DOI: 10.14260/jemds/2015/1049

CASE REPORT

ACUTE BUDD-CHIARI SYNDROME DURING PREGNANCY: CASE REPORT

Venkateshmurthy[1], Kayalvizhi K. B[2], Sahajananda H[3]

HOW TO CITE THIS ARTICLE:
Venkateshmurthy, Kayalvizhi K. B, Sahajananda H. "Acute Budd-Chiari Syndrome during Pregnancy: Case Report". Journal of Evolution of Medical and Dental Sciences 2015; Vol. 4, Issue 41, May 21; Page: 7233-7236, DOI: 10.14260/jemds/2015/1049

ABSTRACT: Budd-Chiari syndrome (BCS) is characterized by occlusion of hepatic veins by a tumor or a thrombus occurring in the vein or an extension from the inferior venacava or right atrium.[1,2] Prevalence of Budd-Chiari Syndrome in pregnant woman varies is about 6.7% throughout the world with an incidence of 7.1% in Asian population.[3] Thereby pregnancy is known to pose risk for occurrence of acute form of BCS, which can prove to be fatal. Here we present one such case with pregnancy related Budd-Chiari Syndrome with acute presentation, posing a challenge to anesthetists & intensivists.

KEYWORDS: Budd- Chiari syndrome, Pregnancy.

CASE REPORT: We hereby report a case of 22 years old primigravida with full term pregnancy coming with mild abdominal pain. She was on regular antenatal checkups in private hospital with no co morbidities and previous investigations being normal with latest ultrasound showing single live fetus with no fetal abnormalities. She had no other co-morbidities. She came to our hospital early in the morning for safe delivery presenting with mild abdominal pain.

The severities of pain accentuated late in the evening, respite all medications and bed rest & associated with bleeding per vagina. Clinical examinations revealed pale and diaphoretic patient, with heart rate 120/min, blood pressure 80/55mm of a suspicion of abruptio placenta.

After securing 2 large bore intravenous lines, crystalloids and colloids were rushed. She was pre-medicated with inj. Ranitidine 50mg i.v, inj. Glycopyrrolate 0.2mg i.v. Rapid sequence induction done using inj. Ketamine 50mg, intubated using inj. scoline 100mg and 7.0mm cuffed oral Endo tracheal tube, maintaining cricoid pressure throughout. Bilateral air entry checked and tube was secured. Anesthesia was maintained with N_2O 4L/min, O_2 4L/min and inj. Vecuronium.

Emergency Lower segment Caesarean section done and live baby was extracted. Placenta was intact with no abruption. Uterus contracted well after placental delivery. But patient continued to bleed from intra-abdominal cavity. Since the sight of bleeding could not be identified and was heavy, good abdominal compression and packed cell transfusions helped maintaining blood pressure. Meanwhile right internal jugular vein was cannulated with central venous catheter until General surgeons arrived for help and decided to do laparotomy. Fig. 1 and 2.

DOI: 10.14260/jemds/2015/1049

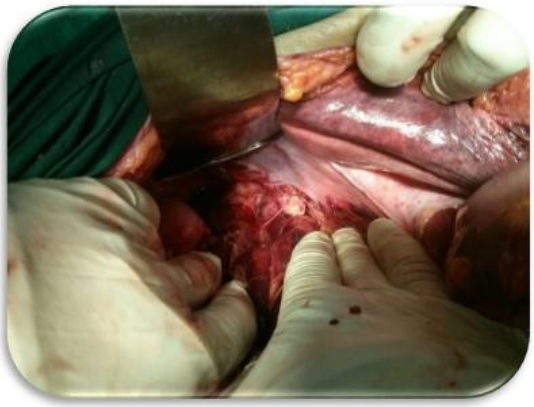

Fig. 1: Securing haemostasis after splenectomy

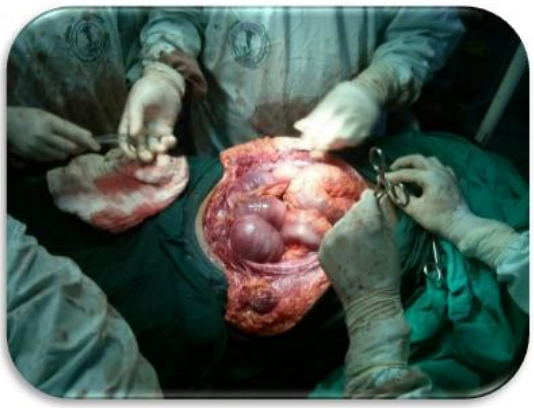

Fig. 2: Enlarged spleen with continuous bleeding

After placing midline incision, severe oozing from abdominal cavity was noted. There was also severe bleeding from the perisplenic area, lesser sac, both the paracolic gutters. After opening the lesser sac, bleeding was seen from the body of the pancreas and superior mesenteric artery, with enlarged spleen, dilated splenic and pancreatic vessels. Splenectomy was done, but bleeding persisted. Vascular surgeon was summoned who tried all the maneuvers to control bleeding from peripancreatic area. Bleeding increased with each meticulous approach. So it was decided to keep 3 abdominal mops soaked with abgel and sepgaurd solution and close the abdomen, to open later, once the patient was stabilized.

The whole surgery lasted for 3 ½ hrs and estimated blood loss was more than 3 liters. 8 litres of crystalloids, 6 packed red blood cells, 5units of Fresh frozen plasma, 2 units of whole blood, 4 units of platelets, 1 cryoprecipitate, 1.5 litres of Hestarch was transfused intraoperatively. Continuous infusion of Dopamine, noradrenaline and dobutamine were used to maintain blood pressure around 90/60mmHg to 110/70mmHg, Mean Arterial Pressure 60 to 65mmHg. Heart rate 110-120/min, Spo_2 95 to 99%.

Patient was shifted to Surgical Intensive care unit for further monitoring, stabilization and ventilatory support. Along with good antibiotic coverage and ionotropes, blood gas analysis revealed pH-6.98, PCO_2-32, PaO_2 135, HCO_3 7.6. Approppriate measures were taken to correct the acidosis. Hb 6.8gm/dl, platelets 60,000/cu.mm creatinine 1.3 with absolute anuria. Central Venous pressure 11 cm of H_2O; PT-29: APTT 27 sec. INR: 2.3.

Liver function tests showed increase in Aspartate amino-transferrase (163) and Alanine amino-transferrase (120) Total protein 1.9; albumin-1.4, globulin-0.5, AG ratio 2.8:1. Serum electrolytes were normal. Echocardiography showed stress induced cardiomyopathy with global hypokinesia, Ejection Fraction 45% and mild pericardial effusion & pleural effusion. As we were suspicious of some occult liver pathology, a venogram and Doppler studies were ordered. But in the mean-time patient developed sudden cardiac arrest and despite all measures, succumbed to death. Postmortem examination revealed a massive thrombus in the post hepatic inferior vena cava extending up to hepatic veins and portal veins. Depending on these Post mortem findings, it was concluded that she had an acute type of Budd Chiari Syndrome from which she never recovered and succumbed to death.

DOI: 10.14260/jemds/2015/1049

DISCUSSION: The Budd-Chiari syndrome is a rare entity with hypercoagulable state[4] Other causes are myeloproliferative disorders, Paroxysmal nocturnal hemoglobinuria, deficiencies of protein C, Protein S, and antithrombin III.[4] The factor V Leiden mutation, the prothrombin gene mutation and methylene tetrahydrofolate reductase mutation have also been noted Hypercoagulable state of pregnancy increases the risk of venous thromboembolism and the use of contraceptive pills (until 6 months before pregnancy) is also said to cause Budd-Chiari syndrome.[4,5]

Abdominal pain is the presenting symptom of acute presentation, Hepatomegaly, jaundice, esophageal/gastric variceal bleeding, ascites, pedal edema are other symptoms depending on the type of BCS. Rarely it-may also present as acute liver failure with hepatic encephalopathy[5] Mild to moderate derangements in liver function tests is noted depending on the severity of the disease.

Pregnancy related Budd-Chiari syndrome is usually an acute disorder with Inferior vena cava and Hepatic vein obstruction with dilatation of the collateral vessels and bleeding[6] along with marked elevation of Alanine transaminase and Aspartate transaminase. Imaging studies recommended are Doppler Ultrasound, hepatic venogram, MRI of liver, and liver biopsy.[7]

Many cases of Budd-Chiari syndrome and Portal venous thrombosis in pregnancy have been reported in the literature. In one case report, fulminant Budd-Chiari syndrome presented with tender liver, marked ascites, markedly elevated Liver function tests–3 weeks after caesarian section which resulted in death of the patient.[8] In another case of fulminant type of Budd-Chiari syndrome in pregnancy, combined caesarian section and orthotopic liver transplantation were performed at 31st week of gestation & thereby patient survived.[8]

Our case presented with an acute onset of abdominal pain with vomiting and acute bleeding at 38th week of gestation–mimicking abruptio placenta. Only after Caesarean section and extraction of the baby, the real problem was suspected and possible investigations were done postoperatively, which revealed slightly enlarged liver, enlarged spleen (For which splenectomy was done), dilated gall bladder, dilated periportal veins, mild ascites, mild bilateral pleural effusion, mild pericardial effusion, markedly elevated liver enzymes, decreased Albumin: Globulin ratio, decreased serum albumin. Despite the hypercoagulable status of pregnancy there is still a risk of fibrinolysis resulting from liver failure.

Patient succumbed to death even before we could send her for further investigations. The pathology might have started early in the pregnancy which remained latent until the uterine contractions set in and increased abdominal pressure might have compressed the retro hepatic Inferior vena cava accelerating the thrombotic process, causing complete obstruction of the Inferior vena cava, portal vein, leading to acute presentation of Budd Chiari Syndrome.[7,8,]

CONCLUSION: Due to high mortality rates in fulminant cases of Budd Chiari Syndrome identifying at risk patients along with special investigations like hepatic venograms, contrast Doppler studies, MRI, and inception of early thromboprophylaxis may help in saving the patient.

REFERENCES:

1. M. Tilicos, A. Tsoulias, D. Tsantoulas et. al. Budd-Chiari Syndrome in pregnancy. A case report. Post graduate Medical journal (Oct. 1978) 54, 686-691.
2. Weirong Ren, Xiang Li, Jia Jia, Yanxia et.al. Prevalence of Budd-Chiari Syndrome during pregnancy or puerperium: A Systematic review and meta-analysis: Gastroenterology Research and practice ID 839875.

DOI: 10.14260/jemds/2015/1049

3. J.M.J Valentine, G. Parkin, S.G. Pollard and M.C. Bellamy combined orthotopic liver transplantation and caesarian section for Budd-Chiari Syndrome: British Journal of Anaesthesia 1995; 75:105-108.
4. K.V. Narayanan Menon, M.D, Vijay Shah, and Patrick S. Kamath: The Budd-Chiari Syndrome; the New England Journal of Medicine 2004; 350: 578-85.
5. C.E. Eapen, Thomas Mammen et. al: changing profile of BCS in India; Indian Journal of Gastroenterology 2007; 26; 77-81.
6. Dipika Mohanty, Shrimati Shetty, Kanjaksha Ghosh et. al: Hereditary Thrombophilia as a cause of Budd-Chiari Syndrome A study from western India; Hepatology 2001; 34: 666-670.
7. Marie-Helene Denninger, Yasmine Chait, Nicole Casadevall et al: cause of Portal or Hepatic Venous Thrombosis in adults: The role of multiple concurrent factors; The American Association for the study of Liver disease; 0270-9139/00/3103-000753.00/0: 587-591.
8. Peter Fickert, Heimo Ramschak, Lukaskenner, Gerald Hoefler et.al: Acute Budd-Chiari Syndrome with Fulminant hepatic failure in a pregnant woman with factor V Leiden mutation; Gastroenterology 1996; 111: 1670-1673.

AUTHORS:
1. Venkateshmurthy
2. Kayalvizhi K. B.
3. Sahajananda H.

PARTICULARS OF CONTRIBUTORS:
1. Professor, Department of Anaesthesia, Rajarajeswari Medical College & Hospital, Bangalore.
2. Assistant Professor, Department of Anaesthesia, Rajarajeswari Medical College & Hospital, Bangalore.
3. Professor & HOD, Department of Anaesthesia, Rajarajeswari Medical College & Hospital, Bangalore.

NAME ADDRESS EMAIL ID OF THE CORRESPONDING AUTHOR:
Sahajananda H,
413, Ragam, 7th Main,
Vijaya Bank Colony, Bangalore-76.
E-mail: sahaj_anand@hotmail.com

FINANCIAL OR OTHER COMPETING INTERESTS: None

Date of Submission: 12/03/2015.
Date of Peer Review: 13/03/2015.
Date of Acceptance: 13/05/2015.
Date of Publishing: 21/05/2015.