CASE REPORT

MASSIVE SPLENOMEGALY IN PREGNANCY: A REVIEW OF THREE CASES

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ABSTRACT: BACKGROUND: Splenomegaly with an incidence of about 2-5% is a challenging medical problem. Splenomegaly during pregnancy presents a further testing situation for the obstetrician. The diagnosis especially during advanced gestation becomes difficult by clinical examination. Further the risk of splenic rupture may increase in pregnancy due to several physiological and mechanical reasons. It is noteworthy that anemia which is a frequent association with splenomegaly is also an important medical condition of pregnancy.

CASE REPORT: We report three cases of massive splenomegaly with pregnancy (all with different etiology and presentation) first with severe anemia, second with dragging pain and third with recurrent fever and its adverse effects on pregnancy outcomes. Though none of our patient had the life threatening splenic rupture but we found association of massive splenomegaly with fetal growth restriction and aggravation of preexisting anemia requiring blood components. To the best of our knowledge, there are no published reports indicating adverse effects of massive splenomegaly on pregnancy outcome.

CONCLUSION: Splenomegaly with pregnancy should be considered as a high risk pregnancy and it is to be dealt under the supervision of senior obstetrician in a tertiary care centre because it can complicate the maternal and fetal outcome.

INTRODUCTION: Splenomegaly with an incidence of about 2-5 % [1] is a challenging medical problem. The causes of massive splenomegaly (spleen more than 8cms below the costal margin) are myriad ranging from chronic infectious and inflammatory conditions to hematological diseases. Outcome can be good in cases of infectious pathology that have been adequately treated in contrast to chronic conditions like hematological malignancies which tend to have a poor outcome. Complications relate to underlying cause responsible for splenomegaly. Spontaneous rupture of an enlarged spleen though rare can be catastrophic [2].

Splenomegaly during pregnancy presents a further testing situation for the obstetrician. The diagnosis especially during advanced gestation becomes difficult by clinical examination. Further the risk of splenic rupture may increase in pregnancy due to several physiological and mechanical reasons [2]. It is noteworthy that anemia which is a frequent association with splenomegaly is also an important medical condition of pregnancy. It becomes all the more important for the treating obstetrician to actively look for splenomegaly in such cases.

This report consists of 3 cases of massive splenomegaly in pregnancy, all presented differently and posed a diagnostic challenge to us- first presented as severe anemia, second as chronic pain abdomen and the third as recurrent fever.

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CASE 1: 26years old primigravida at 34 weeks gestation presented with severe anaemia, history of fever off and on since 12 days, history of 2 units of whole blood transfusion and antimalarial treatment one week back. No history of blood transfusion prior to pregnancy. On examination: pallor + clinically 4-5g%, chest examination was normal, cardiovascular system revealed hyperdynamic flow murmur, uterus was enlarged to 34 weeks with singleton live fetus in cephalic presentation and an enlarged spleen felt of 12cm below the costal margin, Liver was also enlarged 4cm below costal margin, slightly tender.

On investigation Hb was 4g/dl, TLC was 7400/mm³, DLC was P66/L30/M1/E3, Platelets was 1,60,000/mm³, Peripheral smear was suggestive of hemolysis with erythroblast 28%, severely microcytic hypochromic, basophilic stippling and polychromasia, Iron studies were normal (Serum Iron of 100mg%, TIBC of 280mg%, and S. Ferritin of 120mg/dl) but reticulocytosis was present with a count of 15%, LFT was also suggestive of hemolysis with Serum bilirubin Total-2.1mg/dl, Direct-1.3mg/dl, Indirect-0.8, ALT=39u/l, AST-19u/l, ALP-121, LDH was also raised to 1518 IU suggesting hemolysis, Coagulation profile was normal. Malaria and leishmaniasis were also ruled out (MP Smear and malarial antigen - negative, antibodies for leishmaniasis negative). USG revealed mild hepatomegaly, a 22.7cm enlarged spleen and 34weeks normal fetus with normal Doppler findings. Since the above results ruled out the common causes of splenomegaly like leukemia, myeloproliferative disorders, and chronic infections, Hb electrophoresis was done which revealed HbE disease with Beta thalassemia minor. Pt was given 4 units of packed cells over 3 days and Hb increased to 9gms/dl. A course of steroids were given for fetal lung maturity. At 37completed weeks her cesarean section was done in view of contracted pelvis and massive splenomegaly. Girl child of 2.25kg with apgar of 8/10, 9/10 was delivered and post op period was uneventful. At present her anemia has improved and both mother and baby are doing well.

CASE 2: 22years primigravida a 26 weeks pregnancy admitted with dragging pain at left hypochondrium and left lumbar region, fever, no h/o trauma, LPV or BPV present. On examination: uterus was enlarged till 26weeks with singleton live fetus and an enlarged spleen of 12cm below costal margin. Investigations showed hemogram suggestive of pancytopenia (Hb=7.4g/dl, TLC=3800/mm³and Plt=21000/mm³), RBC's on Peripheral smear were microcytic hypochromic; ANA's, anti phospholipid antibodies and RA factor were negative ruling out autoimmune hemolytic anemia, Hb Electrophoresis was also normal, USG whole abdomen suggested no hepatomegaly but with multiple intrahepatic, pericholecystic, peripancreatic and splenic collaterals, a splenomegaly of 22.5cm and a single live fetus of 26weeks. Upper GI Endoscopy revealed no varices. With above positive findings a diagnosis of Extrahepatic Portal Hypertension was made. Medical and Hematological consultations suggested conservative management and active intervention in form of platelet transfusion only in case of bleeding. Patient was discharged and advised regular ANC. She was advised to avoid any trauma on abdomen and report immediately in case of acute pain abdomen. Patient was readmitted at 36weeks in view of FGR and oligohydramnios and her cesarean section was planned. She was given pooled platelets just before cesarean section as her platelets were low, she had boy baby of 2.1 kg with Apgar score of 8/10, 9/10. Her intra-op and post op period was uneventful. Both mother and baby are fine at present.

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CASE 3: 22years unbooked primigravida at term came with labour pains with history of multiple episodes of high grade fever with chills in the past, which had been diagnosed as malaria. On examination 36 weeks uterus was felt with regular uterine contraction, an enlarged spleen of 10cm below costal margin and liver 3cm below costal margin was also palpated. She was in active labour (5 cm dilated, 90% effaced, vertex at 0 station). Decision for normal vaginal delivery under the supervision of senior obstetrician was taken with special care to avoid trauma to spleen. She delivered a 2.2 kg girl baby with Apgar of 8 and 9, no PPH occurred, her post-natal period was uneventful. Her Hb was 8g/dl, TLC= 4500/ mm³, Platelets = 1lakh/mm³. Peripheral smear revealed normocytic hypochromic picture. Hb electrophoresis normal ruling out hemoglobinopathies. Although MP smear was negative but malarial antibodies were positive pointing towards the diagnosis of tropical splenomegaly syndrome or hyperreactive malarial splenomegaly. LFT and KFT were normal, ANA and RA factor done which were negative to rule out autoimmune anemia. This patient could not be followed as she left against medical advice.

DISCUSSION: All our three cases had anemia and FGR and to the best of our knowledge, there are no published reports indicating effects of massive splenomegaly on pregnancy outcome.

DOUBLE HETEROZYGOUS HBE/B THAL: is a rare condition. Hemoglobin E (HbE) is variant hemoglobin with a mutation in β globin chain causing substitution of glutamic acid for lysine at position 26 of β globin chain. HbE is the second common abnormal gene after sickle cell hemoglobin (HbS). HbE disease alone is usually asymptomatic and goes unnoticed. Hb E/ β presents variably with clinical features ranging from thalassemia trait to major requiring blood transfusions. Less severely affected patients usually have splenomegaly, mild anaemia but do not require transfusions. Some patients may manifest as severe anemia first time during pregnancy as was seen in our case. Frequent blood transfusions during pregnancy are required for good maternal and fetal outcome.

EXTRA HEPATIC PORTAL HYPERTENSION: has varied causes congestive, infections, storage disorders, myeloproliferative disorders, hemolytic and anatomical. Acute uncomplicated obstruction of the portal vein presents with sudden onset of ascites, which tends to resolve spontaneously as collateral circulation develops to bypass the block. Splenomegaly is a very common feature and the physical discomfort of this large organ is the only significant clinical complaint. Liver biochemical changes, when present, are trivial but hypersplenism is common and often severe enough to lead to pronounced anaemia. Despite the thrombocytopenia associated with hypersplenism bleeding problems rarely occur in these patients because the platelets are functionally normal. Hemorrhage from esophageal varices in 40% of the cases, is the most common complication which usually occurs in the second and third trimester due to the physiological changes in the circulatory system during pregnancy temporary aggravating the portal hypertension [3]. Treatment is usually conservative, unless too much discomfort and severe anemia warrants splenectomy. Varices to be treated only if symptomatic. Blood products may be required during delivery. Pregnancy is usually well tolerated.

TROPICAL SPLENOMEGALY SYNDROME (TTS): is seen in people living in the malaria endemic area. The most common presentations of TSS are dragging sensation, abdominal swelling, anemia, and hepatomegaly. Spontaneous rupture of the spleen occurs almost exclusively during acute

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infection and usually during the primary attack [3]. *P. vivax* is the species most closely associated with the rupture of spleen [4]. Certain criteria are set to diagnose patients with hyperreactive malarial syndrome (HMS). These criteria includes, residence in malaria endemic area, gross splenomegaly 10cm or more, elevated serum IgM level two standard deviations or more, hypersplenism, clinical and immunologic responses to antimalarial therapy, hepatic sinusoidal lymphocytosis in liver biopsy[5]. The first four criteria were documented in our patient. Medical management with antimalarial drugs for prolonged period is the mainstay. Response to therapy is guided by the splenic size and symptomatic improvement. There is limited data warranting splenectomy as choice of treatment for tropical splenomegaly syndrome / hyperreactive malarial syndrome.

Pregnancy can increase the risk of spontaneous rupture of the enlarged spleen. Firstly pregnancy can worsen preexisting anemia and also trigger red blood cell hemolysis which in turn induces massive extramedullary hematopoiesis giving rise to increase in splenic size and finally splenic rupture [2]. Secondly mechanical factors like reduction in volume of peritoneal cavity and uterine contractions during pregnancy which can cause compression of the diaphragm predisposes to splenic trauma [2]. Thirdly frequent abdominal examinations and manipulation during labour can also give rise to splenic trauma. To prevent this dreaded complication, we did an elective LSCS in the first two cases reported by us. The third case already came in active labour but we took great care to avoid any abdominal trauma and closely monitored the patient during labour and after delivery. Fatal splenic rupture without any history of trauma has been reported in a pregnant woman with HbC/ β thal by Boldorini et al. Various pathogenic mechanisms have been suggested for this increased risk of splenic rupture.

Regarding the fetal prognosis in cases of massive splenomegaly, it was observed that all the three cases reported by us had growth restricted fetuses. To the best of our knowledge there are no published case reports which also reiterate the above observation. The mechanism of IUGR could be due to limited space in the intraperitoneal cavity for the uterine enlargement and also because of associated chronic anemia. Further studies in this direction will give a clearer insight into this problem.

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1. Massive Spleen
2. Uterus
3. Umbilical Cord

Fig A: Showing per-op finding of case -1 just after the delivery of fetus, with umbilical cord still in situ.

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