Case Report

DOI: 10.17354/ijss/2016/41

Systemic Lupus Erythematosus in Pregnancy with Secondary Anti-Phospholipid Antibody Syndrome

K Vishwanath[1], Sanjay Kulkarni[2], N G Ashwini[3], Jolly Anil John[4]

[1]Associate Professor, Department of Medicine, MS Ramaiah Medical College, Bengaluru, Karnataka, India, [2]Professor and Head, Department of Medicine, MS Ramaiah Medical College, Bengaluru, Karnataka, India, [3]Post-graduate Student, Department of Medicine, MS Ramaiah Medical College, Bengaluru, Karnataka, India, [4]Assistant Professor, Department of Medicine, MS Ramaiah Medical College, Bengaluru, Karnataka, India

Abstract

Systemic lupus erythematosus (SLE) is a multisystem autoimmune disease. The clinical spectrum of SLE ranges from benign to a very severe life-threatening illness. It is characterized by an autoantibody response to nuclear and cytoplasmic antigens. SLE can affect any organ system, but it mainly involves the skin, joints, kidneys, blood cells, and nervous systemwe are presenting this case of SLE with anti-ds DNA negative, coombs test negative with severe thrombocytopenia , with secondary APLAin a pregnant women. Thus we are presenting this case of SLEin a pregnant women with anti-ds DNA Negative, coombs test negative with severe thrombocytopenia ,with secondary APLAwhich is not usual.

Key words: Antiphospholipid antibodies syndrome, Lupus hematological flare, Pregnancy, Systemic lupus erythematosus

INTRODUCTION

Systemic lupus erythematosus (SLE) is a multisystem autoimmune disease, characterized by an autoantibody response to various nuclear and cytoplasmic antigens Systemic lupus erythematous affects women of childbearing age and is associated with higher and fetal and maternal risk compared to pregnancy in healthy women. Association of Antiphospholipid syndrome and unexplained fetal death has been explained in women who had two or more pregnancy losses.

CASE REPORT

A 19-year-old female in 23 weeks of gestation was referred to us by her obstetrician with a history of diffuse petechial rashes, easy fatigability, occasional epistaxis, with the previous history of spontaneous pregnancy losses at 6 and 8 weeks, respectively, suspecting immune thrombocytopenia. At admission, her hemoglobin was 4.3 g%, platelet count of 2000. Obstetric scan confirmed a single live intrauterine fetus with fundic placental placement.

Lab parameters showed iron deficiency and Vitamin B12 deficiency anemia (serum iron = 22, total iron binding capacity = 404, transferrin saturation = 5.4%, serum ferritin = 7. 66 Vitamin B12 = 98) with severe thrombocytopenia. Peripheral smear picture showed microcytic hypochromic anemia with a moderate degree of anisopoikilocytosis; pencil shaped and tear drop cells with normal distribution and number of white blood cells with severe thrombocytopenia. Bone marrow study revealed norm cellular bone marrow aspirate with adequate cell trait, increase in a number of megakaryocytes.

Bone marrow aspirate however showed? Hypocellular with few islands of megakaryocytic series. ANA-IF was strongly positive (+++) with a speckled pattern. ANA profile was strongly positive (+++) for SS-A native (60 k Da), Ro - 52 recombinant, and positive (+) for SS-B. APA (phospholipid) –IgM was positive(titer =13.23 MPL U/ml). Titre of ds-DNA was negative in the ANA profile. Both indirect and direct coombs test was negative.

Access this article online

www.ijss-sn.com

Month of Submission : 11-2015
Month of Peer Review : 12-2015
Month of Acceptance : 01-2016
Month of Publishing : 01-2016

Corresponding Author: Dr. K Vishwanath, No 598, 2nd Main 2nd Stage E Block Rajajinagar, Bengaluru, Karnataka, India. Phone: +91-9632611288. E-mail: vishu_ani@yahoo.com

With the above presentation a diagnosis of SLE with? Secondary ANTI-PHOSPHOLIPID ANTIBODY SYNDROME, in G_3A_2 23 weeks ofpregnancy was made. Pulse therapy with high dose methylprednisolone was started. Improvement in her platelet count was seen. Immunologist opinion was sought and advised to continue methylprednisolone pulse therapy for total of 3 days.Therapy option with IV-Ig was also offered to the patient but was deferred in view of cost factor and unwillingness of the patient attenders. Rheumatologist opinion sought, advised to switch over to oral prednisolone at dose of 1 mg/kg body weight/day. A dose of 50 mg per day in 3 divided doses was started. Improvement in platelet count was maintained. Planned to taper the dose of prednisolone by 5 mg/week if target thrombocyte count above 1 lakh was maintained. Was also advised to start tab hydroxychloroquine 200 mg, po , bid dose.Low molecular weight heparin therapy was also started in view of APLA syndrome and the patient' s decision of continuation of pregnancy. Patient was discharged with advice to continue hydroxychloroquine, LWMH, prednisolone oral therapy with regular follow-up with weekly platelet count, regular ANC visits..

DISCUSSION

Systemic lupus erythematosus is an autoimmune connective-tissue disorder with a wide range of clinical features, which predominantly affects women[1] The clinical spectrum of SLE is wide and ranges from benign easily treated disease with rash, arthritis, fatigue, to a very severe life threatening illness with progressive irreversible damage. The course of the disease is variable and is characterized by flares of rampant inflammation that can threaten, in an unpredictable manner, almost any organ in the body Flare can be considered as a reappearance of clinical features which were earlier quiescent. Subtle abnormalities in hematological, cardiac and central nervous system (CNS) clinical, lab parameters and imaging occur either isolated or in combination which help to diagnose flare at an early stage Hematologic disease. In particular thrombocytopenia, is also commonduring pregnancy, with the risk ranging from 10% to 40% in different cohorts.Thrombocytopenia is a major haematological complication in patients with systemic lupus erythematosus (SLE) The pathogenesis of thrombocytopenia in SLE patients is heterogeneous, but the most common mechanism is believed to be increased platelet clearance mediated by anti-platelet autoantibodies, which is analogous to the mechanism seen in patients with idiopathic thrombocytopenic purpura (ITP). Other potential mechanisms include thrombotic thrombocytopenic purpura, disseminated intravascular coagulation, haemophagocytic syndrome, antiphospholipid syndrome and impaired thrombopoiesis3In this case as coombs test was negative, but thrombocytopenia was severe which was unusual as such severe thrombocytopenias are seen usually with lupus flare. Effect of pregnancy on SLE flares usually occur during the last half of pregnancy3 and within the first few weeks after delivery. In this case presented the hematologic manifestations of SLE occurred in first trimester and was associated with APLA positive disease. A few studies have shown decreased ds DNA levels during flare4 Even in this case anti ds DNA was negative but with coombs test negative and normal LDH a diagnosis of hematological flare was not possible.

CONCLUSION

Thus we are presenting this case of SLE in a pregnant women with anti-ds DNA NEGATIVE, coombs test negative with severe thrombocytopenia, with secondary APLA which is not usual.

ACKNOWLEDGMENT

Dr Chandrashekar, Immunologist, Chanrey rheumatology centre, Bangalore.

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How to cite this article: Vishwanath K, Kulkarni S, Ashwini, John JA. Systemic Lupus Erythematosus in Pregnancy with Secondary Anti-Phospholipid Antibody Syndrome. Int J Sci Stud 2016;3(10):193-194.

Source of Support: Nil, **Conflict of Interest:** None declared.