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Secondary Anti-Phospholipid Antibody Syndrome with Lupus Nephritis, Subclinical Hypothyroidism and Vitamin D Deficiency- A Rare Case Report of A Male Patient

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Abstract: *Antiphospholipid antibody syndrome is defined by the presence of thromboembolic complications and/or pregnancy morbidity in the presence of persistently increased titers of anti-phospholipid antibodies. Its clinical presentation can be diverse and any organ can be involved, with a current impact in most surgical and medical specialties. Here we present a case of a 52 years old male who presented to us with features of renal failure and multiple joint pain with history of multiple ischaemic strokes in the past. He was diagnosed by us as a case of secondary APLA syndrome (which is rare in males) and subsequently treated and followed up. We conclude that clinicians need to have a high index of suspicion of APS in patients who present with a thrombotic episode – clinicians should investigate for the presence of antiphospholipid antibodies, as early diagnosis may influence the course of the disease. Furthermore, resources for the detection of antiphospholipid antibodies should be made readily available in resource-limited settings. Finally, patient education on the importance of drug compliance, periodic monitoring, and prevention of thrombosis is indispensable, especially as mortality could be associated with the effects of vascular thrombosis and/or the effects of bleeding due to anticoagulants.*
Key words: *APLA, Coagulation, Male*

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I. Introduction

Anti-phospholipid antibody syndrome is one of the most common thrombocytophilias which was first described in 1986[1]. Unfortunately it is not recognised often enough. Only 1-5% of the population suffering from APLA syndrome has an increased level of the antibodies in serum and 30-50% of them presents with symptoms which are often misdiagnosed[2].The most commonly detected subgroups of anti-phospholipid antibodies are lupus anticoagulant, anti-cardiolipin, and anti-beta-2-glycoprotein 1 antibodies[3]. The biological function of these antibodies, although not well elucidated, is thought to act as a natural anticoagulant[4]. Despite their name, lupus anticoagulant antibodies are associated with thromboembolic events rather than clinical bleeding. Anti-phospholipid antibodies can interfere with both pro- and anticoagulant pathways. In vitro, phospholipid surfaces inhibit pro-coagulant pathways and therefore prolong clotting; in vivo, meanwhile, the microenvironment of cell membranes promotes greater inhibition of anticoagulant pathways and thus also thrombosis[5]. Although the pathogenesis of APS is poorly understood, many mechanisms have been postulated, including the binding of anti-phospholipid antibodies to endothelial cells, which stimulates an upregulation of adhesion molecules, increasing leucocyte adhesion[6] and creating a prothrombotic state[7]. Clinically, APS can be very diverse and can occur in a variety of disease states, rendering diagnosis and treatment challenging, especially in resource-limited settings[8]. The lack of proper prevention in this patients causes severe complications and the most common causes of mortality are cerebrovascular accidents, acute coronary syndrome and infections[9]. Without any other associated autoimmune disease it is marked as primary APLA syndrome whereas while associated with others like SLE it is diagnosed as a secondary APLA syndrome[10].

II. Case report

A 52 years old male patient presented to us in Regional Institute of Medical College, Imphal, Manipur, India with pain in multiple small joints of hands and feet for 6 months and altered mental status for 3 days. He was a known alcoholic for more than 25 years of consumption of locally made alcoholic beverages 2-3 days a week. He was also a known smoker consuming one pack of cigarette per day. He had two episodes of ischaemic stroke in last 3 years. He had no history of Tuberculosis, Diabetes Mellitus or any surgical intervention or any other major medical event.

After admission he was started on conservative treatment and all routine investigations were done. Raised serum creatinine level, proteinuria with altered sensorium and history of recurrent thrombotic stroke made us to look for autoimmune pathologies and the disease was diagnosed subsequently.

Laboratory findings:

1. Complete Blood Count- Hb- 7.1 g/dL (decreased), TLC- 13990 cells/cmm (raised), DLC- Neutrophils- 81%, Lymphocytes 11%, Plt- 3.40 lakhs/cmm (normal)
2. ESR- 130 mm in 1st hour (raised)
3. Liver Function Test- within normal ranges
4. Kidney Function Test- Blood Urea- 107 mg/dL, Serum Creatinine- 6.7 mg/dL
5. Routine Urine Examination- Protein- 2+, rest all- within normal limits
6. Thyroid Function Test- TSH- 5.32 (raised), T4- 112 (normal), T3- 1.74 (normal)
7. RBG- 90 mg/dL
8. HbsAg/ anti- HCV Ab/ HIV-Ab- non reactive
9. Serum Lipid Profile- withinnormal ranges
10. ANA- positive, anti ds-DNA- >200 IU/mL (highly positive)
11. RA factor- negative, Anti CCP Ab- negative
12. APLA- IgG- 36.15 U/mL (raised), IgM- not elevated
13. Anti cardiolipin antibody- 53 units (elevated)
14. Anti beta-2 glycoprotein- Raised
15. Vitamin D- 29 ng/dL (deficiency), Calcium- 8.9 mg/dL, Phosphorus- 5.2 mg/dL

Radiology Findings-

1. Chest X-Ray PAV- Normal.
2. CT Scan of Chest- Cardiomegaly with Pulmonary Hypertension, Moderate Pericardial and Bilateral Pleural Effusions.
3. CT Pulmonary Angiography- Normal pulmonary arteriogram.
4. Ultrasonography of abdomen- Normal.

Management:Patient was given low molecular weight heparin injection subcutaneously 60ug twice daily and methylprednisolone injection intravenously 1g daily for 5 days. This was followed by oral warfarin 1mg once a day and oral Prednisolone tablet 20 mg twice a day. He underwent haemodialysis two times and his serum creatinine came down to 1.1 mg/dL and after stabilization of his clinical conditions he was discharged. On discharge he was also added on vitamin D capsules 60000 IU perweek for 8 weeks followed by maintainance dose.

Follow-up:He was followed up after 1 week for clinical assessment. He was physically well, maintaining INR at 2.1 and was advised for a renal biopsy test which he denied to do. After 13 weeks he was again followed up with a new APLA level report which was still elevated and the diagnosis of secondary APLA syndrome was confirmed. After that patient did not turn up for further follow up.

III. Discussion

Though APLAS is one of the most common thrombocytophilias, unfortunately, it is not recognized often enough. That is why in every case of recurring thrombosis, especially in an atypical localization or an atypical aetiology, APLAS should be considered.Contemporary optimal treatment of APS, especially in the case of the catastrophic form, has three main aims: to treat any precipitating factors (prompt use of antibiotics if infection is suspected, amputation of any necrotic digits, high awareness in patients with APS who undergo an operation or an invasive procedure), to prevent and treat ongoing thrombotic events as well as to suppress excessive cytokine action. The most commonly applied therapy includes anticoagulation (usually intravenous heparin followed by oral anticoagulants), corticosteroids, plasma exchange, intravenous gamma globulins and, if associated with lupus flare, cyclophosphamide. Anticoagulant therapy of an unlimited duration should be recommended in patients with a diagnosed APS. According to actual consent in prevention of the recurrence of thrombotic-embolic disease maintaining INR in the range of 2.0–3.0 gives equal results as in the range of 3.1–4.0. Not smoking and avoiding the hormonal contraception in those patients is also of a great importance. So far, the management of patients with APS has been mainly supportive, generally aimed at avoiding recurrent thrombotic events. It seems that therapy targeted at the triggering factor (antiphospholipid antibodies) including a combination of plasmapheresis, which reduces the serum antibodies level, and anti-CD20 antibody (*rituximab*), which inhibits their production, could offer a promising perspective[1].

IV. Conclusion

In conclusion, clinicians need to have a high index of suspicion of APS in patients who present with a thrombotic episode. Management of APS that involves prevention of thrombosis together with close monitoring of the patient on an anticoagulant could be quite challenging. Therefore, patient education on the importance of drug compliance, periodic monitoring, and prevention of thrombosis is indispensable, especially as mortality could be associated with the effects of vascular thrombosis and/or the effects of bleeding due to anticoagulants.

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