CASE REPORT

Combined caesarean with splenectomy in pregnancy with portal hypertension: defining plausibility

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SUMMARY

24-year-old woman at 28 weeks gestation was referred from peripheral hospital with diagnosis of pregnancy with portal hypertension. She had received multiple transfusion for pancytopenia in the past and had undergone endoscopic sclerotherapy for oesophageal varices. Initially, she was admitted in our hospital at 28 weeks gestation for blood transfusion and was evaluated by multispecialty team of doctors. She was advised splenectomy for transfusion-dependent pancytopenia secondary to hypersplenism in non-cirrhotic portal hypertension. She was readmitted at 36 weeks gestation. A decision for caesarean was taken owing to failed induction of labour at 38 weeks gestation. She underwent combined caesarean with splenectomy. Mother and child had an uneventful postoperative recovery and were discharged on ninth postoperative day. Preconceptional counselling, treatment of oesophageal varices and multispecialty approach was paramount in the management. Combined caesarean with splenectomy is feasible and cost-effective treatment associated with improved quality of life. Prospective clinical trials are essential to prove safety and efficacy of treatment.

BACKGROUND

Pregnancy is not uncommon in women with non-cirrhotic portal hypertension (NCPH).¹ In India portal hypertension (PH) is more commonly due to non-cirrhotic portal fibrosis (NCPF) and extrahepatic portal vein obstruction (EHPVO).² Prevalence of NCPF in the world and in India is 3%–5% and 15%–20%, respectively.³ In-hospital incidence of pregnancy with NCPH in India is 0.09%.³ PH increases the risk of life-threatening bleeding during pregnancy.³ The overall risk of variceal bleeding is almost 400 times greater in pregnancy with PH than in pregnant women without PH.³ NCPF when compared with EHPVO is associated with significant increase in risk of abortion, prematurity, intrauterine growth retardation, preterm birth, still births and increase in the incidence of caesarean delivery.^{4–6} Splenectomy during pregnancy has been reported in pregnant mothers with refractory immune thrombocytopenic purpura, ruptured splenic artery aneurysm and splenic trauma. However, there is no documentation of combined caesarean with splenectomy in pregnancy with PH in English literature. We adopted a multispecialty approach as pregnancy with PH is associated with increased morbidity and mortality when compared

with general population. In this case study, we will be discussing an approach to pregnancy with PH and why we combined caesarean with splenectomy in pregnancy with PH.

CASE PRESENTATION

A 24-year-old woman at 28 weeks gestation was referred from peripheral hospital with complaints of generalised weakness and fatigue since 6 years. She also complained of dull aching pain in left upper abdomen since 3 years, and decreased appetite with early satiety since 3 years. She had an episode of haematemesis about 3 years ago and was admitted in peripheral hospital. Patient received blood transfusion for pancytopenia and had undergone endoscopic sclerotherapy for bleeding oesophageal varices. No history of taking medicines like propranolol or nitrates in the past. On examination she was pale, icteric and haemodynamically stable. Her body mass index was 18.3 (kg/m²). On examination of abdomen, spleen was palpable 15 cm below the left costal cartilage and liver span was 12 cm. Uterus was corresponding to 28 weeks period of gestation.

INVESTIGATIONS

See [table 1](#) for investigation. Our diagnosis was Rh-negative pregnancy with PH in a known case of pancytopenia due to hypersplenism and NCPH.

DIFFERENTIAL DIAGNOSIS

Cirrhosis of liver, extrahepatic portal venous obstruction, tropical splenomegaly.

TREATMENT

Upper gastrointestinal endoscopy (UGIE) was performed during her initial admission at 28 weeks gestation that showed grade 2 oesophageal varices. She received blood transfusion for pancytopenia and was evaluated by multispecialty team of doctors (viz, obstetrician, general surgeons, gastroenterologists, haematologists and radiologist) prior to her discharge. She was advised splenectomy for transfusion-dependent pancytopenia that was secondary to hypersplenism in NCPH. She was readmitted at 36 weeks gestation. Fetal cardiotocography and ultrasonography revealed that the fetus was normal and concordant with gestational age. She was placed on serial fetal monitoring at 38 weeks gestation and her blood parameters were regularly monitored. As she complained of decreased fetal movements during a period of

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Table 1 Investigation report at 36 weeks of pregnancy

Tests	Report	Interpretation
Blood group	B-negative	► Rh-negative pregnancy
Haemoglobin	8.7 gm%	► Pancytopenia
Total white blood cells (WBC)	$2.2 \times 10^9/L$	
Platelet count	$20 \times 10^9/L$	
Total bilirubin	3.2 mg%	► Unconjugated hyperbilirubinaemia
Unconjugated bilirubin	2.8 mg%	
Conjugated bilirubin	0.4 mg%	
Aspartate transaminase	38 U	
Alanine transaminase	12 U	
Alkaline phosphatase	138 U	
Prothrombin time	13.3	
International normalised ratio	1.1	
Peripheral blood smear:	<ul style="list-style-type: none"> ► Microcytic hypochromic anaemia ► WBC: 25 cells ► Platelets: 20 000 ► Reticulocyte count: 4.5% ► No haemoparasites seen 	► Iron-deficiency anaemia
Haemoglobin (Hb) Electrophoresis	<ul style="list-style-type: none"> ► Hb : 8.7 ► Hb A: 96.2% (94-98) ► Hb A2: 2.4% ► Hb F: 0.8% 	<ul style="list-style-type: none"> ► Suggestive of a normal pattern ► No evidence of abnormal haemoglobin peaks
Viral tests	HIV Negative Hepatitis B virus (HBsAg) Negative Hepatitis C virus (HCV-Ab) Negative Syphilis Negative	Negative
Ultrasonography abdomen	<ul style="list-style-type: none"> ► Dilated portal vein (20 mm) with multiple collaterals ► Chronic portal vein thrombosis with recanalisation ► Spleen enlarged (22 cm) with dilated splenic vein (15 mm) ► Normal echotexture liver ► Single live intrauterine fetus corresponding to 28 weeks of gestation 	<ul style="list-style-type: none"> ► Hypersplenism ► Non-cirrhotic portal hypertension ► Normal viable fetus

HBsAg, hepatitis B surface antigen; HCV-Ab, hepatitis C antibody.

continuous monitoring, labour was induced thrice with dinoprostone 0.5 mg. As the patient did not respond to induction of labour, an immediate plan for emergency caesarean section was taken. As the patient requested for combined caesarean with splenectomy, a written and informed consent was obtained after counselling about risks and benefits of surgery. Adequate units of blood were arranged. A linear midline laparotomy incision was placed between xiphoid process and pubic symphysis ([figure 1](#)) and a male baby weighing 2.5 kg was delivered. Splenectomy was performed and liver biopsy was taken to rule out cirrhosis of liver. Abdominal incision was approximated after achieving adequate haemostasis. Surgery was performed under general

anaesthesia ([video 1](#)) that lasted for 3 hours with 400 mL intra-operative blood loss. The maternal heart rate and blood pressure remained within normal range during the entire procedure.

OUTCOME AND FOLLOW-UP

Both mother and child had uneventful postoperative recovery. Baby had no congenital anomaly. On postoperative day (POD)–1: Haemoglobin was 11.5 gm%, white blood cell count was $22.2 \times 10^9/L$, and platelet cell count was $191 \times 10^9/L$. On POD-3 patient developed thrombocytosis with platelet count of $1200 \times 10^9/L$. Therefore, tablet aspirin 75 mg once a day was

Figure 1 Midline laparotomy incision extending from xiphoid process to the pubic symphysis for combined caesarean with splenectomy.

Video 1 Combined caesarean with splenectomy surgical procedure in pregnancy with portal hypertension

Figure 2 Suggested management algorithm for pregnancy with portal hypertension.

prescribed. Triple vaccine (vaccination against *Haemophilus influenzae*, *Pneumococcus* and *Neisseria meningococcus*) was administered on POD-3. Patient received adequate analgesia and antibiotics for initial three postoperative days. Deep breathing exercises using incentive spirometer and chest physiotherapy were continued until POD-9. Liver biopsy showed 'portal tracts with minimal chronic inflammation, no significant interface hepatitis, no periportal fibrosis, normal hepatocytes with maintained lobular architecture'. Patient was discharged on POD-9. UGIE at 6 months follow-up revealed grade II varices. During her follow-up visit at 1.5 years, patient has improved quality of life with no episode of haematemesis, no requirement of blood transfusion, reduced number of hospital visits and no hospital admission.

DISCUSSION

Maternal and perinatal outcome in pregnancy with PH is directly related to extent of liver disease and oesophageal varices at the time of conception. Complications of PH are more common in cirrhotic liver than in non-cirrhotic liver. Variceal bleed is a catastrophic complication of PH during pregnancy and is associated with poor outcome. Among those with pre-existing varices, 78% will have gastrointestinal bleeding during pregnancy with a mortality rate of 18% to 50%.⁷ Variceal bleeding during pregnancy is associated with a higher incidence of abortion (29.4%) and perinatal death (33.3%).⁸ Even though second, third trimester and second stage of labour are associated with the greatest risks of variceal bleed, active variceal bleeding may occur at any stage of the pregnancy. Incidence of variceal bleeding was 93.3% in freshly diagnosed cases of NCPF with PH, and 8.6% when the disease was diagnosed prior to pregnancy.⁹ Patients with NCPH who conceived after the treatment of oesophageal varices by endoscopic sclerotherapy or a decompression

surgery had a successful pregnancy outcome.^{10 11} Beta blockers reduce portal pressure and decrease the risk of variceal haemorrhage but their use in pregnancy is limited due to the risk of fetal growth restriction and fetal bradycardia.¹² Predictors of variceal bleeding include history of variceal bleed in the past, presence of jaundice, hypersplenism, ascites, thrombocytopaenia and endoscopic signs of large varices. In the absence of definite recommended guidelines for treatment of oesophageal varices during pregnancy, screening UGIE can be performed during the second trimester. The main risk of UGIE during pregnancy is fetal hypoxia due to sedation or positioning.¹³ Patients with uncontrolled bleeding not responding to sclerotherapy and variceal ligation will require surgical exploration. Splenic rupture in pregnancy is a life-threatening complication, with fetal and maternal mortality rates of 95% and 70%, respectively.¹⁴ Clinical presentation of splenic rupture may mimic abruptio placentae or uterine rupture. Mortality is commonly due to haemorrhagic shock. Incidence of postpartum haemorrhage in pregnancy with PH vary from 7% to 26% and is generally due to coagulopathy and thrombocytopaenia secondary to hypersplenism.¹⁴ Spontaneous bacterial peritonitis increases the risk of preterm delivery and abruptio placentae. Rupture of splenic artery aneurysm is a rare cause of mortality and may occur in 2.6% of pregnant women with cirrhosis of liver. Duplex ultrasound examination of abdomen is useful investigation to look for a splenic artery aneurysm. Clinical presentation of ruptured splenic artery aneurysm are pain and haemorrhagic shock. Most of the ruptures occur in the third trimester necessitating emergency laparotomy, ligation of the aneurysm or splenectomy.

Management of complications of PH arising during pregnancy is similar to management in the non-pregnant patient. Patient had undergone preconceptional counselling and therapeutic UGIE prior to conceiving. Preconception

counselling is necessary as pregnancy should be planned only when the liver functions are preserved and the patient agrees for regular follow-up and close monitoring. During counselling, patients are explained about the risks associated with recurrence of variceal bleeding, rupture of spleen and postpartum haemorrhage. Surveillance or therapeutic UGIE performed prior to conceiving is associated with good prognosis.^{15 16} Our patient was advised splenectomy for transfusion-dependent pancytopenia that was due to hypersplenism in NCPH. There is increased risk of prematurity, intrauterine growth retardation and preterm labour following abdominal and pelvic surgeries during pregnancy.¹⁷ Therefore, timing of these surgeries requires balancing between maternal and fetal risks. Plan of treatment in our case was intelligent conservatism of spleen with expectant full term normal vaginal delivery, caesarean delivery was reserved only for obstetric indications and surgery for PH will be performed during pregnancy only if it is essential. Vaginal delivery is safe in women with PH and second stage of labour may need to be cut down by instrumental deliver. Operative vaginal delivery should be avoided in thrombocytopenia.¹⁸ Caesarean section may be complicated by the presence of grossly dilated varices on the anterior abdominal wall and precautions to be taken while giving the abdominal incision. Our patient was counselled about safety and feasibility of various procedure (such as postpartum splenectomy, postpartum vascular shunt surgery, postpartum devascularisation procedures and combined caesarean with splenectomy). Theoretically, vascular shunt surgery should be the most successful procedure, but they carry the risk of anaesthesia and technical difficulties during pregnancy. As our patient did not respond to induction of labour, an immediate plan for emergency caesarean section was taken. As the patient requested for combined caesarean with splenectomy, a written and informed consent was obtained after counselling about risks and benefits of surgery. Patient wanted to undergo combined caesarean with splenectomy as two procedures will be performed under single anaesthesia.

While carrying out delivery in patients with PH, team must take care of blood volume, coagulation disorders, raised intra-abdominal pressure and risks associated with anaesthetic medicines to be administered. Adequate units of blood and plasma should be arranged and measures for balloon tamponade for the variceal haemorrhage must be kept handy. Sodium and water retention in pregnancy can cause pulmonary oedema and generalised seizures. Seizures may cause splenic rupture and hypotension. Epidural analgesia could be a preferred mode. Spinal or epidural anaesthesia is contraindication due to low platelet count. Platelet count less than 80 000 cells / μ L or coagulopathy general anaesthesia would be preferred.¹⁹ The hypertensive response to intubation and straining during extubation would increase the risk of variceal haemorrhage.²⁰ Thrombocytopenia may require administration of platelets following ligation of splenic artery. Large uterus and enlarged spleen occupying entire abdomen cause loss of space to operate in the abdomen cavity. Spleen can easily rupture with aggressive manipulation owing to its large size, soft and friable consistency. There is high flow of blood in the splenic artery, short gastric vessels and collateral vessels surrounding spleen. Thus, unnecessary blunt dissection may lead to increased risk of bleeding. Splenic vein is large, dilated and thin walled. A meticulous dissection is desired around splenic vein and its collateral vessels to avoid bleeding. Ligation of

Learning points

- Preconceptional counselling and treatment of oesophageal varices was paramount in the management of pregnancy with portal hypertension (PH).
- Multidisciplinary approach in tertiary care centre (multispecialty care and good blood bank facility) gave the best outcome in pregnancy with PH.
- Combined caesarean with splenectomy is feasible in pregnancy with PH. It is a cost-effective treatment associated with improved quality of life in patients with PH.
- Prospective clinical trials are essential to prove safety and efficacy of combined caesarean with splenectomy in pregnancy with PH.

splenic artery, short gastric vessels, splenic vein is done and spleen is delivered after releasing it from all the ligaments. These patients have to be monitored for bleeding as they are at risk for postpartum haemorrhage. Puerperal fever should be investigated and treated with appropriate antibiotics. All postsplenectomy patients carry risk of infection although vaccination and prophylactic antibiotic regimen have reduced infection-related morbidity and mortality.²¹ Our patient was explained about deep breathing exercises using incentive spirometer and chest physiotherapy. Except for thrombocytosis that she had developed on POD-3, she had uneventful postoperative recovery. She was discharged on POD-9. UGIE at 6 months of follow-up revealed grade II oesophageal varices. Patient has improved quality of life with no episode of haematemesis, no requirement of blood transfusion, reduced number of hospital visits and no hospital admission in the past 1.5 year. Considering the feasibility of combined caesarean with splenectomy and improved quality of life postoperatively, we suggest following management algorithm (figure 2) for treatment in pregnancy with PH. Prospective studies are required to provide conclusion on the safety and efficacy of combined caesarean with splenectomy surgery.

Contributors MMP conceived the design. MMP was the first operating surgeon; LUC and RG were first and second assistant surgeons, respectively. Patient was evaluated, resuscitated and discussed with MMP on telephone. Patient was operated by MMP, LUC and RG. LUC performed caesarean delivery and MMP, RG and YSR performed splenectomy. RG collected the operating steps videography photographs. Demography of the patient, clinical details (data) and video editing was done by RG and YSR and further it was analysed by MMP. Manuscript was prepared by MMP, RG and YSR. Editing of image and video was performed by RG. Case report was written, critically analysed and uploaded by MMP. During first revision of case report, MMP and LUC revised and edited the case report completely. Final approval of the case report is provided by MMP, RG, LUC and YSR. Overall responsibility and corresponding author is MMP.

Competing interests None declared.

Patient consent Obtained.

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