Case Report

A Rare Presentation of Probable Catastrophic Antiphospholipid Syndrome with Bilateral Adrenal Haemorrhage Secondary to Systemic Lupus Erythematosus

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Abstract :

Background :

Catastrophic antiphospholipid syndrome (CAPS), a rare thrombotic manifestation of antiphospholipid syndrome (APS), carries high mortality. Bilateral adrenal haemorrhage (BAH), an unusual complication of APS, is seldom reported. This case highlights a rare presentation of bilateral adrenal haemor- rhage due to probable CAPS secondary to systemic lupus erythematosus (SLE).

Case Presentation :

A 57-year-old diabetic female from India, presented with hypovolemic shock, abdominal pain, and vomiting. Clinical evaluation, imaging (CECT abdomen), and laboratory investigations (anti-â2GP1 IgG, lupus anticoagulant, anticardiolipin antibodies) were conducted. Secondary causes like sepsis, trauma, and coagulopathies were excluded. SLE workup included ANA and anti-dsDNA testing.

CECT revealed bilateral adrenal haemorrhage. Persistent positivity for antiphospholipid antibodies confirmed APS. Elevated ANA (1:100) and anti-dsDNA suggested underlying SLE, despite absent typical symptoms. Serum cortisol (9.2 µg/ dL) and ACTH (180 pg/mL) indicated evolving adrenal insufficiency. The patient received methylprednisolone pulses, IVIG, anticoagulants, and immunosuppres-sants (MMF), achieving rapid stabilisation without mineralocorticoid replace- ment.

Conclusion :

This case underscores Bilateral Adrenal haemorrhage as a rare and atypical manifestation of CAPS and any case of APS should always be evaluated for background SLE. Early diagnosis via antibody testing and imaging, combined with aggressive immunosuppression and anticoagulation, improves outcomes.

This report enriches the limited literature on CAPS-related BAH and reinforces multidisciplinary management in thrombotic-autoimmune syndromes.

Keywords :

Anti Phospholipid Syndrome (APS); Bilatera Adrenal Haemorrhage (BAH); Systemic Lupus Erythematosus (SLE)

1. Introduction :

APS (antiphospholipid syndrome) is an autoantibody mediated acquired throm-bophilia

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characterised by recurrent arterial or venous thrombosis and/or pregnancy morbidity. It could be Primary or Secondary to other autoimmune disease.

Currently the incidence of APS is 5 cases out of 1,00,000 population.[1] 1% of APS patients present with Catastrophic APS.[2] Mortality accounted for 37% cases of CAPS with CAPS associated with SLE accounting to higher mortality (48%).[3] Here we will discuss a rare presentation of probable CAPS in a back- ground of SLE.

2. Case Summary & Discussion :

A 57 years old diabetic, non-hypertensive, Indian female presented with severe prostration and pain abdomen associated with vomiting for 1 week. The pain was acute and generalised, there was no aggravating or relieving factor, no history of radiation, dysuria, or fever. On examination features of hypovolaemic shock were present and she was resuscitated in ICU. But there was persistently low blood pressure. Routine Investigations are as in Table 1. Any infectious or surgical aetiology was ruled out through extensive evaluation.

Table 1. Laboratory Investigations

Parameter	Value	Parameter	Value
Total Leucocyte Count	18,000/mm³ (88% neutrophils)	Serum Albumin	4.2 mg/dL
Platelet Count	85,000/mm³	Serum Globulin	3.2 mg/dL
ESR	100 mm/hr	SGOT	57.8 U/L
CRP	101.2 mg/L	SGPT	39.8 U/L
Procalcitonin	0.03 ng/mL	Amylase	41 U/L
Sodium	126 mEq/L	Lipase	63 U/L
Potassium	3.3 mEq/L	Urine R/E & M/E	OBT positive, RBC 60-80/HPF, Albumin ++, Pus cells 2-3/HPF
Urea	19.7 mg/dL	Blood Cul- ture & Urine Culture	No growth
Creatinine	0.9 mg/dL	Arterial Blood Gas (ABG)	No acidosis; Lactate 1.4 mmol/L

USG Whole Abdomen showed suspected Suprarenal lesion. So, **CECT Abdomen** was done, which showed hyper dense, non-enhancing lesion in B/L adrenal with perilesional fat stranding – likely Bilateral **Adrenal Haemorrhage.**

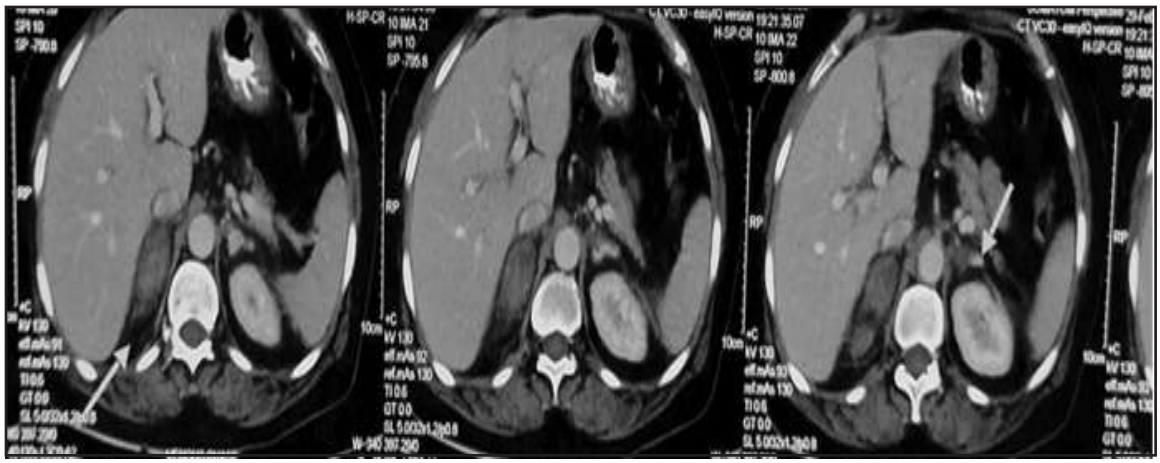

Figure A1. CECT Abdomen showing Adrenal Haemorrhage (marked by yellow arrows)

Then we revisited the case. There was no history of anti-coagulation, gram-negative septicaemia, trauma, no previous history of foetal demise, no previous history of recurrent stroke. There was no precipitating history suggestive of Disseminated Intravascular Coagulation, Thrombotic Microangiopathy. Peripheral Blood Smear was rechecked to rule out Microangiopathic changes like schistocytes. Also, platelet count was low, Prothrombin time (PT) normal, Activated Partial Thromboplastin Time was mildly raised which goes against Disseminated Intravascular Coagulation. There was also no history suggestive of Small Vessel Vasculitis. So, though rare, but to rule out APS (which could be a potential cause of B/L Adrenal Haemorrhage), we did the following investigations: **Anti-B2GP1 IgG (14.00 U/ml), Lupus Anticoagulant, Anti Cardiolipin Antibody IgG was positive (55 GPL)**. These antibodies were repeated after 12 weeks and came to be positive. APS was diagnosed. There are very few reported cases of B/L Adrenal haemorrhage in APS, as by Hana Aldajaani *et al*[4] and Dr. Rashika Bansal *et al* recently.[5]

To rule out any secondary cause, Blood for ANA (IIFT) – Hep 20-10 Cell Line was done, which showed Positive result in 1:100 dilution with **Dense Fine Speckled Nuclear** pattern. **Anti-ds DNA was positive in high titre**.

But in this case, we didn't get any history of skin rash, recurrent oral ulcers, any vasculitic rash, low back pain or any inflammatory polyarthritis.

So, a **Probable Catastrophic APS** secondary to SLE was diagnosed. Simultaneously Serum 8 am Cortisol was 9.2µg/dl with Serum ACTH value 180 pg/ml which showed an evolving stage of Primary Adrenal Insufficiency.

Table A1: Criteria for the Classification of Catastrophic Antiphospholipid Syndrome (CAPS)

Criteria	Description
1. Multi-organ involvement	Evidence of involvement of three or more organs, systems and/or tissues†
2. Rapid development	Development of manifestations simultaneously or in less than a week
3. Histopathological confirmation	Confirmation by histopathology of small vessel occlusion in at least one organ or tissue‡
4. Laboratory confirmation	Laboratory confirmation of the presence of antiphospholipid antibodies (lupus anticoagulant and/or anticardiolipin anti- bodies)§

Classification Categories :

Category	Criteria Required
Definite Catastrophic APS	All four criteria
Probable Catastrophic APS	All four criteria, except for only two organs, systems and/or tissues involvement
	All four criteria, except for the absence of laboratory confirmation at least six weeks apart due to the early death of a patient never tested for aPL before the cat- astrophic APS
	Criteria 1, 2 and 4
	Criteria 1, 3 and 4 and the development of a third event in more than a week but less than a month, de- spite anticoagulation

Notes :

- Usually, clinical evidence of vessel occlusions, confirmed by imaging techniques when appropriate. Renal involvement is defined by a 50% rise in serum creatinine, severe systemic hypertension (>180/100 mmHg) and/or proteinuria (>500 mg/24 hours).
- For histopathological confirmation, significant evidence of thrombosis must be present, although vasculitis may coexist occasionally.
- If the patient had not been previously diagnosed as having an APS, the laboratory confirmation requires that presence of antiphospholipid antibodies must be detected on two or more occasions at least six weeks apart (not necessarily at the time of the event), according to the proposed preliminary criteria for the classification of definite APS.

Source: Adapted from 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.

Patient was started on Methyl prednisolone pulse therapy followed by IVIG. Subsequently the treatment was followed by maintenance steroid, immunosup- pressants like MMF, Anticoagulant. Surprisingly patient's Plasma Renin Activity was normal, so, mineralocorticoid replacement therapy was not needed. Patient responded dramatically and followed up. Now patient is in absolutely stable con- dition with oral steroid replacement.

Though rare, but Bilateral Adrenal haemorrhage could be a presentation of APS.

4. Discussion :

The temporal relationship between adrenal haemorrhage and evolving adrenal insufficiency in this case aligns with the "two-hit hypothesis" proposed for CAPS, where an initial triggering event (possibly subclinical SLE activity) leads to widespread endothelial damage and subsequent thrombotic microangiopathy.[6] Antiphospholipid antibodies, particularly anti-β2-glycoprotein I and anti-cardiolipin antibodies, play a crucial role in initiating the thrombotic cascade, by disrupting the annexin A5 shield on endothelial surfaces, exposing phosphatidylserine and creating a prothrombotic environment.[7] In SLE-associated APS, these effects are potentially amplified by complement activation and immune complex deposition, creating a hypercoagulable state that predisposes to microvascular thrombosis.[8]

The adrenal glands are particularly vulnerable to thrombosis and subsequent haemorrhage due to their unique vascular architecture. They receive rich arterial supply but drain through a single vein, creating a vascular watershed area susceptible to congestion and thrombosis.[9] When antiphospholipid antibodies trigger thrombosis in the adrenal veins, venous congestion ensues, leading to haemorrhagic infarction.[10] This pathophysiological mechanism explains the bilateral adrenal involvement observed in our patient.

The absence of typical SLE manifestations in our patient represents what some researchers have termed "serositis-dominant lupus," where visceral or serological manifestations predominate over cutaneous or articular features.[11] This phenotypic heterogeneity underscores the importance of comprehensive immunological evaluation in unusual thrombotic presentations, particularly involving the adrenal glands.[12]

5. Conclusion :

Although rare, bilateral adrenal haemorrhage should prompt consideration of APS in the differential diagnosis, particularly when associated with thrombocytopenia and positive antiphospholipid antibodies. This case highlights the importance of recognising CAPS as a potentially life-threatening condition requiring aggressive immunomodulatory and anticoagulant therapy. Early diagnosis and appropriate management can significantly improve outcomes in this high-mortal- ity condition.

6. Declaration :

Author Contributions : Jyotirmay Maji and Ajitesh Roy both contributed to the conception and design of the case report as well as in the

acquisition of the patient's data and imaging readings interpretation of data and the writing of the manuscript; both authors have read and agreed to the published version of the manuscript.

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7. Abbreviations :

The following abbreviations are used in this manuscript:

CAPS	Catastrophic Antiphospholipid Syndrome
BAH	Bilateral Adrenal Haemorrhage
SLE	Systemic Lupus Erythematosus
CECT	Contrast Enhanced Computed Tomography
β2G-	Beta 2 Glycoprotein P1
ANA	Anti Nuclear Antibody
dsD-	Double Stranded DNA NA
ACT	Adreno Corticotrophic Hormone H
IVIG	Intra-veinous Immunoglobulin
MMF	Mycophenolate Mofetil

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