Case Report

Idiopathic thrombocytopenic purpura with Ebsteins Anamoly in pregnancy: A case report

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Abstract

We describe a pregnant patient with Ebsteins Anamoly with idiopathic thrombocytopenic purpura who responded to methyl prednisolone and intravenous immunoglobulin

Key words: Idiopathic thrombocytopenic purpura, Ebsteins Anamoly, methyl prednisolone, Intravenous immunoglobulin.

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INTRODUCTION

Idiopathic thrombocytopenic purpura is defined as isolated low platelet count [thrombocytopenia] with normal bone marrow and the absence of other causes of thrombocytopenia. It causes a characteristic purpuric rash and an increased tendency to bleed. Ebsteins Anamoly is a congenital heart defect in which the septal and posterior leaflets of tricuspid valve are displaced towards the apex of the right ventricle of the heart. We report here a case of pregnant woman with ITP with Ebsteins Anamoly who posed a therapeutic challenge.

CASE REPORT

The patient was 22 year old G_2A_1 at 35 weeks of GA, who presented with history of purpuric rashes. On examination she was found to be moderately anaemic. She had generalised purpuric rashes and loud P2 and PSM in tricuspid area (prior h/o exertional breathlessness). She had no icterus or lymphadennopathy. The blood pressure was 140/90 mmHg. Fundal height was corresponding to POG. Her investigestions revealed Hb- 8.8g%, platelet count- 3000\c.mm, PT-13.2, INR-0.89, peripheral smear- microcytic hypochromic anemia with neutrophilic leucocytosis with thrombocytopenia. Kidney and liver function tests were within normal limits. Antiplatelet antibody sent was positive. 2Decho showed Ebsteins anamoly with severe TR, LVEF- 65%, tricuspid valve is 1.5 cm below mitral valve. Obstetric ultrasonography showed SLIUF of 33-34 wks GA, EFW - 2282 g. 4 platelet transfused. Inj dexamethasone 8 mg IV twice a day was started. T. aldactone 25mg OD and T lasix 40mg 0-1/2-0 started. IVIg 1gm/kg over 6-8 hrs given. There was no much improvement in platelet count. Inj Dexamethasone was stopped. Inj methylprednisolone [solumedrol] 1 gm in 100ml NS over 1 hr for 3 days given. At 35 wks, she was admitted to the Obstetrics ward with increased BP recordings with eclampsia. Inj MgSO4 loading dose given. Inj labetalol 20 mg slow iv stat given. IE prophylaxis was started. Arrangments were made for emergency caesarean section inview of nonreassuring fetal heart rate. Platelet count then was 45,000|c.mm. Intraoperatively, there was no bleeding. Inj dexamethasone 8mg IV twice a day was continued postoperatively. Inj tranexemic acid was given. Postoperative period was uneventful.

DISCUSSION

Thromboctopenia complicates up to 10% of all pregnancies. Thrombocytopenia in ITP occurs because of

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platelet destruction mediated by platelet autoantibodies directed against cell surface antigens. The reticulo endothelial sysytem destroys platelet /antibody complexes. These autoantibodies can cross the placenta, thus both mother and new born can be affected. Intraoperative or intrapartum bleeding complications are unusal if the platelet count is greater than 50,000|µl. Due to their efficacy and cost, many consider corticosteroids to be first line treatment for ITP in pregnancy. The typical therapeutic dose of prednisolone is 1mg /kg/day which after achieving a response is gradually titrated to the lowest effective dose perhaps 20-30mg/day How ever since steroids may increase the incidence of pregnancy induced hypertension and exacerbate gestational diabetes, ASH guidelines consider IVIg (i.e., 1gm/kg once daily for 2 days) over 6-8 hrs to be an appropriate first line agent for severe thrombocytopenia or thrombocytopenic bleeding in third thrimester. A subset of patients who fail to respond satisfactorily to corticosterod or IVIg alone may respond to high doses of these agents when administered in combination (methyl prednisolone 1gm/kg, IVIg 1-2gm/kg). Splenectomy may be considered as another option for patients who fail to adequately respond to corticosteroids or IVIg and it is to be performed in the 2nd trimester as it has been associated with an increased incidence of premature labor when performed earlier and may be technically difficult later in gestation. Ebsteins anamoly is a rare heart defect that present at birth that may be associated with cyanosis and arrhythmias. The septal and posterior leaflets of the tricuspid valve are downwardly displaced into right ventricular cavity causing an abnormal function in the right ventricle. Pregnancy in women with Ebsteins anamoly is well tolerated. It is associated with an increased risk of prematurity, fetal loss and CHD in the offspring.

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