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ORIGINAL RESEARCH

Splenomegaly in Pregnancy-Evaluation of Causes and Pregnancy Outcome- A Case Series

Dr. V. Prathyusha[1], Dr. Anumpama Hari[2], Dr Shalini[3] , Dr. K. Vishali [4]

[1]Assistant Professor,Department of Obstetrics and Gynaecology,Mallareddy Medical College for women, Hyderabad,Telangana
[2]Professor,Department of Obstetrics and Gynaecology,Mallareddy Medical College for women, Hyderabad,Telangana

[3]Assistant Professor ,Department of Obstetrics and Gynaecology,Mallareddy Medical College for women,Hyderabad, Telangana
[4] Corresponding Author, Assistant Professor,Department of Obstetrics and Gynaecology,Mallareddy Medical College for women, Hyderabad,Telangana

ABSTRACT

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

Materials and Methods: We reviewed the course of pregnancy, maternal and fetal outcomes of 6 pregnant women with massive splenomegaly who were managed in our unit during 2019-2021.

Results: We reviewed the course and outcome of 6 pregnant women with massive splenomegaly {>17cm}. 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); 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 peripheral smear for malaria parasite was done as indicated. The course of pregnancy, mode of delivery, and maternal and fetal outcome were noted.

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

Keywords: splenomegaly, portal hypertension, esophageal varices.

Corresponding Author: Dr. K. Vishali, assistant professor

INTRODUCTION

Pregnancy associated with massive splenomegaly is an infrequent entity and when associated with anaemia, thrombocytopenia, ascites and jaundice poses grave complications in maternal and fetal aspects.[6] Splenomegaly can be asymptomatic or can be a clinical presentation of underlying diseases like liver diseases, hematologic malignancies, infections like Malaria or right heart failure. The literature presents sparse information on obstetric aspects of Pregnancy while cases of splenic disorders during pregnancy and diseases causing

splenomegaly during pregnancy are well documented .In our case series we report 6 cases of Splenomegaly presenting in different trimesters of pregnancy. Many complications were encountered during the pregnancy which were tackled by a multidisciplinary team comprising of obstetrician, gastroenterologist, physician, intensivist and neonatologist. We present our experience of pregnancy with massive splenomegaly in this case series.

MATERIALS & METHODS

In the present study, we have reviewed the course and outcome of 6 pregnant women with massive splenomegaly who were managed during 2019–2021 at the Department of Obstetrics and Gynecology, Mallareddy medical college for women. Massive splenomegaly was defined as a spleen with craniocaudal length of 17 cm or more on ultrasonography.(2)Patients' characteristics including age, obstetric and medical history(fever, gastrointestinal haemorrhage, blood transfusions), 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 and splenic vein), upper gastrointestinal (GI) endoscopy autoimmune work-up (lupus anticoagulant and anti-phospholipid antibody) and work-up for infections(peripheral smear for malaria parasite) was done as indicated. Treatment was given to all the patients after consulting a physician/gastroenterologist. Fetal surveillance was done with fortnightly growth monitoring and weekly umbilical artery Doppler indices. The course of pregnancy, mode of delivery, and maternal and fetal outcome were noted. Indications for preterm termination of pregnancy were documented.

RESULTS

Table 1: Case details

	Case 1	**Case 2**	**Case 3**	**Case 4**	**Case5**	**Case 6**
Age	**27yrs**	**25yrs**	**20y**	**20yr**	**28yr**	**30yr**
Parity	G4P3L2D1 , at 34 weeks	G2P1D1 at 18 weeks	Primi at 20 weeks	Primi with 20 weeks	Primi with 37weeks	G2A1 with 20 weeks
Etiology	Non cirrhotic portal hypertension	Cirrhosis with portal hypertension	Non cirrhotic portal hypertension	B-thalassemia intermedia	SLE (ANA and anti-ds DNA positive.	SLE (ANA, ds DNA positive)
Spleen size and USG features	17.5cm	25cm	20cm. Portal vein 19mm.Portal htn with recanalised paraumbilical veins.	18cm	20cm	21cm. Dilated splenic vein-16mm Dilated portal vein at porta hepatis-19mm
Upper GI endoscopy	Normal	Grade 2 esophagealvarices, PHG(Stomach-small discrete clumps of veins in fundus	Portal hypertensive gastropathy	Normal	normal	Grade 2 esophagealvarices
Complications	Severe PE	Gestational HTN , gross ascites	Pancytopenia	FGR, hypothyroidism	CCF , severe anaemia	Ascites, Pre eclampsia, suture site

						hematoma.
Transfusions	nil		PRBC and SDP	8 PRBC (in antenatal and post natal period)	9 PRBC, 2 FFP,2 RDP	2 SDP
Pregnancy outcome	Induced with prostaglandin gel, Preterm delivery at 34weeks	Termination of pregnancy	Termination of pregnancy	Prostaglandin gel induction ,term vaginal delivery	PGE2 induction, term vaginal delivery	Elective lscs. ind: cpd
Baby weight	2kg	200gm	350gm-	2.3kg	2kg	2.6kg
Apgar	7-9	-	-	6-8	8-9	8-9
Management	-	Diuretics, beta blockers. Variceal banding performed after termination of pregnancy	-		Prednisolone	Prednisolone and hydroxyl chloroquine

During the study period, 6 pregnant women were diagnosed with massive splenomegaly [Table 1]. The age of the women ranged from 20 to 30 years. Three of them presented in third trimester and three cases of splenomegaly were diagnosed during routine obstetric examination and investigation in second trimester.

One patient had cirrhosis with portal hypertension with gross ascites (case 2).Two patients (case 1and case 3) were diagnosed with portal hypertension due to non-cirrhotic portal fibrosis (NCPF) and one of them had pancytopenia (case 3).One patient was diagnosed with beta thalssemia intermedia at 20 weeks of gestation and had history of blood transfusions during the antenatal period. Two patients were diagnosed with SLE. Presentation varied among the patients. 2 of them presented with gestational hypertension/ pre eclampsia.

1 patient (case 5) presented with severe anaemia and congestive cardiac failure. Three patients were diagnosed on examination and investigations. All the patients were admitted in high risk obstetric unit for evaluation and management. All 6 patients were diagnosed with anaemia (haemoglobin<10 g/dl) and 2 of them had severe anaemia(Hb< 7g/dl). 2 of them had thrombocytopenia (platelet count 30 000 to 50 000cells/cmm) and 2 had leucopenia (2400–3100 cells/cmm).1 patient had pancytopenia. Coagulation profile (prothrombin time and APTT) was normal in all the patients .Liver function tests were normal in two patients, but 4 patients had raised serum bilirubin .Liver enzymes were marginally raised in 2 patients. Viral markers for hepatitis were found to be negative in all patients.2 patients has positive ANA and anti-ds DNA antibodies. On ultrasonography, the size of spleen ranged from 17 cm to 25 cm .Portal vein diameter range is from 18-20mm. The liver is cirrhotic in one patient. One patient had gross ascites [Table 1]. Upper GI endoscopy showed grade 2 eophageal varices in 3 patients. Of the three, in two patients esophageal banding was done in 2nd trimester. In 1 patient banding was done after delivery. Patients who were stable were advised regular and frequent antenatal checkups. Those patients with complications and abnormal investigation results were admitted for further multidisciplinary management .One patient with cirrhosis and portal hypertension (case 2) had massive ascites and was advised medical management followed by drainage of ascitic fluid. One patient (case 6) developed moderate ascites after cesarean section and was managed medically. Maternal complications observed in the

patients were gestational hypertension, ascites, cytopenias, abnormal liver function and jaundice, need for multiple blood transfusions, wound hematoma in 1 patient (case 6), prolonged hospital stay .The most common perinatal complications were Intrauterine fetal growth restriction (IUGR), prematurity and small for gestational age infants .

In 2 patients (case 2 and 3), pregnancy was terminated at 20 weeks as indicated by maternal complications. In 2 patients vaginal delivery was planned and labour was induced with pgE2 gel (case 4 and 5). Intrapartum electronic fetal monitoring was done and they had assisted vaginal deliveries. 1 patient had emergency cesarean section in view of severe pre eclampsia and fetal distress (CASE 1).In one patient (case 6) elective cesarean section was performed for obstetric indication (CPD). Patients (cases 3, 4 5) required multiple blood and blood products transfusion in the antenatal and post natal period.

The mean birth weight was 2.2 kg and babies needed NICU support and admission in view of preterm delivery and respiratory distress. One patient (case 2) with cirrhosis and portal hypertension had gross splenomegaly and ascites in the antenatal period and was medically managed for the condition and pregnancy terminated. One patient (case 6) developed ascites after delivery and was medically managed. Patient (case 6) developed wound hematoma which was drained and resuturing performed .The hospital stay was prolonged in all the patients (minimum 3 weeks). All the mothers and babies were discharged and advised follow up for a period of 12 weeks.

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. The common causes of massive splenomegaly seen in pregnancy are haemolytic anaemia, thalassaemia, myelofibrosis, malaria, kalaazar, tuberculosis, portal hypertension (cirrhosis, extra hepatic portal vein obstruction, and non-cirrhotic portal fibrosis) and collagen disorders (systemic lupus erythematosus).[4]

Two patients in our case 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. Thalassemic women have an increased risk for thrombosis, as the disease entity is a chronic hypercoagulable state with high incidence of thromboembolic episodes. Additional risk factors for venous thromboembolism during pregnancy and the postpartum period are the reduction in venous flow velocity during gestation, inherited thrombophilias, antiphospholipid syndrome, and previous history of thrombosis. Thrombophylaxis might be essential during pregnancy and the postpartum period in cases of nontransfused TI, splenectomy, or a history of recurrent abortions.[1]

NCPH is the most common cause of portal hypertension in developing countries. The prognosis of this disease is better than cirrhosis of liver. The increased blood volume and cardiac output in pregnancy increases the portal venous flow and aggravates portal hypertension and its complications. Patients with NCPH present with upper gastrointestinal bleed, ascites, splenomegaly, and jaundice and growth restriction. Other squeal includes hyper dynamic circulation, pulmonary complications and portosystemic encephalopathy secondary to increased portosystemic collateral circulation. Prenatal obliteration of varices by either endoscopic variceal ligation (EVL) or endoscopic sclerotherapy (EST) reduces the risk of complications and certainly improves the pregnancy and perinatal outcome.[6,7,9,11]

The mode of delivery in women with massive splenomegaly remains a concern for obstetricians. There is a risk of variceal bleeding because of repetitive

Valsalvamanoeuvre,[5] and hence some experts advise an elective caesarean section in those with high-grade varices.[6]

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. 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.[7]

Antibiotic use needs to be individuazised with spontaneous bacterial peritonitis in mind for patients with cirrhosis of liver. 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 extramedullaryhemopoiesis giving rise to increase in splenic size and finally splenic rupture. 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. Thirdly frequent abdominal examinations and manipulations during labour can also give rise to splenic trauma.

CONCLUSION

A multidisciplinary team approach in tertiary care center with availability of intensive care units is likely to yield best pregnancy outcome in pregnant women with cirrhosis and portal hypertensive despite various associated complications.

REFERENCES

1. Petrakos G, Andriopoulos P, Tsironi M. Pregnancy in women with thalassemia: challenges and solutions. Int J Womens Health. 2016;8:441-451. Published 2016 Sep 8. doi:10.2147/IJWH.S89308.
2. Tolunay HE, Aydın M, Cim N, Boza B, Dulger AC, Yıldızhan R. Maternal and Fetal Outcomes of Pregnant Women with Hepatic Cirrhosis. Gastroenterol Res Pract. 2020 Jan 8;2020:5819819. doi: 10.1155/2020/5819819. PMID: 32190039; PMCID: PMC7064848.
3. Tan, J., Surti, B. and Saab, S. (2008), Pregnancy and cirrhosis. Liver Transpl, 14: 1081-1091. https://doi.org/10.1002/lt.21572
4. Hoffbrand AV, Moss PA (eds). The spleen. In: Hoffbrand'sEssentialHaematology. Oxford:John Wiley & Sons; 2015:116–21.
5. Gala AR, Surapaneni T, Aziz N, Kallur SD. A Review of Outcomes in Pregnant Women with Portal Hypertension. J ObstetGynaecol India. 2018 Dec;68(6):447-451. doi: 10.1007/s13224-017-1016-1. Epub 2017 Jun 5. PMID: 30416270; PMCID: PMC6207547.
6. Sandya MR, Manjula SK, SrAncy. Pregnancy outcome in Non-cirrhotic portal hypertension. Int J ReprodContraceptObstetGynecol 2017;6:4534-7.
7. Keepanasseril A, Gupta A, Ramesh D, Kothandaraman K, Jeganathan YS, Maurya DK. Maternal-fetal outcome in pregnancies complicated with non-cirrhotic portal hypertension: experience from a Tertiary Centre in South India. Hepatol Int. 2020 Sep;14(5):842-849. doi: 10.1007/s12072-020-10067-5. Epub 2020 Jun 25. PMID: 32588317.
8. Vettorazzi, J. ,Valério, E. , Reis, A. , Torres, H. and Petermann, A. (2018) Massive Splenomegaly in Pregnancy: Case Report. Open Journal of Obstetrics and Gynecology, 8, 163-169. doi: 10.4236/ojog.2018.83019.

9. Khanna R, Sarin SK. Non-cirrhotic portal hypertension - diagnosis and management. J Hepatol. 2014 Feb;60(2):421-41. doi: 10.1016/j.jhep.2013.08.013. Epub 2013 Aug 23. PMID: 2397871
10. Singhal S, Roy KK, Kumar S, Meena J, Bharti J, Shekhar B. Pregnancy with massive splenomegaly: A case series. Natl Med J India 2018;31:146-148
11. Subbaiah M, Kumar S, Roy KK, Sharma JB, Singh N. Extrahepatic portal-vein obstruction in pregnancy. Taiwan J ObstetGynecol 2015;54:394–7.
12. Carreras LO, Defreyn G, Machin SJ, Vermylen J, Deman R, Spitz B, Van Assche A. Arterial thrombosis, intrauterine death and "lupus" antiocoagulant: detection of immunoglobulin interfering with prostacyclin formation. Lancet. 1981;1:244–6. doi: 10.1016/S0140-6736(81)92087-0.
13. Schouten JN, Verheij J, Seijo S. Idiopathic non-cirrhotic portal hypertension: a review. Orphanet J Rare Dis. 2015 May 30;10:67. doi: 10.1186/s13023-015-0288-8. PMID: 26025214; PMCID: PMC4457997.
14. Rajekar H, Vasishta RK, Chawla YK, Dhiman RK. Noncirrhotic portal hypertension. J ClinExpHepatol. 2011 Sep;1(2):94-108. doi: 10.1016/S0973-6883(11)60128-X. Epub 2011 Nov 9. PMID: 25755321; PMCID: PMC3940546.
15. Pratima Mittal, JyotsnaSuri, ReetaBansiwal. "Massive Splenomegaly in Pregnancy: A Review of three cases". Journal of Evolution of Medical and Dental Sciences 2013; Vol2, Issue 27, July 8; Page: 5042-5046.