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Case Report

Aorto to alveoli: catastrophic antiphospholipid syndrome masquerading as multisystem failure

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ABSTRACT

Catastrophic Antiphospholipid Syndrome (CAPS) is a rare, fulminant variant of antiphospholipid syndrome, characterized by rapid multi-organ involvement due to widespread thrombosis. We report a 65-year-old male, with prior comorbidities such as diabetes, hypertension, dyslipidemia, cerebrovascular accident and coronary artery disease who presented with complete thrombosis of the distal abdominal aorta and common iliac vessels. During hospital stay, he developed acute respiratory distress, right-sided haemothorax with massive pleural effusion, diffuse alveolar haemorrhage, acute kidney injury with microscopic haematuria, and neurological deterioration requiring mechanical ventilation. Initial autoimmune workup was negative for ANA, ANCA, and anti-GBM antibodies. However, β2-glycoprotein IgG was strongly positive, and anticardiolipin antibody was borderline positive, raising suspicion of CAPS. Given the constellation of multiorgan dysfunction with diffuse alveolar haemorrhage and vascular thrombosis, a diagnosis of catastrophic APS was considered. The patient underwent aggressive multimodal therapy including pulse corticosteroids, anticoagulation, renal replacement therapy, immunomodulators, and ventilatory support. Plasma exchange/IVIG was planned in case of further worsening. With multidisciplinary management and intensive supportive care, the patient showed gradual recovery, and was discharged in a stable condition. This case highlights the diagnostic challenge of CAPS, which often masquerades as vasculitis, sepsis, or other autoimmune diseases. It also underlines the management hurdles, requiring timely recognition, multidisciplinary coordination, and combined immunosuppression and anticoagulation. Despite its rarity, CAPS should be considered in patients with rapidly progressive multiorgan thrombosis, as early diagnosis and intervention can be life-saving.

Keywords: Catastrophic APS, Diffuse alveolar haemorrhage, Haemothorax, Multiorgan thrombosis, β2-glycoprotein

INTRODUCTION

Antiphospholipid Syndrome (APS) is an autoimmune disorder characterized by recurrent arterial or venous thrombosis and pregnancy morbidity, associated with persistent antiphospholipid antibodies.[1] Catastrophic Antiphospholipid Syndrome (CAPS), also called Asherson's syndrome, is a rare but life-threatening variant, occurring in less than 1% of APS cases.[1] It is defined by rapidly progressive, widespread small-vessel thrombosis leading to multiorgan dysfunction within days.[1] Despite the advancement of aggressive multimodal therapies, mortality remains high, ranging from 40%-50%.[2] The disease frequently presents as a clinical masquerade, mimicking conditions such as systemic sepsis, vasculitis, or thrombotic microangiopathies.[2,3] This often results in critical diagnostic delays, particularly when standard autoimmune workups— including ANA, ANCA, and anti-

GBM antibodies return negative results.[3,5] One of the most rare and fatal complications is diffuse alveolar haemorrhage (DAH), which creates a profound therapeutic dilemma regarding the safe initiation of the anticoagulation.[5-6]

CASE REPORT

A 65-year-old male with past history of diabetes mellitus, hypertension, dyslipidemia, cerebrovascular accident and coronary artery disease was admitted under cardiology for evaluation of complete thrombosis of the distal abdominal aorta and bilateral common iliac arteries.

Clinical course

Upon admission, the patient was conscious and oriented, with stable vital signs. His temperature was 98°F, and there were no signs of pallor, cyanosis, clubbing, oedema, or lymphadenopathy. A physical examination of his respiratory system revealed decreased air entry in the right lower zone, accompanied by bilateral crepitations. The initial laboratory workup revealed significant hematologic and renal abnormalities (Table 1).

Table 1: Summary of admission laboratory investigations.

Investigation	Patient value	Normal / reference range
Haemoglobin	8.8 g/dl	13.5-17.5 g/dl
Total leukocyte count (TLC)	13,000/µl (n:82%, l:16%)	4,000-11,000/µl
Platelet count	1.0 lakh/µl	1.5-4.5 lakh/µl
Serum urea/ creatinine	68 mg/dl / 1.6 mg/dl	15-45/ 0.7-1.3 mg/dl
Serum procalcitonin	0.12 ng/ml	<0.5 ng/ml
Urinalysis (RBC)	30-40 /hpf	0-2/hpf
B2-glycoprotein I (IgG)	160	<20 (negative)
Anticardiolipin antibody (IgG)	18	<12 (negative)
ANA/ANCA/ anti-GBM	Negative	Negative

Chest X-ray revealed a significant right-sided moderate pleural effusion (Figure 1). The initial working diagnosis was an exudative pleural effusion, and a diagnostic and therapeutic pleural tapping was performed.

This is where the first significant diagnostic hurdle was encountered. The pleural tapping revealed the fluid to be haemorrhagic, which immediately shifted the differential diagnosis away from a simple transudative or typical exudative effusion. Following this finding, an intercostal drainage (ICD) tube was inserted, and a repeat CT thorax confirmed a massive right-sided haemothorax (Figure 2).

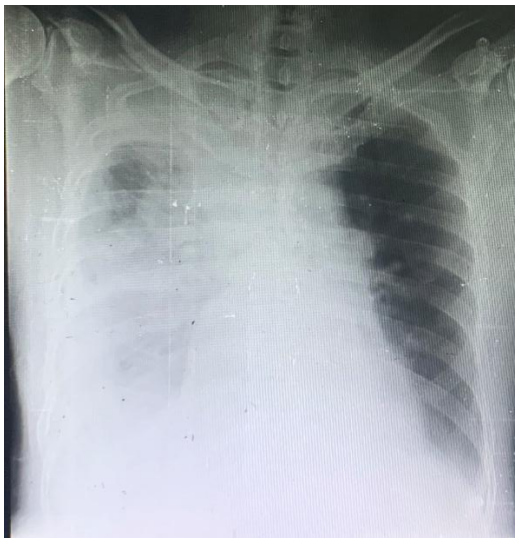

Figure 1: Chest X ray shows significant opacification of right hemithorax.

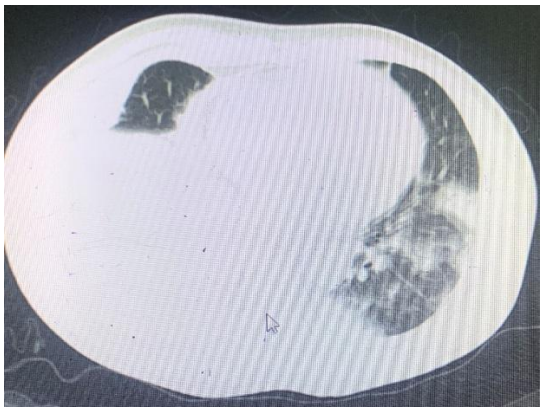

Figure 2: Axial CT thorax showing massive right sided haemothorax and pulmonary compression.

The patient's condition continued to deteriorate, with worsening respiratory status disproportionate to the haemothorax alone, leading to intubation and mechanical ventilation. At this critical juncture, a bronchoscopy with bronchoalveolar lavage (BAL) was performed. The lavage returned progressively more haemorrhagic fluid, and cytological/biopsy examination definitively identified diffuse alveolar haemorrhage (DAH) (Figures 3 and 4).

Concurrently, the patient developed deranged renal function along with microscopic haematuria. Initial autoimmune workup, including ANA, ANCA, and anti-GBM antibodies, was negative. The combination of large-vessel thrombosis, haemorrhagic pulmonary manifestations, and acute renal failure created a diagnostic conundrum that mimicked several critical conditions, including vasculitis or sepsis. The breakthrough in diagnosis came with the results of the specific

antiphospholipid antibody panel, which showed a strongly positive β2-glycoprotein IgG and a borderline positive anticardiolipin antibody. This finding, combined with the multisystem thrombotic and haemorrhagic events, led to the diagnosis of Catastrophic Antiphospholipid Syndrome (CAPS).

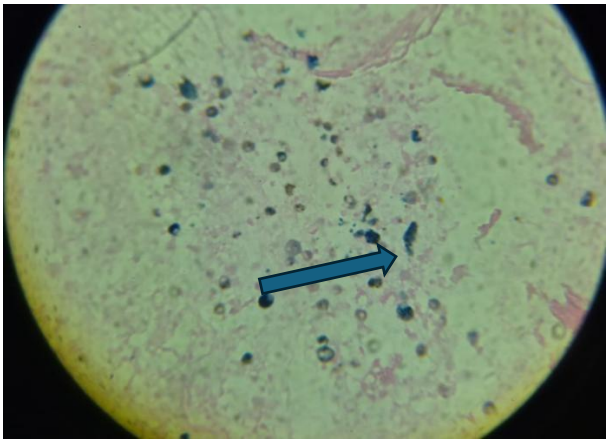

Figure 3: Prussian blue stain showing hemosiderin-laden macrophages. BAL cytology demonstrating extensive intra-alveolar haemorrhage with numerous hemosiderin-laden macrophages.

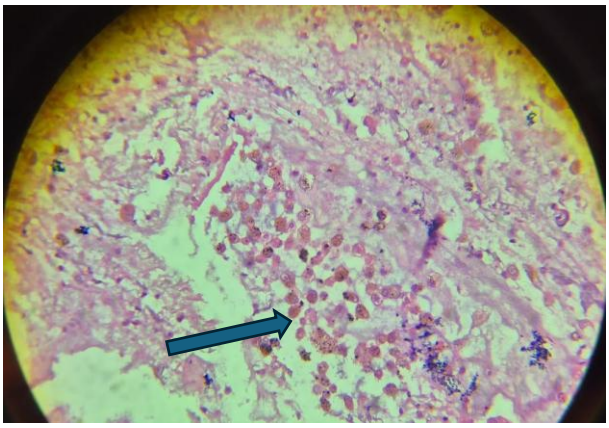

Figure 4: Haematoxylin and eosin stain showing hemosiderin-laden macrophages. Lung biopsy section stained with haematoxylin and eosin (H and E) showing extensive intra-alveolar haemorrhage and hemosiderin-laden macrophages.

The management of this case was a continuous struggle between competing therapeutic goals. The patient's life-threatening pulmonary haemorrhage made the use of therapeutic anticoagulation to address the extensive thrombosis particularly risky. The decision to initiate unfractionated heparin, despite the ongoing bleeding, was a high-stakes clinical judgment that required careful risk-benefit analysis. Furthermore, the patient's lack of response to initial oral steroids and subsequent improvement only after high-dose pulse steroids highlights the need for aggressive immunosuppression in this fulminant condition. With this comprehensive and multidisciplinary approach, the patient showed a gradual recovery. He was eventually extubated, continued with physiotherapy, and exhibited improving laboratory trends. At the time of discharge, he was conscious, oriented, obeying commands, tolerating oral feeds, and had stable vital signs. The successful outcome is a testament to the crucial role of a coordinated, multidisciplinary team in managing the complex and often fatal manifestations of CAPS.

Overall treatment given

He was managed with anticoagulation using unfractionated heparin as per the RASCHKE protocol. Immunosuppressive therapy was initiated with intravenous methylprednisolone 500 mg once daily for 3 days, followed by intravenous dexamethasone 10 mg per day, after which he was transitioned to oral steroids once his condition stabilized. Renal replacement therapy was provided in the form of three sessions of haemodialysis to address acute kidney injury and deranged renal function. In addition, respiratory support was escalated to mechanical ventilation following the diagnosis of diffuse alveolar haemorrhage (DAH) confirmed on bronchoalveolar lavage.

DISCUSSION

CAPS represents the most extreme manifestation of APS, characterized by a "thrombotic storm" that results in widespread microvascular and macrovascular occlusions.[7,8] As demonstrated in our patient, CAPS is a diagnostic masquerade, often confused with systemic vasculitis or sepsis.[3-7] The hallmark of CAPS is the rapid progression of multiorgan failure within a very short timeframe.[7] Our patient met the preliminary classification criteria (Asherson criteria) by exhibiting involvement of four distinct organ systems: vascular(aorto-iliac thrombosis), pulmonary (haemothorax and DAH), renal (AKI with haematuria), and neurological (deterioration requiring ventilation).[5,9,10] According to the 2023 ACR/EULAR Antiphospholipid Syndrome Classification Criteria, the inclusion of high-titer beta2-glycoprotein I antibodies—which were strongly positive in our patient—carries significant diagnostic weight, even when other markers are negative.[11] The presence of DAH in this case, confirmed via bronchoalveolar lavage, presented a profound therapeutic dilemma.[4,5] While DAH involves bleeding, in the context of CAPS it is often driven by microvascular thrombosis of the alveolar capillaries; thus, anticoagulation remains essential despite the pulmonary hemorrhage.[5-7] Management of CAPS typically requires a triple therapy approach: Anticoagulation, high-dose corticosteroids, and plasma exchange (TPE) or IVIG.[8] Data from the CAPS registry indicates that patients who receive this combined triple therapy have significantly higher survival rates compared to those receiving single or double therapy.[10] In our case, the patient's recovery after pulse methylprednisolone and unfractionated heparin suggests that early, aggressive intervention can halt the

thrombotic process.[4,5,7] This highlights the necessity of a coordinated, multidisciplinary team involving cardiology, nephrology, pulmonology, and rheumatology to navigate the complex balance of immunosuppression and anticoagulation.[5,10]

Clinical significance

This case is clinically significant as it represents a rare and fulminant presentation of CAPS, involving simultaneous large-vessel thrombosis, diffuse alveolar haemorrhage, renal dysfunction, and neurological impairment. It highlights the diagnostic challenge in differentiating CAPS from mimicking conditions such as vasculitis, sepsis, or thrombotic microangiopathy, especially in the context of negative ANA, ANCA, and anti-GBM antibodies.

The relevance lies in the fact that β2-glycoprotein antibody positivity provided the critical clue, reminding clinicians of the importance of targeted antibody testing in unexplained multiorgan failure. Management of this case also underscores the therapeutic dilemma of balancing anticoagulation in the presence of life-threatening haemorrhage, emphasizing the role of early recognition, aggressive multimodal therapy, and multidisciplinary collaboration in improving survival outcomes in CAPS.

CONCLUSION

This case illustrates the clinical masquerade of CAPS, which presented initially with large vessel thrombosis and later progressed to pulmonary haemorrhage, renal failure, and neurological impairment. Diagnostic delay is common due to overlap with other autoimmune and infectious diseases. A multidisciplinary team approach, early recognition, and aggressive multimodal therapy are essential to improve outcomes in this otherwise often fatal condition.

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