Short Report

Pregnancy with massive splenomegaly: A case series

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ABSTRACT

Background. Pregnancy with massive splenomegaly is a rare entity and is associated with increased risk to both mother and foetus. There is paucity of studies in the literature to guide clinicians for the management of this condition.

Methods. We reviewed the course of pregnancy, maternal and foetal outcomes of 5 pregnant women with massive splenomegaly who were managed in our unit during 2015–16.

Results. All 5 women had anaemia and thrombocytopenia, and had different causes for splenomegaly. One patient had chronic malaria, 2 had portal hypertension with cirrhosis and the remaining 2 had non-cirrhotic portal hypertension. Life-threatening complications were present in 2 patients; one of them had severe pre-eclampsia complicated by pulmonary oedema, cardiac arrest and the other patient developed spontaneous bacterial peritonitis. Intrauterine growth restriction and meconium-stained liquor were the most common perinatal complications. Two patients had vaginal delivery and 3 required emergency caesarean section. Postpartum haemorrhage was present in 2, and the hospital stay was prolonged in all the patients. All mothers and babies were discharged in a satisfactory condition.

Conclusion. Pregnancy with massive splenomegaly poses a challenge because of diverse aetiology and potentially adverse outcomes. Multidisciplinary care in a tertiary centre can help optimize the outcome.

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INTRODUCTION

Pregnancy with massive splenomegaly is a rare entity and is complicated by anaemia, thrombocytopenia, ascites and jaundice.[1] There is an increased risk of complications for the mother and foetus. Anticipation of risks and lack of sufficient studies to guide the management lead to anxiety in both the treating obstetrician and the patient.

We managed 5 pregnant women with massive splenomegaly over a period of 12 months. Many complications were encountered during the pregnancy, but we were able to achieve successful outcomes with a multidisciplinary approach involving the obstetrician, gastroenterologist, physician, intensivist and neonatologist. We present our experience of pregnancy with massive splenomegaly leading to successful maternal and foetal outcomes.

METHODS

We reviewed the course and outcome of 5 pregnant women with massive splenomegaly who were managed during 2015–16 at the Department of Obstetrics and Gynaecology, All India Institute of Medical Sciences, New Delhi. Massive splenomegaly was defined as a spleen with craniocaudal length measuring 17 cm or more on ultrasonography.[2] Patients' characteristics including age, history (fever, thrombotic episodes, gastrointestinal haemorrhage), and treatment history were recorded. Laboratory investigations (complete blood counts, liver function tests and viral markers including hepatitis B surface antigen and anti-hepatitis C virus); ultrasonography with colour Doppler (to assess the echotexture of the liver, and measure the spleen size, liver span and diameter of the portal vein), upper gastrointestinal (GI) endoscopy; autoimmune work-up (lupus anticoagulant and antiphospholipid antibody) and work-up for infections (RK-39, peripheral smear for malaria parasite) was done as indicated. Treatment was given to all the patients after consulting a physician/gastroenterologist. Foetal surveillance was done with fortnightly growth monitoring, biweekly biophysical profile, and weekly umbilical artery Doppler indices. The course of pregnancy, mode of delivery, and maternal and foetal outcome were noted.

RESULTS

During the study period, a total of 903 high-risk women delivered in our unit, of which 5 pregnant women presented with massive splenomegaly (Table I). Their ages ranged from 21 to 24 years. Three of them presented in late trimester and splenomegaly was diagnosed incidentally during routine obstetric examination. One had a history of recurrent malaria (case 1). Two patients (case 2 and case 3) had a history of recurrent jaundice and fever in the past 2–3 years and were diagnosed with portal hypertension with cirrhosis during the pregnancy.

The other two patients (case 4 and case 5) were diagnosed with portal hypertension due to non-cirrhotic portal fibrosis (NCPF) at 13 and 17 years of age as they had recurrent episodes of haematemesis. One of them had a history of variceal banding 15 times in the past 5 years and also had coeliac disease.

All the patients had anaemia (haemoglobin 6.7 g/dl to 10.5 g/dl) and thrombocytopenia (platelet count 30 000 to 50 000 cells/cmm) and 4 had leucopenia (2400–3100 cells/cmm). Peripheral blood smear showed normocytic, normochromic anaemia. Liver function tests were within normal limit in four patients, but one patient with cirrhosis had serum bilirubin of 6.1 mg/dl with marginally raised liver enzymes (alanine amino transferase 52 IU/L, aspartate amino transferase 44 IU/L, alkaline phosphatase 413 IU/L). Coagulation profile (prothrombin time, activated partial thromboplastin) was normal, viral markers for hepatitis, infective and autoimmune work-up was negative in all

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TABLE I. Characteristics and outcome of cases

Item	Case 1	Case 2	Case 3	Case 4	Case 5
Parity	Primi	Primi	Primi	G2P0L0A1	Primi
Aetiology	Malaria	Cirrhosis	Cirrhosis	NCPF	NCPF
Size of spleen (cm)	21	18	17	23	24
Onset of labour	Spontaneous	Induced with prostaglandin gel	Induced with prostaglandin gel	—	Induced with prostaglandin gel
Gestational age at delivery (weeks)	39^{+4}	39^{+3}	39^{+4}	32^{+4}	37^{+3}
Mode of delivery	Vaginal	Emergency LSCS	Vaginal	Emergency LSCS	Emergency LSCS
Birth weight (g)	2615	2282	2484	1800	2391
Apgar score	9/9	8/8	9/9	3/5	9/9
Transfusion	8 PRP	4 PRP, 1 SDP	2 SDP, 2 PRBC	2 PRBC, 4 FFP, 4 PRP	2 PRBC, 2 SDP
Complications	None	IUGR, MSL, fever, SBP, pleural effusion	SFD, atonic PPH (balloon tamponade)	Jaundice, severe pre-eclampsia, pulmonary oedema, cardiac arrest, prematurity	IUGR, oligoamnios, MSL, atonic PPH
Duration of hospital stay (days)	50	112	13	35	12

NCPF non-cirrhotic portal fibrosis LSCS lower segment caesarean section PRP platelet-rich plasma SDP single donor platelet PRBC packed red blood cell FFP fresh frozen plasma IUGR intrauterine growth restriction MSL meconium-stained liquor SBP spontaneous bacterial peritonitis SFD small for date PPH postpartum haemorrhage

the patients. The size of spleen ranged from 17 cm to 23 cm on ultrasonography. The liver appeared to be cirrhotic in two patients. Two patients had gross ascites (Table I). Upper GI endoscopy in the patients with NCPF showed residual non-bandable varices in one patient and grade 2 varices in the other. None of them had variceal bleeding during pregnancy. Patients were hospitalized at the gestational age of 28–32 weeks for maternal and foetal surveillance and supportive management. One patient with cirrhosis and portal hypertension had massive ascites and required drainage for symptomatic relief.

Intrauterine foetal growth restriction (IUGR), small for date, prematurity and meconium-stained liquor (MSL) were the most common perinatal complications. One patient developed severe pre-eclampsia and pulmonary oedema. She had a cardiac arrest at 32^{+4} weeks of gestation. However, she was revived and had an emergency lower segment caesarean section (LSCS) due to severe foetal bradycardia. She was managed in the intensive care unit with ventilatory support for 4 days and supportive therapy thereafter. The other 4 patients were planned for a vaginal delivery. The sole indication for induction of labour was IUGR at term. Intrapartum electronic foetal monitoring was done for all the patients. Two patients had an emergency LSCS done for MSL and foetal bradycardia. Platelet counts were maintained above 50 000 cells/cmm during labour with transfusion of single donor platelets and platelet-rich plasma. Atonic postpartum haemorrhage (PPH) occurred in two patients and was managed with medical therapy, one patient also required balloon tamponade. The mean birth weight was 2.3 kg (range 1.8–2.6 kg). One patient who had portal hypertension and cirrhosis with massive ascites developed unexplained high-grade fever and tenderness over the abdomen 20 days postpartum. She was diagnosed with spontaneous bacterial peritonitis (SBP) and was managed with injectable ceftriaxone for 7 days. The hospital stay was prolonged in all the patients (range 13–112 days). All the mothers and babies were discharged in a satisfactory condition.

DISCUSSION

Pregnancy with massive splenomegaly poses a unique challenge to obstetricians because physiological changes of pregnancy and underlying pathology both contribute to deterioration of the haemodynamic status.[3] The common causes of massive splenomegaly seen in pregnancy are haemolytic anaemia, thalassaemia, myelofibrosis, malaria, kala azar, tuberculosis, portal hypertension (cirrhosis, extrahepatic portal vein obstruction, non-cirrhotic portal fibrosis) and collagen disorders (systemic lupus erythematosus).[4] Three of the patients in our series were diagnosed incidentally during pregnancy despite having massive splenomegaly, probably because examination during pregnancy was their first interface with specialized healthcare services. This emphasizes the importance of a thorough systemic examination of pregnant women by obstetricians on their first antenatal visit.

Hyper-reactive malarial splenomegaly or tropical splenomegaly syndrome is a common cause of splenic enlargement in endemic areas. This is associated with increased incidence of anaemia, thrombocytopenia and low birth weight as seen in case 1. There is increased risk of infections and splenic rupture. Treatment consists of a prolonged course of effective antimalarial therapy that leads to reduction in splenic mass, but there is always an increased apprehension of teratogenicity with most antimalarial drugs. In a retrospective study, pregnant women were given mefloquine 250 mg weekly for a median of 9 weeks (range 2–25 weeks) and a reduction in the size of spleen along with fall in antimalarial antibody titres were observed.[5] The Center for Disease Control and Prevention has recommended mefloquine for prevention and treatment of malaria in pregnant women with no increase in the risk of teratogenicity.[6] Our patient (case 1) was also given mefloquine 250 mg weekly for 12 weeks. On follow-up, a reduction in the size of spleen was observed after 6 months. To reduce the risk of rupture, it is advisable to palpate the abdomen carefully and avoid any fundal pressure during labour.

Pregnancy is rarely seen in women with cirrhosis because of associated endocrine abnormalities.[7,8] Maternal deterioration occurs in 30%–50% of these cases because of variceal haemorrhage, hepatic decompensation, ascites, SBP, PPH and splenic artery aneurysm rupture.[9] Although none of our patients with portal hypertension and cirrhosis had variceal bleeding or hepatic encephalopathy, we encountered two life-threatening complications, namely, SBP and atonic PPH. SBP is rare and occurs in patients with long-standing cirrhosis.[10] There is also increased risk of abortions (15%–30%), prematurity (25%), MSL,

foetal distress and still births.[9,11] Both these patients had IUGR, one of them also had MSL during labour and required a caesarean section.

NCPF is more commonly reported from eastern, underdeveloped countries.[12] The aetiopathogenesis is not yet known and autoimmunity, infections, prothrombotic states and antiretroviral drugs have been implicated as its causative factors.[13] One of our patients had underlying coeliac disease suggesting the possibility of an immunological association. These patients are usually diagnosed at a younger age because of repeated episodes of variceal bleeding, splenomegaly, ascites and thrombocytopenia similar to our patients. The prognosis of pregnant women with NCPF is relatively better than those with cirrhosis because of preserved liver functions and lower complication rates.[14] However, our patients with NCPF had life-threatening complications, namely, pulmonary oedema, cardiac arrest in one, and atonic PPH in the other.

The mode of delivery in women with massive splenomegaly remains a concern for obstetricians. There is a risk of variceal bleeding because of repetitive Valsalva manoeuvre[15] and hence some experts advise an elective caesarean section in those with high-grade varices.[12,14] However, the risk of bleeding from dilated collaterals over pelvic or abdominal wall during or after caesarean section is also reported and therefore decision regarding the mode of delivery should be individualized.[14] Vaginal delivery is advised with epidural analgesia along with the use of ventouse or forceps to cut short the second stage of labour. Caesarean section should be done only for obstetric indications.[16] Epidural analgesia is useful as it can be continued in case caesarean section is required and general anaesthesia should be avoided because of higher risk of encephalopathy.[16,17] We used epidural anaesthesia after correction of thrombocytopenia in two of our patients and general anaesthesia was given to only one patient who was operated after cardiopulmonary resuscitation. Platelet counts were maintained at more than 50 000 cells/cmm during labour and caesarean section.[3] Active management of the third stage of pregnancy was followed for all the patients, but 2 of 5 patients had atonic PPH. A possible explanation for this higher incidence of PPH could be associated coagulopathy, anaemia and thrombocytopenia.[9] For management of PPH, we preferred oxytocin over methyl ergometrine because of an increased risk of variceal bleed with the latter.[9]

Conclusion

Massive splenomegaly in pregnancy is rare and obstetricians should be aware of the aetiology and modalities for management. These women can have life-threatening complications throughout pregnancy and also during labour and postpartum period. The foetus is also at risk of increased perinatal morbidity and mortality. Therefore, pregnant women with massive splenomegaly should be managed at tertiary care hospitals which have multidisciplinary teams.

Conflicts of interest. None declared

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