

## Case Study

### Antiphospholipid Antibody Syndrome in Pregnancy presenting with Severe Thrombocytopenia - A success story

Sunil V. Kapur<sup>1</sup>, Dr Shreya Gowri<sup>2</sup>

#### Author's Affiliation:

1- Consultant Rheumatologist and Immunologist: Asian Rheumatology Center-Warangal- Telangana-India.

2- Matrika Fertility Center- Warangal- Telangana-India.

#### Correspondence:

Dr. Sunil V. Kapur, Email: sunil.v.kapur@gmail.com

Received on: 14-May-2024

Accepted for Publication: 24-Sep-2024

#### Introduction

Venous or arterial thrombosis and/or pregnancy morbidity in patients with persistent antiphospholipid antibody (aPL) presence are characteristics of the antiphospholipid syndrome (APS). Antithrombotic therapy is the cornerstone of treatment in APS as it is an autoimmune thrombophilic condition. Even though APS is a prothrombotic condition, some patients have thrombocytopenia, which can make treatment more challenging in terms of using antithrombotic medication. Thrombocytopenia does not seem to reduce the risk of thrombosis in patients with APS, who may experience thromboembolic complications requiring antithrombotic therapy. In these situations, thrombocytopenia treatment might be required to make it easier to administer antithrombotic medications. We describe a case of a pregnant woman who had a history of repeated miscarriages, who manifested symptoms of severe thrombocytopenia and bleeding, and who was eventually diagnosed with antiphospholipid antibody syndrome. Initially treated with steroids, antithrombotic therapy was initiated when her platelet count began to rise. She had an uneventful and successful pregnancy, with no complications during the follow-up.

#### Case Report:

A 26-year-old woman, gravida 3, para 1, living 1, abortus 1, had her first pregnancy end spontaneously at 10 weeks gestation. Her second pregnancy was complicated by severe oligohydramnios and intrauterine growth restriction (IUGR), resulting in a preterm indicated caesarean section at 28 weeks. The baby, a girl who weighed just 700 g, needed to spend three months in the Neonatal Intensive Care Unit (NICU) before being released in better health. In her current pregnancy, she experienced petechial rashes and gum bleeding throughout her body. There was no history of hematemesis, melena, epistaxis, or bleeding per vaginam. There was not history of migraines, arthralgias, myalgias, red eyes, mouth ulcers, weight loss, or constitutional symptoms. Laboratory investigations showed platelet count—10,000/cumm, haemoglobin—7 gms %, and total leucocyte count (TLC) of 7000/cumm. She had normal liver and kidney function tests. She had a persistently low platelet count ranging from 20,000/cumm to 37,000/cumm, prolonged bleeding time, normal clotting time, normal prothrombin time (PT), and activated partial thromboplastin time (aPTT). Her HIV, HBV, and HCV antibodies were all negative. Her dengue serology and malaria parasite tests were negative. The D-dimer study was found to be negative. Her APS profile revealed lupus anticoagulant positivity with negative anti B2 glycoprotein 1 antibody and anticardiolipin antibody. Her anti-nuclear antibody (ANA) by immunofluorescence assay (IFA) and Anti-double strand- (ds) DNA was negative. Bone marrow aspiration examination revealed normal marrow cellularity and increased megakaryocyte (MK) proliferation. She was diagnosed as primary APS and put on oral steroids (60

mg/day) and platelet transfusions were given following which her platelet normalized in 2 months' time. She was later started on Tab aspirin 75 mg OD, tablet hydroxychloroquine 200 mg OD and injection enoxaparin 60 mg/0.6 ml s/c once daily for APS syndrome. As the pregnancy progressed, injection Clexane was stopped at 37 weeks and aspirin was stopped at 36 weeks. At 38 weeks, an elective cesarean section was performed. The patient delivered a live, term, small for gestational age (SGA) baby girl weighing 2 kg, with APGAR scores of 8 and 9 at one and five minutes, respectively. The infant was admitted to the NICU for observation due to their low birth weight. Twelve hours after the cesarean section, enoxaparin was administered, and the patient remained stable throughout the postoperative period. All routine laboratory tests, including coagulation profiles, were found to be normal.

## Discussion

Antiphospholipid syndrome (APS) is a hypercoagulable and autoimmune clinical condition brought on by antiphospholipid antibodies.<sup>1,2</sup> Similar to our patient, primary APS patients do not exhibit clinical signs of any secondary autoimmune disease. A class of antibodies known as antiphospholipid antibodies reacts with a wide range of antigens, such as anticardiolipin antibodies, lupus anticoagulation antibodies and anti-acid phospholipid antibodies.<sup>3,4</sup> These antibodies primarily result in thrombocytopenia and arteriovenous thrombosis, which can cause a number of clinical symptoms, including kidney damage, stroke, heart attack, deep vein thrombosis, pulmonary embolism, and thrombocytopenia. <sup>3,4</sup> APS can result in stillbirth, premature birth, and miscarriage in expectant mothers. Approximately 20 to 40 percent of patients with APS have been reported to have thrombocytopenia, which is typically mild and does not require clinical intervention. However, 5–10% of patients may have severe thrombocytopenia (platelet count <50,000/cumm). It's interesting to note that thrombocytopenia is not usually linked to hemorrhagic consequences in APS patients.<sup>3,4</sup> In our instance, there was severe thrombocytopenia and signs of bleeding. Treatment for APS associated thrombocytopenia is generally indicated in cases of overt bleeding irrespective of platelet count. In addition, despite being thrombocytopenic, patients with APS experience thromboembolic complications. As a result, before beginning antithrombotic therapy, treatment is required to raise the platelet count to at least 30-50,000/cu.mm. Treatment options include glucocorticoids, intravenous immunoglobulin, rituximab, and immunosuppressive medications such as azathioprine and cyclophosphamide. <sup>5, 6</sup>

In our case, the expectant mother had severe thrombocytopenia with bleeding, a poor obstetric history, and APS. She responded well to steroid therapy and platelet transfusion with good recovery of platelet count. Patient was successfully started on antithrombotic treatment and delivered a healthy baby at term.

## References

1. Mialdea M, Sangle SR, D'Cruz DP. Antiphospholipid (Hughes) syndrome: beyond pregnancy morbidity and thrombosis. *J Autoimmune Dis.* 2009;6:3–7.
2. Miyakis S, Lockshin MD, Atsumi T, et al. International consensus statement on an update of the classification criteria for definite antiphospholipid syndrome (APS). *J Thromb Haemost.* 2006;4(2):295–306.
3. Dusse LM, Silva FD, Freitas LG, et al. Antiphospholipid syndrome: a clinical and laboratorial challenge. *Rev Assoc Med Bras.* 2014;60(2):181–186.
4. Kutteh WH. Antiphospholipid antibody syndrome and reproduction. *Curr Opin Obstet Gynecol.* 2014;26(4):260–265.

5. M. J. Cuadrado, F. Mujic, E. Muñoz, M. A. Khamashta, and G. R. V. Hughes, "Thrombocytopenia in the antiphospholipid syndrome," *Annals of the Rheumatic Diseases*, vol. 56, no. 3, pp. 194–196, 1997.
6. G. Finazzi, "The Italian registry of antiphospholipid antibodies," *Haematologica*, vol. 82, no. 1, pp. 101–105, 1997.