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# Male Apla Syndrome: Case Report

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***ABSTRACT***
*Anti-phospholipid syndrome is an autoimmune multisystem disorder characterized by arterial, venous or small vessel thromboembolic events and /or pregnancy morbidity in the presence of persistent antiphospholipid antibodies.(1) It may occur as a primary condition or may be secondary to an underlying autoimmune disease, most commonly, SLE. In addition to the hallmark features of venous/ arterial or small vessel thrombosis and pregnancy related complications, other features of APS includes TIA, thrombocytopenia, livedo reticularis.(2) In rare cases APS results in multiorgan failure to small vessel thrombosis, a condition referred to as catastrophic APS.(3)*

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## I. INTRODUCTION

Antiphospholipid antibodies are a heterogeneous group of antibodies, including anti cardiolipin antibody, lupus anticoagulant, anti beta 2 glycoprotein 1 antibody, which are directed against phospholipid binding proteins.(4) Presence of these antibodies results in an autoimmune multisystem disorder more commonly found in females with increased pregnancy related complications.(5) The disease is characterized by the presence of thrombosis, thrombocytopenia, recurrent pregnancy loss, livedo reticularis or even cardiac valvular diseases. The disease may manifest as Myocardial Infarction, Deep vein thrombosis or ischaemic stroke in young patients.(6) It may be an idiopathic primary disorder or maybe secondary to an underlying autoimmune disease most commonly SLE.(7) Through years of study it has been found that many of the antibody positive laboratory proven cases of APLA are clinically silent. Hence it is critical to find out those cases of APLA which are prone to develop thrombotic complications. It has been found that patients with elevated levels of IgG Anti Cardiolipin antibodies are at a much higher risk of developing thrombotic events as compared to patients with drug related / infection induced APLA.(8) This case in particular has been evaluated, followed up and presented since male APLA syndromes are rarely found.

## II. CASE REPORT

46 year old male, 2nd child of a non-consanguineous marriage, electrician by occupation, non smoker, non alcoholic with no relevant past medical or surgical history and no history of prior hospital admissions with family history of coronary artery disease in father at 60 years of age and no other relevant family history presented with complaints of Insidious onset, gradually progressive dyspnea on exertion since 9 months with acute worsening in the past 3 weeks from NYHA Fc 2 to Fc 4 associated with bilateral pedal edema for the past 3 months. He presented to the casualty of a tertiary care centre in south India with complaints of chest discomfort, exertional breathlessness and palpitation. On immediate evaluation cardiac biomarkers were found to be elevated and patient got admitted as a case of Non ST elevation MI.

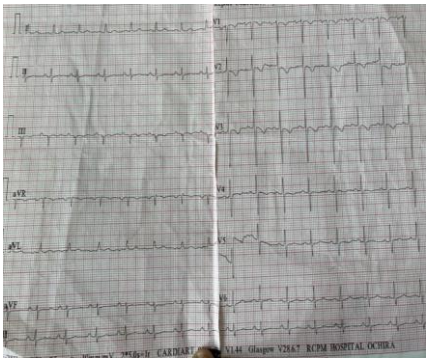


**FIG 1. ECG recording of the patient on admission**

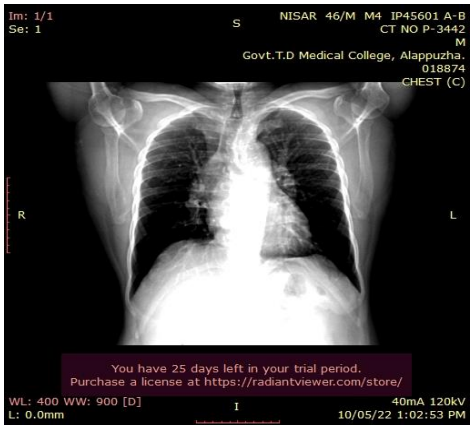
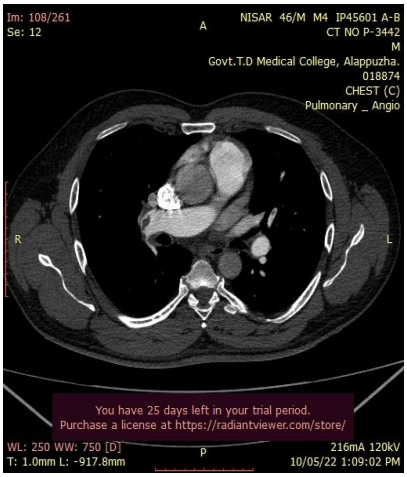



**FIG 2. Chest X Ray of the patient at the time of admission**

In view of the provisional diagnosis the patient was started immediately on Heparin, Dual anti platelet therapy and statins along with other supportive measures. There was no worsening of symptoms. On routine cardiology evaluation ECHO was done on the 4th day of admission and patient was detected to have severe PAH and TR. To rule out any valvular heart disease like ASD patient was scheduled for a detailed echo with bubble contrast soon. After a course of 5 days heparin was discontinued. Patient was by now symptomatically better. 2 days later an elaborate echo with bubble contrast was done and this showed an intact septum and ruled out any valvular heart disease. Now the probability of pulmonary embolism, though seemed less likely in a young patient with no comorbidities, had to be entertained to explain PAH. An emergency CTPA was done. The Provisional reports revealed the presence of embolus in both R and L pulmonary artery in segmental and sub segmental branches.

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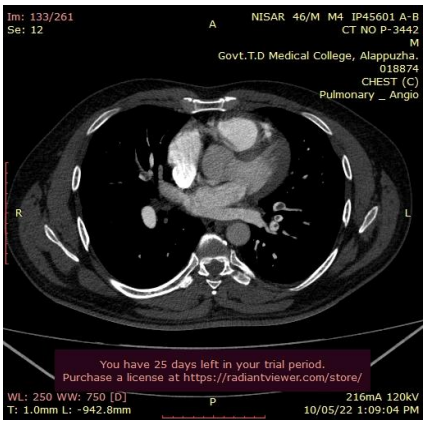


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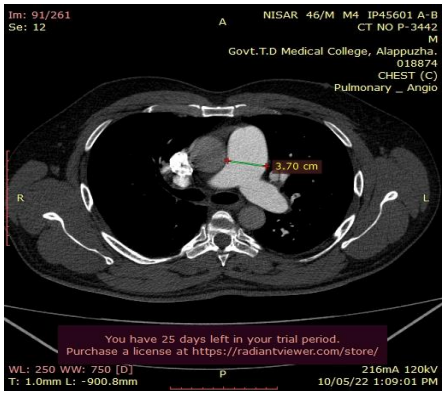


C

FIG 3. CT Pulmonary angiogram showing emboli in the R main pulmonary artery and L segmental arteries and © the dilated pulmonary trunk suggestive of pulmonary artery hypertension

Patient was restarted on heparin. B/L LL arterial Doppler was done and found to be normal. Detailed history taken and evaluation done for procoagulable states. Patient underwent screening for autoimmune conditions. Was found to have elevated Serum Antiphospholipid Ab IgM levels of **225.6U/L [>10],** Anti Cardiolipin Ab IgM **– 12.3 AU/ml [>7 ]** and Beta 2 Glycoprotein 1 IgM **– 64.9 U/ml [>10 ].** Serum homocysteine, Anti dsDNA, Protein C, Protein S levels and ANA profile were found to be within normal limits. His routine blood and urine investigations were within normal limits and viral markers negative. On evaluation his tumor markers were found to be negative, Thyroid function tests normal, and USG abdomen and pelvis showed fatty hepatomegaly alone. Patient was counseled regarding the disease and started on Warfarin, Heparin tapered off.(9) The rest of the hospital stay was uneventful and patient was discharged with advice

## III. DISCUSSION

Anti phospholipid syndrome is an autoimmune multisystem disorder characterized by arterial, venous or small vessel thromboembolic events and /or pregnancy morbidity in the presence of persistent antiphospholipid antibodies. Antiphospholipid antibodies are a heterogeneous group of antibodies which are directed against phospholipid binding proteins.(10) It may occur as a primary condition or may be secondary to an underlying autoimmune disease, most commonly, SLE. Although clinical presentations of both types of APLA are similar, patients with SLE associated APS are more likely to have arthritis, livedo reticularis, heart valve disease, thrombocytopenia and leucopenia.(11)

In addition to the hallmark features of venous/ arterial or small vessel thrombosis and pregnancy related complications, other features of APS includes TIA, thrombocytopenia, and livedo reticularis. In rare cases APS results in multiorgan failure to small vessel thrombosis, a condition referred to as catastrophic APS.

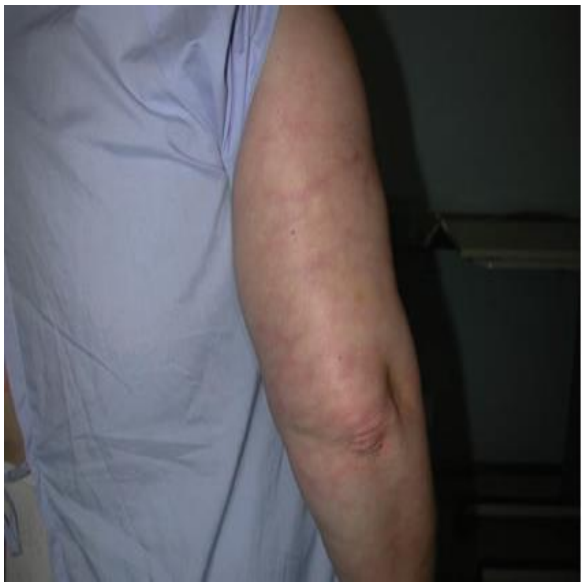

**FIG 4. Livedo reticularis**

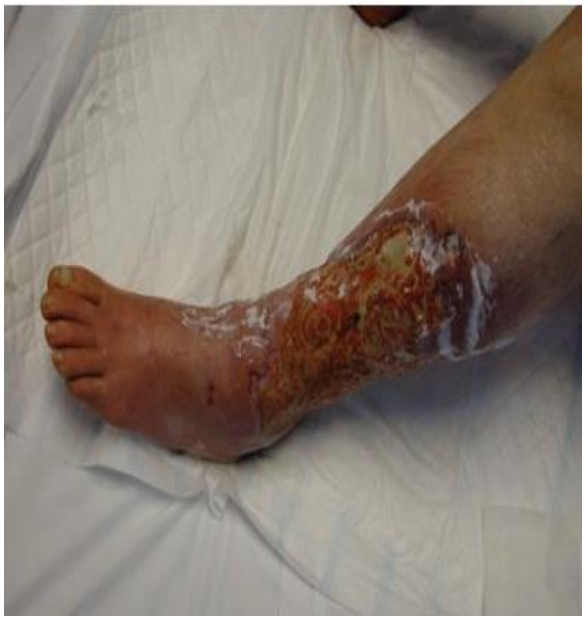

**FIG 5. Skin ulceration secondary to vascular thrombosis**

DIAGNOSTIC CRITERIA- SYDNEY CRITERIA(12)

**Clinical criteria**

1. Vascular thrombosis

   One or more clinical episodes of arterial, venous, or small-vessel thrombosis, in any tissue or organ.
   Thrombosis must be confirmed by objective validated criteria (i.e. unequivocal findings of appropriate imaging studies or histopathology). For histopathological confirmation, thrombosis should be present without significant evidence of inflammation in the vessel wall.

2. Pregnancy morbidity

   (a) One or more unexplained deaths of a morphologically normal fetus at or beyond the 10th week of gestation, with normal fetal morphology documented by ultrasound or by direct examination of the fetus, *or*
   (b) One or more premature births of a morphologically normal neonate before the 34th week of gestation because of: (i) eclampsia or severe pre-eclampsia defined according to standard definitions or (ii) recognized features of placental failure, *or*
   (c) Three or more unexplained consecutive spontaneous abortions before the 10th week of gestation, with maternal anatomic or hormonal abnormalities and paternal and maternal chromosomal causes excluded.

   In studies of populations of patients who have more than one type of pregnancy morbidity, investigators are strongly encouraged to stratify groups of subjects according to (a), (b), or (c) above

**Laboratory criteria**

1. Lupus anti-coagulant present in plasma, on 2 or more occasions at least 12 weeks apart, detected according to the guidelines of the International Society on Thrombosis and Hemostasis (Scientific Subcommittee on LA/phospholipid-dependent antibodies).
2. Anti-cardiolipin antibody of IgG and/or IgM isotype in serum or plasma, present in medium or high titres (i.e. >40 GPL or MPL, or greater than the 99th percentile), on 2 or more occasions, at least 12 weeks apart, measured by a standardized ELISA.
3. Anti-β2 glycoprotein I antibody of IgG and/or IgM isotype in serum or plasma (in titres greater than the 99th percentile), present on 2 or more occasions, at least 12 weeks apart, measured by a standardized ELISA, according to recommended procedures.

*Note:* Definite anti-phospholipid syndrome may be diagnosed if at least one of the clinical criteria and at least one of the laboratory criteria are met.
Modified from Miyakis et al.[13]

TREATMENT(12)

**General measures for anti-phospholipid antibody carriers**

- A strict control of cardiovascular risk factors should be accomplished in all individuals with a high-risk antiphospholipid antibody profile,[a] irrespective of the presence of previous thrombosis, concomitant SLE or additional APS features
- All APA carriers should receive thromboprophylaxis with usual doses of LMWH in high-risk situations, such as surgery, prolonged immobilization, and puerperium

**Primary thromboprophylaxis in SLE patients with anti-phospholipid antibodies**

- In patients with SLE and positive lupus anti-coagulant or isolated persistent anti-cardiolipin antibodies at medium–high titres, primary thromboprophylaxis with hydroxychloroquine ± low-dose aspirin is recommended

**Primary thromboprophylaxis in APA-positive individuals without SLE**

- In non-SLE individuals with APA and no previous thrombosis, long-term primary thromboprophylaxis with low-dose aspirin is recommended in those with a high-risk APA profile, especially in the presence of other thrombotic risk factors

**Secondary thromboprophylaxis**

- Patients with either arterial or venous thrombosis and APA who do not fulfil criteria for APS should be managed in the same manner as APA-negative patients with similar thrombotic events
- Patients with definite APS and a first venous event should receive oral anti-coagulant therapy to a target INR 2.0–3.0
- Patients with definite APS and arterial thrombosis should be treated with warfarin at an INR >3.0 or combined anti-aggregant–anti-coagulant therapy (INR 2.0–3.0)
- The patient's bleeding risk should be estimated before prescribing high-intensity anti-coagulant or combined anti-aggregant–anti-coagulant therapy
- Non-SLE patients with a first non-cardioembolic cerebral arterial event, with a low-risk APA profile[b] and the presence of reversible trigger factors could individually be considered candidates for treatment with antiplatelet agents.

**Duration of treatment**

- Indefinite antithrombotic therapy is recommended in patients with definite APS and thrombosis
- In cases of first venous event, low-risk APA profile[b] and a known transient precipitating factor, anti-coagulation could be limited to 3–6 months.

**Refractory and difficult cases**

- In patients with difficult management due to recurrent thrombosis, fluctuating INR levels, major bleeding or at a high risk for major bleeding, alternative therapies could include long-term LMWH, hydroxychloroquine, or statins.

TREATMENT OF APS IN PREGNANCY(12)

**APS with poor obstetric outcomes**

Recurrent early (pre-embryonic or embryonic) miscarriage

- Low-dose aspirin *alone, or plus*:
- LMWH at thromboprophylactic doses

Fetal death (>10 weeks' gestation) or prior early delivery (<34 weeks' gestation) due to severe pre-eclampsia or placental insufficiency

- Low-dose aspirin, *plus:*
- LMWH at thromboprophylactic doses

**APS with thrombosis**

- Low-dose aspirin, *plus:*
- LMWH

## IV. CONCLUSION

APS can manifest in numerous forms and a proper understanding of the various presentations can help in early detection to prevent any thrombotic complications.(13) Though typically found in females especially in the reproductive age group, male APS should also be looked out for as they may manifest as life threatening vascular thrombotic events.

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