

Portal Cavernoma in Pregnancy

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INTRODUCTION

A portal cavernoma is a consequence of chronic PVT and occurs when myriads of collateral channels develop to bypass the occlusion. It often leads to portal hypertension because the collateral veins are not able to adequately handle the splenic and mesenteric inflow. Pregnancy in a patient with portal hypertension presents a special challenge to the obstetrician because physiological hemodynamic changes associated with pregnancy, needed for meeting demands of the growing fetus, worsen the portal hypertension thereby putting mother at risk of potentially life-threatening complications.

CASE REPORT:

A 29 year old Primigravida conceived after 6 cycles of ovulation induction. An ultrasound was done at 16 weeks and it incidentally detected portal cavernoma with splenomegaly. Due to the rarity of occurrence of a portal cavernoma with pregnancy the patient was referred to our tertiary care hospital. Patient was asymptomatic and admitted at 21 weeks for evaluation. A detailed and focused history taken revealed that the patient had a past medical history of jaundice 1 and a 1/2 years back and had a hospital stay of 4 days, what was thought to be for Viral hepatitis. In our current evaluation, patient was hemodynamically stable and a moderate degree of pallor was present. Abdominal examination revealed a splenomegaly (19 cms, firm in consistency, smooth surface, and no tenderness). There was no evidence of free fluid or engorged veins per abdomen. Uterine height corresponded to a gestation of 22 weeks.



Her laboratory investigations revealed the following:

HAEMOGRAM	
HAEMOGLOBIN	8.5
TLC	2,100
DLC	N75 L13 E 2 B1M9
ESR	9 mm/hr
RBC COUNT	3.19 *10 ⁶ /cmm
PLATELET	29,000/cmm
PERIPHERAL SMEAR	Pancytopenia(mild normocytic normochromic anemia with neutropenic leukopenia and severe thrombocytopenia)

TOTAL PROTEIN	6.26g/dl
SERUM ALBUMIN/GLOBULIN	3.71 / 2.5 g/dl
SERUM A/G	1.5
SERUM TOTAL BILIRUBIN	1.98g/dl
SERUM CONJUGATED BILIRUBIN	0.64 g/dl
SERUM UNCONJUGATED BILIRUBIN	1.34g.dl

Prothrombin time (TEST /CONTROL)	14.5/21.1
Hb electrophoresis	Normal CA, A, A2 Pattern
Bonemarrow smear	Hypercellular marrow with erythroid hyperplasia
ANA	NEGATIVE
Hepatitis markers	Negative
Serum compliment C3	76 mg/dl
Ceruloplasmin	36.4

Abdominal ultrasound showed a portal vein that was 18mm dilated & showed evidence of thrombosis with recanalization and multiple collaterals around portal vein were seen.



Fig. 1: Portal Cavernoma (white arrow)



Fig. 4: Splenomegaly

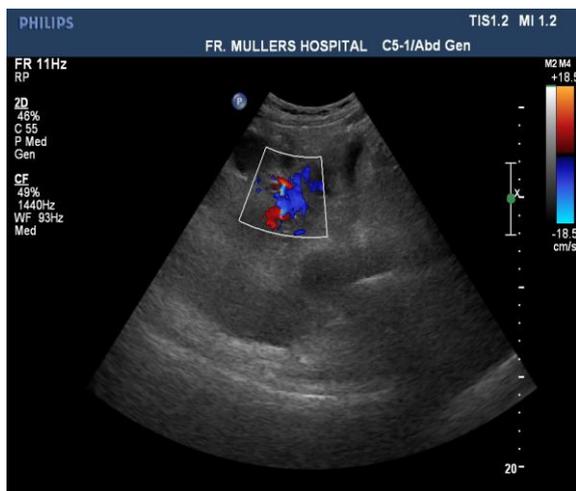


Fig. 2: Portal collaterals (doppler)

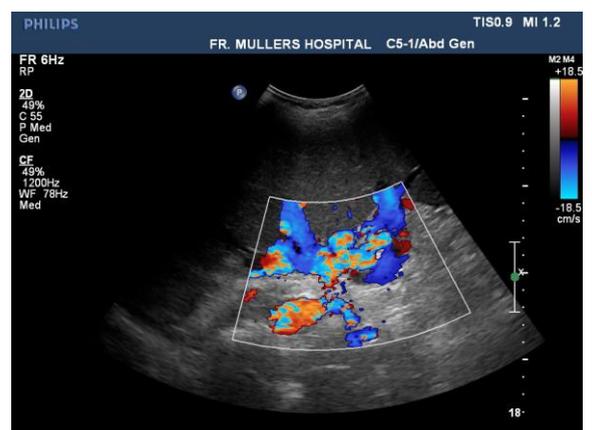


Fig. 5: Splenic varices (doppler)



Fig. 3 Splenic Varices



Fig. 6: Esophageal varices on gastroscopy

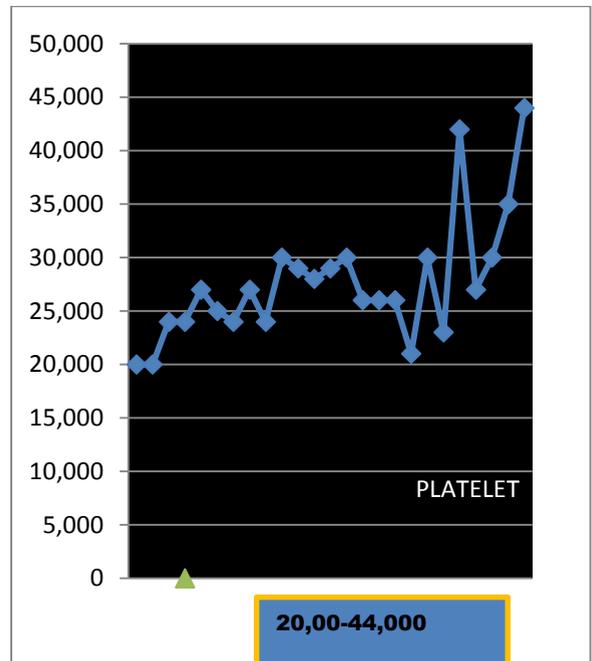
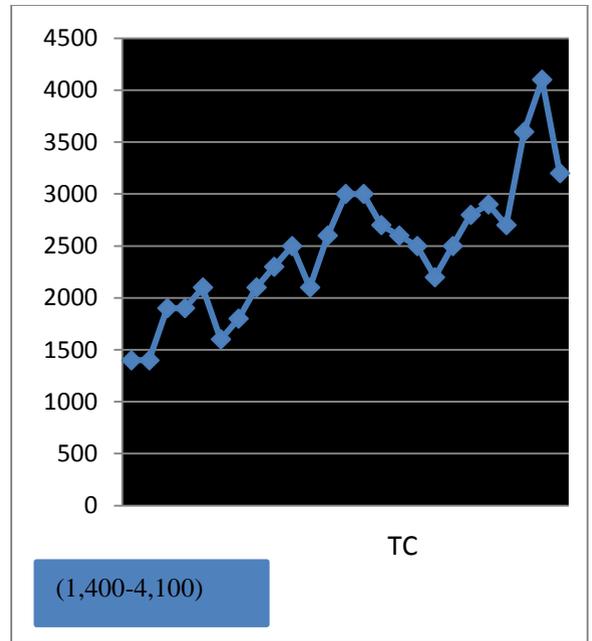
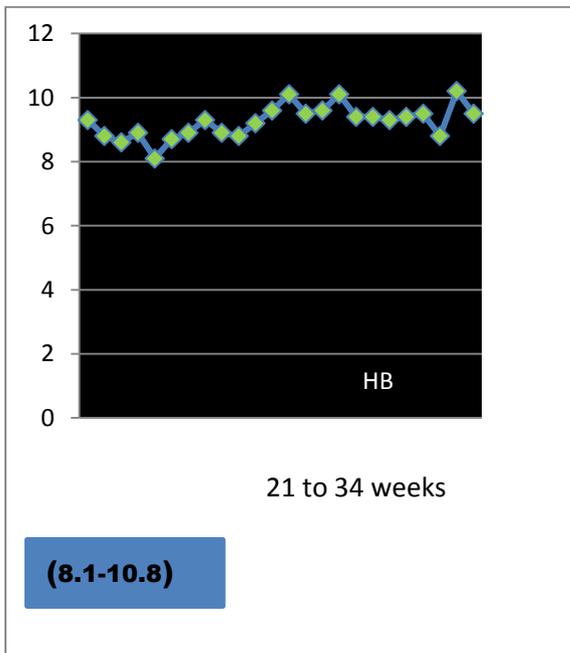
A gastroscopy was done and the following findings were noted: Small to medium sized varices noted in esophagus (GRADE 2) noted in fundus & cardia.



Fig. 7: Varices extending upto the Fundus

Following the above investigations, no underlying cause for portal venous hypertension or any cause for extra hepatic portal vein occlusion could be demonstrated in our case. All maternal and fetal risks were explained. Followup was continued and serial monitoring by ultrasonography revealed no fetal growth restriction. Hemoglobin, white cell counts & platelets were monitored twice weekly.

As a part of treatment, propranolol 20mg twice daily was started. INJ.METHYL COBALAMIN 1000ug intravenously once daily for 7 days and maternal corticosteroids for fetal lung maturity was administered.



Patient looked icteric at 36 weeks 4days and in view of worsening liver function (total bilirubin 4.85 mg/dl, direct bilirubin: 1.17 mg/dl, indirect bilirubin: 3.68mg/dl), a plan for induction of labor was made. 1 apheric platelet was transfused followed by induction of labor after 48 hrs with dinopristone gel 0.5mg intracervically & after 6 hrs augmented with oxytocin. During labor 2units of platelets transfused. An emergency caeserean was done in view of fetal bradycardia and a healthy male baby of weight 2.160 kg & apgar 8 and 9 was delivered.

Postoperatively 2 units of platelets & 1 unit of packed cell were transfused.

On post-operative day three, the following was noted: platelets: 47,000 Total bilirubin: 7.52mg/dl Indirect 6.63 mg/dl, LDH: 327, peripheral smear (normocytic to macrocytic with moderate lymphopenia and severe thrombocytopenia). In view of hypersplenism and decreasing counts, despite transfusions a plan for splenectomy was made. The patient recovered well postoperatively, and on discharge was advised two doses of pneumococcal vaccine.



Fig. 8: Massively enlarged spleen

The management dilemmas that we faced in the management of this case were issues regarding the management of varices, to do a splenectomy vs a conservative approach, the need for anticoagulation, the need for apheresis platelets, the need induction of labor and the mode of delivery and the appropriate postoperative workup.

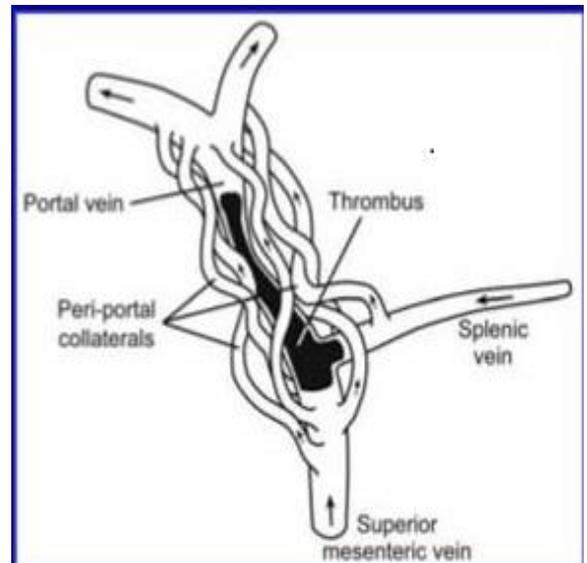
DISCUSSION

Balfour and Stewart first described portal cavernoma in 1869 as thrombosis and varicose dilatation of the portal vein leading to splenomegaly and ascites.

Kobrich coined the term “cavernoma” to describe the peculiar spongy appearance of the portal vein & the phrase cavernomatous transformation of the portal vein” to imply that this condition followed thrombosis of the portal vein.¹

The pathogenesis of this condition remains elusive. The following causes have been postulated:

- mechanical-obstructive
- congenital
- neoplastic



- Arterial rescue
- Venous rescue(6 to 20 days to 3-5weeks)^{2,3}

The causes for portal hypertension can be local (70%) and systemic(30%).⁴ Two large studies on pregnant women with chronic portal venous thrombosis, showed that variceal bleeding is the most common clinical complication followed by thrombosis, abdominal pain, jaundice, and incidental splenomegaly^{5,6}

Hypersplenism and the consequent pancytopenia can be present in chronic PVT⁷. The complications of portal cavernoma in pregnancy are severe anemia, hepatic decompensation leading to progressive liver & renal failure, hepatic encephalopathy, splenic artery aneurysm rupture, ascites, subacute bacterial peritonitis & post partum hemorrhage. Currently the American Association for the Study of Liver Disease (AASLD) recommends screening endoscopy in the second trimester as that is the time of maximum increase in the portal pressure.⁹ Mishra et al recommended primary prophylaxis with non-selective beta blockers in patients with large and high risk gastric varices to reduce the risk of first bleed and mortality.¹⁰

The Asian Pacific Association for the Study of the Liver (APASL) has developed consensus statement on various aspects of extra hepatic portal vein occlusion (EHPVO) including pregnancy and recommended that vaginal delivery can be anticipated in most of these women. Cesarean is usually reserved for obstetrical indications.¹¹ Preconception counselling they say is an integral part of management. Role of anticoagulation is not proven in chronic PVT.¹²

Till now in literature 6 cases of portal cavernoma in pregnancy were identified. To our knowledge, this is the first case of a portal cavernoma

with hypersplenism that had a continued pregnancy to term.

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