intensive care unit (ICU) for close monitoring and blood pressure control. Subsequently, she developed multi-organ failure, necessitating hemodynamic support and mechanical ventilation.

During her ICU stay, there was a satisfactory progression, with gradual resolution of all organic failures. Vasopressor support was titrated, and sedation and analgesia were gradually discontinued. The patient remained stable in the ICU while being treated for *Burkholderia cepacia* bacteremia with a 10-day course of Trimethoprim-Sulfamethoxazole, to which she responded favorably. Following her ICU stay, after achieving the target INR (2.0-3.0), she was discharged on Warfarin. On a follow-up visit 3 months post-initial testing, the aPL remained positive, and her INR was measured at 2.3; the patient remained asymptomatic.

Discussion

Catastrophic antiphospholipid syndrome is a rare and often rapidly progressive condition, as exemplified by the 36-year-old female patient who presented with a dramatic constellation of pulmonary embolism, renal infarction, and subsequent neurological involvement. The challenges in systematically studying CAPS due to its rarity led to the establishment of "The CAPS Registry," which provides information on demographics, precipitating factors, and clinical and laboratory features from over 700 patients. In this analysis, most patients (70%) were female, with a mean age of 38 years, aligning with the patients' demographics. Roughly half of all patients in the registry suffered from SLE or presented with a manifestation of it. Clinical manifestations varied by organ, with the renal system being the most commonly

Main Points

- Catastrophic antiphospholipid syndrome (CAPS) is a rare and severe variant of antiphospholipid syndrome that is marked by the development of microthrombotic events resulting in multi-organ failure.
- A 36-year-old woman with a history of hypertension and preeclampsia presented with CAPS, with a rapidly deteriorating condition.
- The basis of CAPS treatment consists of therapeutic anticoagulation, high doses of corticosteroids, and either plasmapheresis or intravenous immunoglobulin.
- This report highlights the critical need for vigilance in identifying this condition, particularly in patients who present with thrombotic events in multiple organ systems, as well as the potential for successful outcomes with prompt and multidisciplinary intervention.

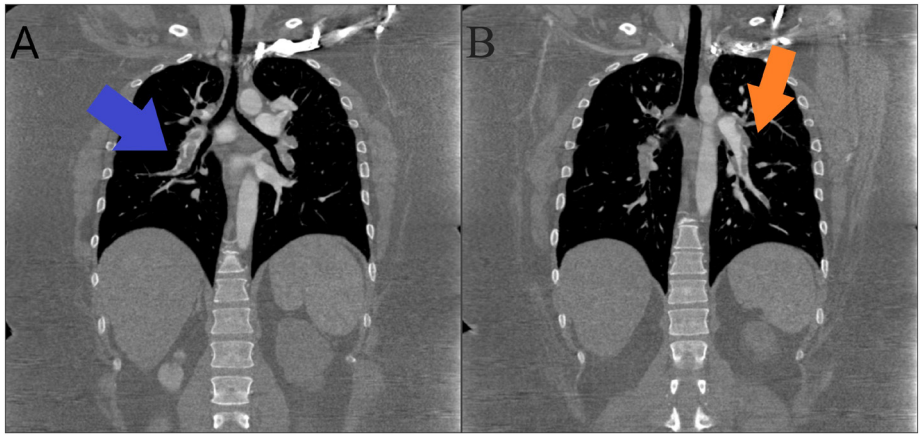

Figure 1. Contrast-enhanced computed tomography of the chest. (A) Thrombosis of the right pulmonary artery (blue arrow). (B) Thrombosis of the left pulmonary artery (orange arrow).

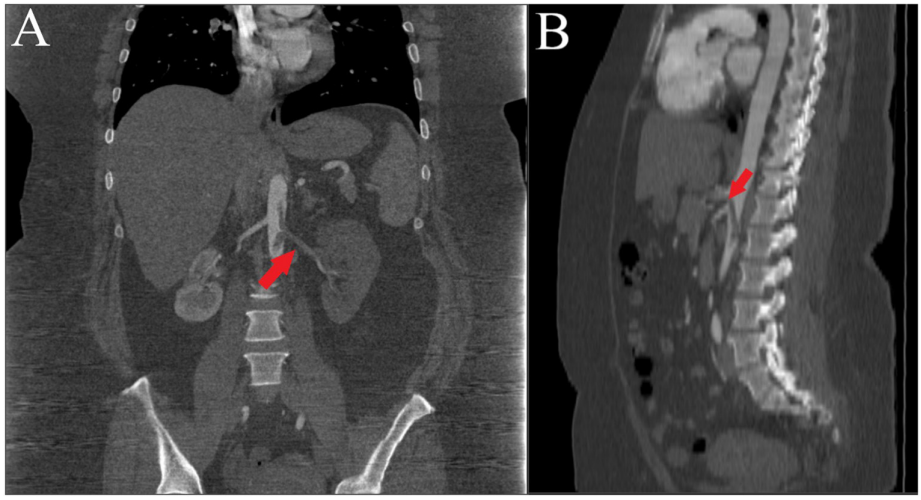

Figure 2. Contrast-enhanced computed tomography of the abdomen. (A) Thrombus from the aorta extending into the left renal artery with secondary extensive renal infarct (red arrow). (B) Partial occlusive thrombosis of the aorta extending into the celiac artery (red arrow).

affected (73%), followed by pulmonary (60%), nervous system (56%), heart (50%), skin (47%), and liver (39%). Thrombocytopenia was the leading laboratory manifestation (67%); other findings included thrombotic microangiopathy, disseminated intravascular coagulation, and elevated LA. While the patient's presentation involved prominent renal, pulmonary, and neurological features consistent with these registry findings, she did not exhibit thrombocytopenia initially, highlighting the variability in CAPS presentation.[6,7]

This phenomenon is often triggered by specific factors, with a known precipitant identified in 65% of cases in the CAPS Registry. Infections are the most common trigger, present in 49% of cases, followed by surgery (17%), malignancy (especially in older individuals, 16%), and pregnancy-related complications (8%).[6]

Current literature supports a 3-step approach to managing CAPS: initially, supportive measures; subsequently, management of any recognized triggers; and lastly, specific treatment. Treatment guidelines emphasize the use of anticoagulants, corticosteroids, and TPE or intravenous immunoglobulin (IVIG), often termed triple therapy. Initial anticoagulation therapy with unfractionated heparin or LMWH represents the pillar of CAPS management. Subsequently, therapy with Warfarin should be started in clinically stable patients, with an INR target of 2.0-3.0 for most patients. However, a higher target (3.0-4.0) has been proposed as an alternative in high-risk aPL patients with arterial thrombosis. DOACS are generally contraindicated in CAPS. High doses of glucocorticoids, used to mitigate the excessive inflammatory response, are fundamental for the treatment of CAPS. Typically, methylprednisolone is started at 1-2 mg/kg/day for at least 3 days, followed by a transition to oral or parenteral therapy with prednisone once clinical improvement is achieved. Alternatively, pulse therapy with methylprednisolone may be employed. Data from the CAPS Registry indicate a roughly equal use of both regimens. The mechanisms by which TPE acts in CAPS are unclear, but the removal of aPL has been postulated as the possible mechanism. IVIG diminishes complement activation, further diminishing the inflammatory response. The best survival rate (70%) has been observed with triple therapy in comparison to mono and dual therapy, which only achieved a survival rate of 59%.[3-9]

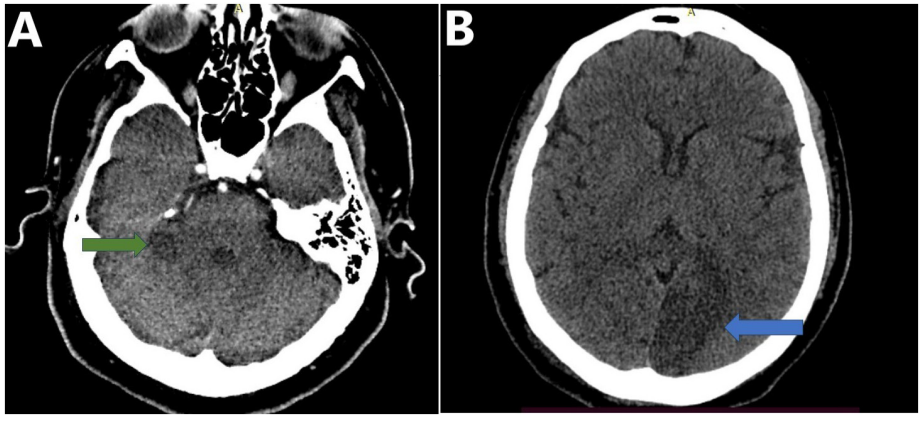

Figure 3. Computed tomography of the brain. (A) Ischemic stroke affecting the right cerebellum (green arrow). (B) Ischemic stroke compromising the left occipital lobe (blue arrow).

New evidence suggests the utility of biological agents, such as Rituximab, a monoclonal antibody against CD20, which has shown promise against refractory and relapsing CAPS cases. Eculizumab, a C5 inhibitor, has been used in severe therapy-resistant CAPS. Ongoing therapies continue to explore different therapeutic targets to improve outcomes, but given the complexity of this condition, a multidisciplinary approach is fundamental.[9,10]

Despite treatment optimization, CAPS mortality remains considerable (33%, higher at 48% in SLE patients). Certain phenomena, such as older age (>36 years), pulmonary and renal compromise, and positive antinuclear antibody titer, have been associated with worse outcomes. The main causes of mortality in the CAPS Registry are cerebral (19.5%), cardiac (14.1%), infections (14.1%), pulmonary (7.1%), and abdominal (4.5%) involvement. The patient, despite being within the older age range defined by the registry for worse outcomes and having pulmonary and renal involvement, had a positive outcome, highlighting the potential for recovery with timely and comprehensive care.[11]

Conclusion

This case underscores the critical need to include CAPS in the differential diagnosis of thrombosis presenting as multi-organ failure. Patients with a significant history of unexplained thrombosis or recurrent pregnancy loss should be tested for aPL. Notably, young patients with a history of vascular events are typically at higher risk for an APS diagnosis and should be evaluated accordingly. Despite the absence of TPE, the patient achieved a favorable outcome with prompt and multidisciplinary intervention, highlighting the potential for recovery even in severe presentations of CAPS. A multidisciplinary team encompassing critical care, nephrology, hematology, and rheumatology consultants remains essential to achieve the best outcomes.

Data Availability Statement: The data that support the findings of this study are available on request from the corresponding author.

Informed Consent: Written informed consent was obtained from the patient who agreed to take part in the study.

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References

1. Rodziewicz M, D'Cruz DP. An update on the management of antiphospholipid syndrome. *Ther Adv Musculoskelet Dis*. 2020;12:1759720X20910855. [CrossRef]
2. Knight JS, Branch DW, Ortel TL. Antiphospholipid syndrome: advances in diagnosis, pathogenesis, and management. *BMJ*. 2023;380:e069717. [CrossRef]
3. Siniscalchi C, Basaglia M, Riva M, et al. Catastrophic antiphospholipid syndrome: a review. *Immuno*. 2024;4(1):1-13. [CrossRef]
4. Jacobs L, Wauters N, Lablad Y, Morelle J, Taghavi M. Diagnosis and management of catastrophic antiphospholipid syndrome and the potential impact of the 2023 ACR/EULAR antiphospholipid syndrome classification criteria. *Antibodies (Basel)*. 2024;13(1):21. [CrossRef]
5. Asherson RA, Cervera R, de Groot PG, et al. Catastrophic antiphospholipid syndrome: international consensus statement on classification criteria and treatment guidelines. *Lupus*. 2003;12(7):530-534. [CrossRef]
6. Rodríguez-Pintó I, Moitinho M, Santacreu I, et al. CAPS Registry Project Group (European Forum on Antiphospholipid Antibodies). Catastrophic antiphospholipid syndrome (CAPS): descriptive analysis of 500 patients from the International CAPS Registry. *Autoimmun Rev*. 2016;15(12):1120-1124. [CrossRef]
7. Rodriguez-Pintó I, Espinosa G, Cervera R. What we know and what we don't know about catastrophic antiphospholipid syndrome. *Rheumatol (Oxf Engl)*. 2024;63(SI):SI46-SI53. [CrossRef]
8. Capecchi M, Abbattista M, Ciavarella A, Uhr M, Novembrino C, Martinelli I. Anticoagulant therapy in patients with antiphospholipid syndrome. *J Clin Med*. 2022;11(23):6984. [CrossRef]
9. Cervera R, Rodríguez-Pintó I, Espinosa G. The diagnosis and clinical management of the catastrophic antiphospholipid syndrome: a comprehensive review. *J Autoimmun*. 2018;92:1-11. [CrossRef]
10. Parepalli A, Sarode R, Kumar S, Nelakuditi M, Kumar MJ. Antiphospholipid syndrome and catastrophic antiphospholipid syndrome: a comprehensive review of pathogenesis, clinical features, and management strategies. *Cureus*. 2024;16(8):e66555. [CrossRef]
11. Bucciarelli S, Espinosa G, Cervera R, et al. European Forum on Antiphospholipid Antibodies. Mortality in the catastrophic antiphospholipid syndrome: causes of death and prognostic factors in a series of 250 patients. *Arthritis Rheum*. 2006;54(8):2568-2576. [CrossRef]