Annals of Case Reports
Li R and Yue Q. Ann Case Report 6: 723.
www.doi.org/10.29011/2574-7754.100723
www.gavinpublishers.com

Case Report

Primary Antiphospholipid Syndrome with Thrombocytopenia in the Second Pregnancy: A Case Report

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Citation: Li R, Yue Q (2021) Primary Antiphospholipid Syndrome with Thrombocytopenia in the Second Pregnancy: A Case Report. Ann Case Report 6: 722. DOI: 10.29011/2574-7754.100722

Received Date: 23 December, 2021**; Accepted Date:** 27 December, 2021**; Published Date:** 30 December, 2021

Abstract

Primary antiphospholipid syndrome (PAPS) is a multiple system clinical syndrome caused only by antiphospholipid antibodies (aPL). Currently, patients with typical clinical manifestations in recurrent miscarriage have been widely screened for the disease get diagnosed. We present a case of PAPS with thrombocytopenia in the second pregnancy, had thrombosis twice without habitual abortion occurring in a 34-year-old Chinese patient. She had discovered thrombosis in the lower limbs 9 years and 6 years ago, gave birth to a healthy baby boy four years ago without a history of miscarriage. There was no other cause for thrombocytopenia in the middle-late stages in this pregnancy. Cesarean section immediately after diagnosis, a healthy 3200g male baby was delivered. Our case shows that the PAPS should be considered when only occurring thrombocytopenia and thrombosis, without typical symptoms such as recurrent miscarriage.

Keywords: Primary antiphospholipid syndrome; Thrombocytopenia; Thrombosis; Recurrent miscarriage

Introduction

PAPS most frequently occurring recurrent miscarriage, but also have other symptoms. However, only manifested as thrombocytopenia and thrombosis is exceedingly rear. It may be easily misdiagnosed because of its atypical clinical symptoms. Primary antiphospholipid syndrome with thrombocytopenia and a history of thrombosis without recurrent miscarriage not been reported so far. The present paper describes an interesting case of severe PAPS and review the literature in order to improve our understanding of the disease, avoid misdiagnosis and provide evidence for its clinical treatment and prognosis.

Case Report

A 34-year-old Chinese woman was admitted to the hospital due to thrombocytopenia was found for more than 3 months, and platelets decreased slightly for 1 day, the minimum value of the platelets 54 × 10 ^ 9 / L. She had a history of deep vein thrombosis in the left lower extremity and placed filter in the right femoral vein and removed 14 days later 6 and 9 years ago. She was in the 40nd week of her second pregnancy. She gave birth 4 years ago. Routine clinical biochemistry showed platelet count 67×10^9/L. D- dimer: 1.22mg / L, partial thromboplastin time: 28.7×10^9/L, ratio of prothrombin time: 0.94. Liver and kidney were all within normal ranges. Ultrasound examinations showed: bilateral carotid and vertebral artery blood flow is still smooth, structural heart prompted echocardiography showed no abnormalities (Figure 1). Tests of thrombotic risk including anti-β2 glycoprotein IgG 1784.2 U/ml, anti-cardiolipin antibody IgG: 287.5 U/L. Lupus anticoagulant factor DVRRT screened 50.3S, Lupus anticoagulant factor DVRRT confirmed: 38.5S, DRVVT ratio: 1.31, antinuclear antibody were within normal limits. Laboratory tests support the final diagnosis of PAPS.

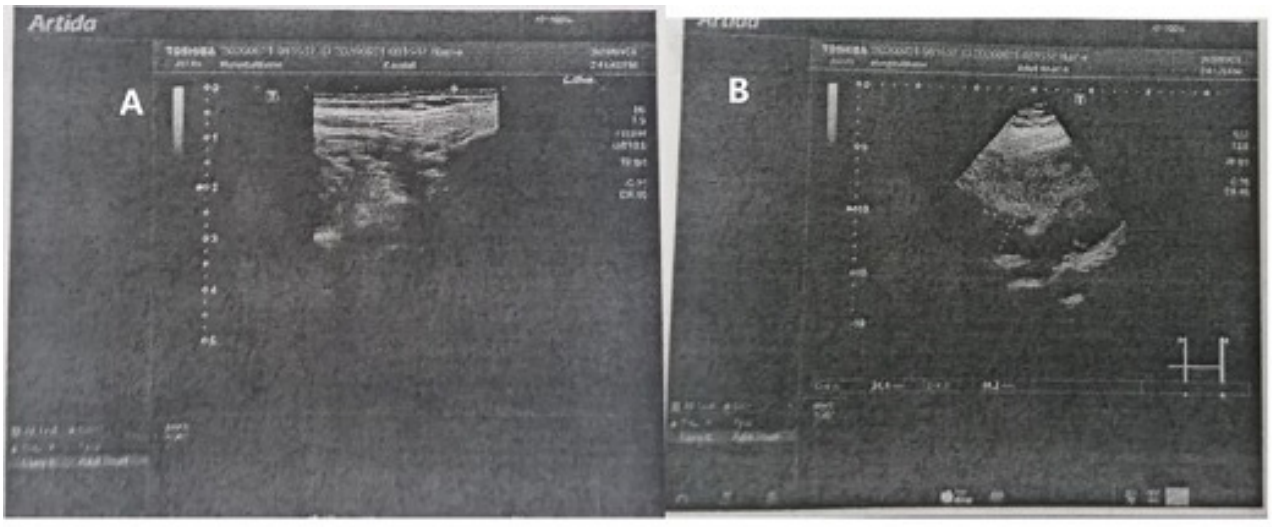

Figure 1: Structural heart prompted echocardiography showed no abnormalities.

After an urgent cesarean section, the patient successful deliver a 3200g male baby and received uneventful recovery. Postoperative color Doppler ultrasound examination showed that the blood in both carotid and vertebral arteries was still unobstructed (Figure 2). On the right leg veins dilate, lower extremity venous blood flow is still smooth over. Heart color Doppler ultrasound prompt: no abnormalities, She received administration of low molecular weight heparin 5000IU / d until 42 days postpartum. Subsequently, Tests of blood coagulation function, platelet aggregation function and blood routine were within normal limits. 42 days postpartum review, lower limbs No fresh thrombosis (Figure 3).

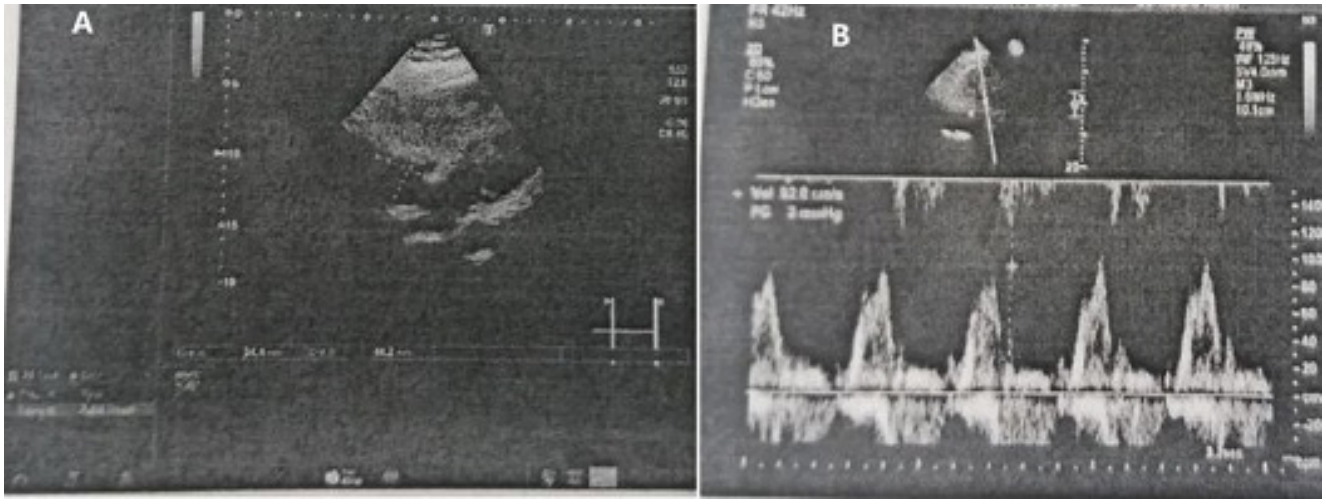

Figure 2: The blood in both carotid and vertebral arteries was still unobstructed.

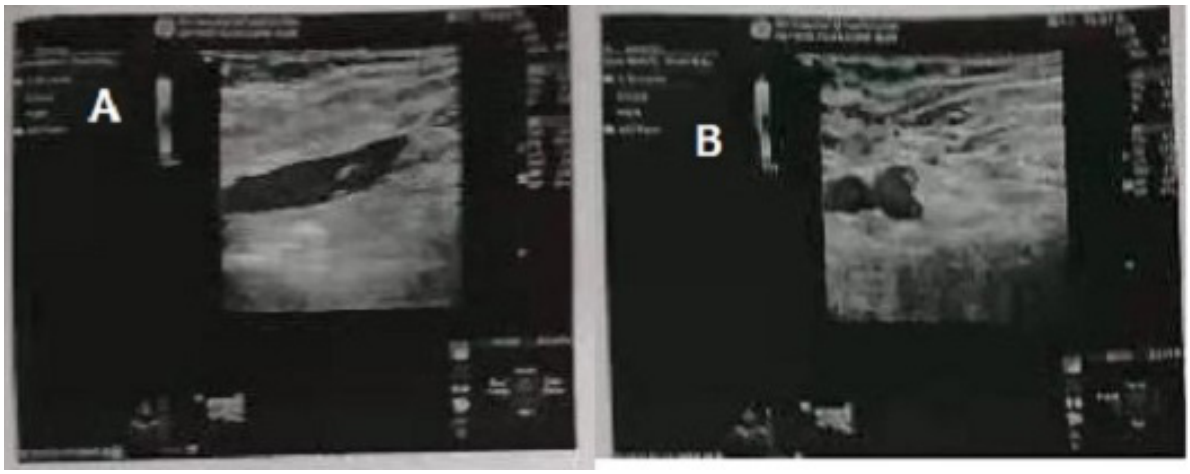

Figure 3: Lower limbs No fresh thrombosis.

Discussion

The incidence of habitual abortion among women with PAPS in women of childbearing age who has been diagnosed is as high as 60% [1]. It is extremely rare for women of childbearing age with PAPS thrombocytopenia during the second pregnancy as the first symptom and had a history of thrombosis and without a history of habitual abortion. We thus summarize, in this paper, the clinical characteristics and histopathologic manifestations of this unusual PAPS case. PAPS is a multiple system clinical syndrome caused by antiphospholipid antibodies (aPL) that occurs alone, its common targets are cardiolipin antibodies, β2-GP1 and prothrombin. aPL profile is an important factor in determining the risk of thrombosis and obstetric events ,high-risk antiphospholipid antibody profile (aPL) is associated with thrombotic and obstetric greater risk of APS (Tektonidou et al, 2019) [1]. aPL leading to placental infarction and impaired maternal-fetal blood exchange promotes placental thrombosis [2], and may mediate inflammatory events in early pregnancy [3]. Among the anti-phospholipid antibodies, lupus anticoagulant is the most closely related to thrombosis [4], and the anti-β2-GP1 antibody plays a key role in platelet activation related to thrombocytopenia [5].

In this case, high level of APL supports the diagnose of APS. Negative antinuclear antibody profile demonstrated no evidence for secondary to other diseases such as rheumatoid arthritis (RA) and systemic lupus erythematosus (SLE), and she had no history of rheumatism supporting, and got the final diagnose of PAPS. Once the diagnosis of APS was established, an urgent cesarean section operated. Due to the increased risk of thrombosis during the puerperium, women who received a preventive dose of heparin during pregnancy should continue to use the same dose of heparin for 6 weeks after delivery. Using heparin is negatively correlated with the risk of obstetric complications [6]. Patients with thrombotic APS and thrombocytopenia should use oral anticoagulants (DOAC) with caution, especially considering their half-life period [7] .The DOAC anticoagulant effect lasts for more than 48 hours, LMWH should be considered. It is not recommended that patients taking LMWH undergo routine anti-Xa monitoring during pregnancy, unless under certain circumstances: <50 kg or >90 kg, or other complicated factors, such as kidney injury or recurrent venous thromboembolism (VTE) [8]. PAPS is easy to be missed clinically, Although auxiliary examinations such as ultrasound and angiography can find thrombosis and the existence of old thrombosis. Female patients of childbearing age with recurrent miscarriage or with thrombocytopenia and previous thrombosis also need screening for the disease. PT can be prolonged by LA induction ("Lupus anticoagulant-hypoprothrombinemia syndrome: report of two cases and review of the literature," 2015) [9], which has become an important clinical idea for the discovery of PAPS. Declaration of Conflicting Interests: This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Disclosure: None declared.

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