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Antiphospholipid Antibody Syndrome: A Rare Case of Catastrophic APLA

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Abstract: *CATASTROPHIC APLA (CAPS) is defined as rapidly progressive thrombo-embolic disease involving simultaneously three or more organs, organ system or tissue leading to functional defects. It is characterized by multiple arterial and/or venous thrombotic events, including the microcirculation, occurring in a short period, and can affect any system. This syndrome can occur in individuals with known APS under treatment, or it can be its first manifestation; in most cases, there is a triggering factor that can be identified. In the present case report, patient presented with sudden onset blackening of tip of index finger and big toe of left lower limb associated with continuous pain, ischemic in nature indicating microangiopathy and livedo reticularis.*

Keywords*: CAPS, Thrombus, Rare, Emboli*

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I. Introduction

Antiphospholipid syndrome (APS) is a multisystem autoimmune condition characterized by vascular thromboses and/or pregnancy loss associated with persistently positive antiphospholipid antibodies (aPL; measured with lupus anticoagulant [LA] test, anticardiolipin antibody [aCL] enzyme linked immunosorbent assay [ELISA], and/or anti-β2-glycoprotein-I antibody [aβ2GPI] ELISA) [Miyakis et al. 2006][1]. APS may occur alone (primary) or in association with any other autoimmune disease (secondary). In its most severe form, a minority of patients develop life-threatening multiple organ thromboses, usually associated with microthrombosis, recognized as catastrophic APS (CAPS) [Asherson et al. 2003[2]; Vora et al. 2006[3]]. The prevalence of CAPS is currently estimated to be 1%, with a mortality rate of 37%[4].

II. Case Report

A 24 year female patient presented to subharti hospital with complaint of high grade fever continous in nature since 5 days associated with cough not associated with expectoration followed by loose stool watery in consistency ,3-4 episodes daily but no h/o blood in stool and multiple episodes of vomiting projectile in nature containing food particles but no h/o blood in vomiting associated with dull aching continous pain in abdomen with petechiae present all over body. patient presented with sudden onset blackening of tip of index finger and big toe of left lower limb associated with continous pain, ischemic in nature.

On Examination: The following parameters were observed on examination:

a. General examination:
1. BP-112/70 mm hg pulse,
2. 98bpm,
3. Pallor-present,
4. Icterus-present
5. No palpable lymph nodes
6. Cyanosis present
7. Petechiae present all over body no edema present
8. Lace like purplish discoloration of skin (LIVEDO RETICULARIS).

b. Systemic examination
1. CVS – S1,S2 Present,
2. No murmur heard,

3. Tachycardia present.

c. Respiratory system

1. Bilateral air entry present,
2. Normal vesicular breath sound heard.

d. Abdomen system

1. Soft, distended,tenderness present all over abdomen,
2. No organomegaly present,
3. Blanching present on palpation,
4. Bowel sound present.

e. CNS examination

1. Patient conscious oriented to time,place, person,
2. Higher mental function intact,
3. Bilateral planter flexor,
4. Power in all four limb normal,
5. Tone normal.

Investigations at present:HB-10.1, Urea-101, Total bil.-0.6, TLC-27500,Creatinine-1.8, Total protein-5.2, PLT-25,000, Sodium-138, AST-146, Pt-16.7, Potassium-2.9, ALP-133, Inr-1.44, Calcium-8.1, Albumin-2.4, APTT-39.2, Globulin-2.8, Cardiolipin antibody(IgA)-7.02, Cardiolipin antibody(IgM)-8.10, Partial Thromboplastin Time-Lupus anticoagulant specific-39.80 seconds (normal range is 25.78-32.58).

BETA 2 GLYCOPROTEIN I, PANEL	Bio.Ref.Interval
Beta 2 glycoprotein IgG-8.62 SGU	<20
Beta 2 glycoprotein IgM-33.78 SMU	<20
Beta 2 glycoprotein IgA-214.24 SAU	<20

Lupus Anticoagulants are heterogenous IgG or IgM auto-antibodies which interfere with phospholipid dependent coagulation tests,particularly PTT. These are associated with SLE, thrombocytopenia,recurrent spontaneous abortion and thrombo- embolic disease states. Abnormal PTT-LA tests have been reported in approximate 30% risk of developing symptoms characteristic of APLA syndrome.

III. Discussion

Catastrophic antiphospholipid syndrome wasfirst described by Asherson et al[5] in 1992.24 year female patient presented with complaints of fever with loose stool and vomiting. On general examination petechia present all over body and blackening of index finger and big toe of left lower limb indicating microangiopathy and livedo reticularis (lace like reticular pattern present over b/l lower limb.In this case initial differential diagnosis thought of were vasculitis,sepsis with dic,antiphospholipid syndrome,thrombotic thrombocytopenic purpura/hemolytic uremic syndrome.

A criterion for classification of catastrophic antiphospholipid antibody is presented in box 1[6].On the basis of presentation,examination of the patient and the diagnostic criteria (box 1), the present case was diagnosed as a case of a rare case of catastrophic APLA.

In reviewingthe CAPS literature, we found three reportedcases of thromboses in the thyroid, two in the stomach, one in the bladder, and three in muscles in CAPS patients[5, 7-10]. Chinnery etal.[10] reported a female SLE patient with multiplethrombosis, including gastric and bladderinvolvement as observed in our patient, withpositive tests for LA and IgG aCL. Cisternas et al.[7] described a CAPS case that presented withthyroid and muscle thromboses, in addition tointestinal, adrenal, pancreatic, and renal thromboses,with LA and IgG aCL positivity. Mizunoet al.[8] also reported a female SLE patientwith multiple unusual sites of thromboses, includingthe thyroid, brain, fingers, liver, spleen,pancreas, and kidneys with positive assays forLA and IgG aCL. Asherson et al.[5]reported anSLE patient with gastric thrombosis who alsohad renal and splenic thromboses with positiveIgG aCL.

IV. Conclusion

Since CAPS is a rare disease, case reports are important to provide a better understanding of its various clinical aspects to facilitate future diagnosis. CAPS is truly a rheumatologic emergency, requiring prompt recognition and diagnosis as well as aggressive treatment to avoid the fulminant irreversible complications related to its devastating prognosis.

1. Evidence of involvement of 3 organs, systems, and/or tissues[b].
2. Development of manifestations simultaneously or in <1 week
3. Confirmation by histopathology of small-vessel occlusion in at least one organ/tissue[c].
4. Laboratory confirmation of the presence of aPL (lupus anticoagulant and/or aCL an/or anti β2 GP I)

Definite CAPS
All four criteria
Probable CAPS

- Criteria 2, 3, and 4, plus evidence of involvement of only two organs, systems, and/or tissues
- All four criteria, except for the absence of laboratory confirmation of the presence of aPL at least 6 weeks after a first positive result (because of the early death of a patient never tested for aPL before onset of CAPS)
- Criteria 1,2, and 4
- Criteria 1,3, and 4, plus the development of a third event in >1 week but <1 month, despite anticoagulation treatment

[a]Proposed and accepted during the 10th International Congress on Antiphospholipid Antibodies
(aPL), September 2002.
[b]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 mm Hg), and/or proteinuria (>500 mg/24h).
[c]For histopathologic confirmation, significant evidence of thrombosis must be present, although vasculitis may coexist occasionally.

Box 1: Preliminary criteria for the classification of CAPS[a]

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