Original Research Paper

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Gynaecology

A CASE OF TROPICAL SPLENOMEGALY WITH HYPERSPLENISM AND PANCYTOPENIA IN PREGNANCY

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ABSTRACT Splenomegaly may be a clinical sign of many systemic diseases like Liver diseases , haematologic disorders, malignancies, AIDS, endocarditis, CCF, Parasitosis. Patients may be asymptomatic or may present with abdominal pain, fever, chest pain, back pain,early satiety, anaemia. In pregnancy it can pose a risk to mother and baby. In this case the lady presented in the first trimester with threatened miscarriage .On routine tests she was diagnosed as severe Thrombocytopenia with anaemia and leucopenia . After finding out all the causes of thrombocytopenia and opinion of the Haematologist, she was diagnosed as a case of Tropical splenomegaly with hypersplenism. The platelets were getting manufactured normally but there was accelerated destruction in the spleen which resulted in severe Thrombocytopenia. She was managed in liasion with haematologist in the antenatal period and delivered by caesarean section at 38 weeks for severe oligohydramnios with breech and previous section under general anaesthesia. Steroid administration did not help to raise the platelet count. She was operated under cover of single donor platelet infusion which was started at the incision.All the measures like using a cauterisation method while cutting the layers of abdomen, use of uterotonics to prevent extra blood loss were followed.she and the baby did well in the intra and post partum period.

KEYWORDS :

Introduction

Splenomegaly is one of the 4 cardinal signs of Hypersplenism. It includes reduction in the number of granulocytes, erythrocytes or platelets. It is associated with a complimentary proliferative response in the bone marrow and potential for correction of the abnormalities by splenectomy. Splenomegaly is associated with haemolytic anaemia where there is destruction of RBCs with increased workload on spleen. The incidence of splenomegaly is about 2-5%. Splenomegaly during pregnancy presents a high risk situation. The diagnosis becomes difficult in advanced gestation. There is always a risk of splenic rupture due to physiological and mechanical reasons.Hypersplenism associated with pancytopenia poses additional risk to the mother and fetus.

Classification based on the splenic size radiologically is
Normal less than 11 cms
Moderate 11 to 20 cms
Severe more than 20 cms

Causes of massive splenomegaly are
1.Leishmaniasis
2.CML
3.Myelofibrosis
4.Malaria

Causes of splenomegaly based on Pathophysiology
1. Congestive - By pooled blood as in Portal Hypertension
2. Infiltrative - Metastasis, Myeloid Neoplasms
3. Immune - Endocarditis, Sarcoidosis, Rheumatoid arthritis
4. Neoplastic - Lymphoma

This case was high risk as she was diagnosed with severe Thrombocytopenia in early pregnancy .She was to be carried till at least 37 weeks without any complications . she was at risk of bleeding episodes, antepartum haemorrhage, IUGR, severe anaemia with need for blood transfusion, IUD, need for operative delivery or caesarean section, Post partum haemorrhage, need for Hysterectomy .she did well throughout pregnancy and he intrapartum and postpartum period was uneventful.

Case history

A 24 year old second gravida with previous one caesarean section came to the OPD with bleeding per vaginum in early pregnancy. On routine blood investigation it was seen that her platelets were 40,000/cumm. She had no bleeding diathesis,easy bruisability, bleeding gums, petechiae or ecchymotic patches. First thought was that it was a spurious result, but on repeating the test it was found to be 36,000 / cu mm. It was alarming. There was no family history of bleeding tendency, and no such history in her previous pregnancy.There was no history of fever ,rash , chest pain, heart disease, liver disease , malaria, enteric fever.

Ultrasound was done for her which showed 8 weeks live pregnancy with massive splenomegaly.. There were multiple periportal collaterals, splenic hilar collaterals. It showed dilated portal vein cavernoma formation . LIver was normal in ultrasound .

On Examination

She was concious oriented afebrile . she was pale . There was no oedema or icterus. Her vital signs like pulse , blood pressure were normal.There was no evidence of ecchymotic patches, generalised lymphadenopathy. Respiratory and cardiovascular systems were normal . Liver was not palpable. Spleen was palpable more than half way to the umbilicus , but not below a line running horizontally through it.. This was grade 3 according to Hacket's classification.

Blood investigations showed

Haemoglobin of 8 gms/dl
TLC count of 3200/cumm
Serum bilirubin was 1.76mg/dl
Direct was 0.59mg/dl
Indirect was 1.17mg/dl
AST and ALT were in normal range
Coagulation profile was bit deranged with INR of 1.6
HB electrophoresis showed AA PATTERN
Malarial antigen and widal tests were negative

Urine routine examination was normal
Serum creatinine was 0.59mg/dl
Blood glucose was 75mg/dl
HBsAg was negative
HIV was non reactive
HCV wasnegative
ANA was negative
APLA was negative

Bone marrow study was done which was suggestive of normocellular bone marrow.It was suggestive of Hypersplenism.so the diagnosis was made as Tropical splenomegaly

She was informed of the condition and possible pros and cons were explained. She was managed in the antenatal clinic in liason with the haematologist. She was given Iron calcium folic acid and multivitamin supplements. she was regularly observed in the ANC clinic with monitoring of weight, blood pressure,haemoglobin levels, regular fetal ultrasound for fetal growth assessment. At 37 weeks she had breech presentation with oligohydramnios with AFI of 7. It was decided to do a elective caesarean section at 38 weeks. The indication was second gravida with previous caesarean section with breech presentation with hypersplenism and severe Thrombocytopenia. The case was discussed with Anaesthesiologist and Haematologist. She was given corticosteroids for achieving fetal lung maturity. It was

thought that steroids may help to raise the platelet counts . As there were no antiplatelet antibodies which were causing destruction of platelets, use of steroid was futile. The ultimate remedy for her was splenectomy.

She was operated under General anaesthesia. Neuraxial anaesthesia was having a risk of developing spinal haematoma as the safe level of platelets for spinal anaesthesia is 80, 000/ cumm. .At the time of incision Single donor platelet infusion was started. SDP is obtained from a single donor by apheresis. It is useful to treat acute haemorrhage secondary to Thrombocytopenia. She was given Joel Cohen incision .All the small vessels were fulgurated while entering the abdomen. A full term male child was delivered by breech. Baby cried well after birth. Placenta and membranes were delivered completely. She was given IV oxytocin 15 units in drip. Injection Methylergometrine 0.4 mg was given. Bleeding was well controlled. Abdomen was closed after complete haemostasis.

In the ward she had one episode of heavy bleeding with passage of big clots. There was no bleeding from the wound. She wa shifted to HDU and was given nasal oxygen as her SpO2 was dropping. , Inj methylergometrine was repeated and oxytocin was continued in the drip.Two units of Packed red cells was transfused. She was completely well after that and was discharged from the hospital on 5th postoperative day. At the time of discharge her platelet count was 50, 000/cumm.

She was followed in the Haematology clinic on 8th postoperative day and her platelets were 72000/cumm.

Discussion

TSS (tropical splenomegaly syndrome) is seen in malaria endemic area. Pregnancy is usually well tolerated in this condition. . There is a risk of spontaneous rupture of the enlarged spleen. TSS is an autoimmune phenomenon that follows episodes of malaria in the past . It is known as Hyperreactive malarial splenomegaly. The most common presentation is abdominal swelling, anaemia, thrombocytopenia. Spontaneous rupture of spleen occurs during acute infection and during primary attack. Pregnancy can worsen the anaemia, , can trigger haemolysis, more extramedullary haemopoiesis. , giving rise to enlargement of spleen and splenic rupture . Plasmodium vivax infection is commonly associated with this syndrome.

The criteria to diagnose tropical splenomegaly are

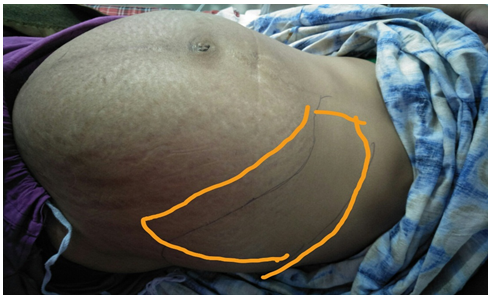
1. Residence in malarial endemic areas
2. Gross splenomegaly
3. Hypersplenism
4. Hepatic sinusoids with lymphocytosis
5. Presence of malarial antigen IgM more than 2SD above the mean.

In this case three criteria were fulfilled.

The exact assessment for TSS is difficult and condition should be differentiated from other causes of splenomegaly, like Leishmaniasis, schistosomiasis, Thalassemia, Leukemia, myelofibrosis, Non Tropical Idiopathic Splenomegaly, Felty's syndrome.. Genetic factors, Pregnancy and malnutrition are documented as predisposing factors. The exact mechanism is not clear but evidence suggests that exposure to malaria elicits exaggerated stimulation of polyclonal B lymphocytes leading to production of IgM. T cell infiltration of hepatic and splenic sinusoids accompanies this process. Serum cryoglobulin and Autoantibodies rise as does the high molecular weight immune complexes. The result is anaemia, deposition of large immune complexes in Kupffer cells in the spleen and LIver . Reticuloendothelial cells hyperplasia results in splenomegaly.They do well with Antimalarial Therapy which is given for prolonged period. Chloroquine and Proguanil are equally effective. Splenectomy has definitely earned a place as a therapeutic measure.

Conclusion

Despite the potential complications during pregnancy and delivery due to splenomegaly, the best mode of delivery is not yet established.In a recent prospective cohort study from the United Kingdom, the rate of caesarean delivery was 45% in those case with splenomegaly. In my case she was breech with previous caesarean section. So the caesarean was indicated irrespective of splenomegaly.There is a role of multidisciplinary management in this case .In our case she was managed with the haematologist and myself with multivitamins , folic acid and iron therapy..There was no role of steroids as there was no antibody induced destruction of platelets. She was not a case of Immune Thrombocytopenia or Thrombotic Thrombocytopenia. The only thing was excess bleeding at the time of caesarean section and splenic rupture. So she was given SDP at the time of surgery and platelet concentrate on the second postoperative day.sh edid well in the post partum period and the platelet counts were 72,000/cumm. Will she be benefited by just prolonged antimalarial therapy or splenectomy will be needed to normalise her haematological parameters is the question to be solved.



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