Severe preeclampsia with HELLP syndrome with Wilson's disease.

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Abstract

Severe Preeclampsia is characterized by systolic blood pressure ≥ 160 mmHg or diastolic pressure >110 mmHg, protein excretion >5 gm/24 hour or 3+ or more on random samples, oliguria <500 ml/24 hr, platelet count <1,00,000/mm^3, hemolysis, elevated alanine aminotransferase and aspartate aminotransferase, cerebral and visual disturbance, persistent severe epigastric pain, retinal hemorrhages, exudates and papilledema, intrauterine growth restriction of the fetus and pulmonary edema. HELLP (Hemolysis, elevated liver enzymes and low platelets) syndrome is the variant of preeclampsia. Definitive treatment of HELLP syndrome is delivery of the baby.

Wilson's disease is autosomal recessive disorder of copper metabolism. It is characterized by the toxic accumulation of copper mainly in liver and brain but also in cornea and kidney due to defective biliary excretion of copper. It is one of the major causes of cirrhosis of liver. Excess copper levels are associated with the disorder of the menstrual cycle and infertility. Increased copper levels are also associated with the preeclampsia, intrauterine growth restriction and neurologic damages in the fetus.

However Pregnancy is not contraindicated in well treated Wilson's disease. A case of decompensated cirrhosis due to Wilson's disease, presented in third trimester of pregnancy, with Preeclampsia and HELLP syndrome is discussed.

Keywords: HELLP syndrome, Wilson's disease, Preeclampsia, Autosomal recessive disorder, Copper metabolism, Pregnancy

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Introduction

HELLP syndrome was coined by Dr. Louis Weinstein in 1982- H (hemolysis, which is the breaking down of red blood cells) EL (elevated liver enzymes) LP (low platelet count) [1] Patients with HELLP syndrome are usually seen before term complaining of malaise (90%), epigastric or right upper quadrant pain (90%) and nausea and vomiting (50%) [2]. The Mississippi classification of HELLP is Class I (severe thrombocytopenia): platelets under 50,000/mm^3, Class II (moderate thrombocytopenia): platelets between 50,000 and 100,000/mm3, Class III (AST >40 IU/L, mild thrombocytopenia): platelets between 100,000 and150,000/mm^3 [3]. HELLP syndrome occurs in 0.1-0.6% of all pregnancies (70% of antepartumm cases and 30% of postpartum cases) [1]. HELLP syndrome occurs in 4-12% with severe Preeclamsia [1]. HELLP syndrome is considered as the complication of preeclampsia [3] Wilson's disease is autosomal recessive disorder of copper metabolism. It is characterized by the toxic accumulation of copper mainly in liver and brain but also in cornea and kidney due to defective biliary excretion of copper. It is one of the major causes of cirrhosis of liver [4]. In pregnancy excess copper levels can be associated with preeclampsia [5] Liver disease in pregnancy is uncommon but when presents it is very severe.

Case Presentation

27 years old gravida 1 para 0 married, hindu, house wife from butwal, Nepal; unbooked case supervised in butwal hospital presented with the chief complaints of amenorrhoea for 8 months, swelling of body for 1 month and focal twitching movement of hands and face; spasticity on left upper limb for 1 day. It was a spontaneous unplanned pregnancy, patient did not take folic acid and abdominal ultrasonography was not done. Patient was taking iron and calcium; however she did not receive tetanus toxoid injection. She started having swelling of body since 7 months period of gestation which started from lower limbs, later progressed to facial region, vulva and then rest of the body. She was delirious during presentation with facial twitch in the left side and twitches in the left upper limb. There was no history of loss of consciousness or seizures. Patient was married for 1.5 years and did not use any contraception. On enquiring about the patient's family history, her sibling died due to jaundice 2 years ago. There was no history of allergies to any medications and patient was non vegetarian. On general examination patient was ill looking, had poor vision with Glasgow coma scale of 15/15. Her blood pressure was 160/100 mmHg in right upper limb and 130/100 mmHg in left upper limb. Pulse rate was 84/min, axillary temperature was 98 degree F and respiratory rate was 18/min. Pitting edema was present in the bilateral lower limbs. Mild pallor was present. On abdominal examination, uterus was 28 weeks size, lie was longitudinal, cephalic presentation, fetal heart rate was 152/min, splenomegaly was present with tip of spleen palpated 14 cm below the left subcostal margin, shifting dullness was present. On Per Vaginal examination cervical OS was closed, cervix was soft and posterior, vulval edema was present and head station was high up. On ophthalmological examination no significant findings were present. On doing blood tests following results were obtained-Hb: 11 g/dL, PCV: 33% , WBC: 3190/mm^3, platelets: 27,000/mm^3, PT: 18 sec, INR: 1.5, sugar (R): 3.9 mmol/L, urea-3.2 mmol/L, creatinine-55

µmol/L, sodium: 141 mmol/L, potassium: 4.4 mmol/L, total bilirubin: 83 mg/dL, direct bilirubin: 10 mg/dL, albumin: 2.4 g/dL, SGOT: 36 U/L, SGPT: 40U/L, ALP: 177 U/L LDH: 778 U/L, fibrinogen: 451 mg/dl, D-dimer: 4800, serum copper: 191 mg/dL (70-145), Viral Serology-Non reactive, blood Culture-no growth. On urinalysis, albumin was 3+. On abdominal ultrasonography: placenta was posterior, Amniotic fluid Index (AFI) was 12.8 cm, moderate ascitis was present and there was massive splenomegaly measuring 18.1 cm, CT scan of abdomen revealed splenic artery aneurysm and cirrhosis of liver Magnetic Resonance Imaging of brain revealed T1 high and T2 low signal intensity in bilateral basal ganglia. Patient was managed in Intensive care unit, magnesium sulphate was started according to Pritchard regimen and nifedipine was given for high blood pressure. Injection dexamethasone was given. On second day of admission, due to fetal distress, emergency caesarean section was performed under general anesthesia. 3 units platelets was transfused per operatively following which platelet count rose to 53000/mm^3 postoperatively, random blood sugar level was 7.1 mmol/L postoperatively. Per-operative findings were: straw coloured ascitic fluid of 1700 ml lower uterine segment was well formed, liquor was mild meconium stained and adequate, baby presented by cephalic, placenta was fundic and anterior, bilateral fallopian tubes and ovaries were normal, enlarged spleen was present with white patches on surface. Baby was given CPR as there was no pulse rate, intubated with 3 mm endotracheal tube, there was return of spontaneous circulation, and baby was then managed in Neonatal intensive care unit. Patient was managed in the Intensive care unit post operatively. Patient was referred to the center where liver biopsy was available. Reports traced later on lead to the diagnosis of Wilson's disease and patient is now waiting for liver transplantation in that center.

Discussion

Severe Preeclampsia is characterized by systolic blood pressure ≥ 160 mmHg or diastolic pressure >110 mmHg**,** proteinexcretion >5 gm/24 hr or 3+ or more on random samples**,** oliguria <500 ml/24hr**,** platelet count <1,00,000/mm^3, microangiopathic hemolysis, elevated alanine aminotransferase and aspartate aminotransferase, cerebral and visual disturbance**,** persistent severe epigastric pain**,** retinal hemorrhages, exudates and papilledema**,** intrauterine growth restriction of the fetus and pulmonary edema [1]. HELLP syndrome is the variant of preeclampsia. Definitive treatment of HELLP syndrome is delivery of the baby. Well Compensated cirrhosis is not a contradiction for Pregnancy. Decompensated cirrhosis during pregnancy results in variceal bleeding, hepatic encephalopathy, splenic artery aneurysm and rupture, hepatic failure,

Conclusion

As presented in this case. Wilson's disease is autosomal recessive disorder of copper metabolism. It is characterized by the toxic accumulation of copper mainly in liver and brain but also in cornea and kidney due to defective biliary excretion of copper. It is one of the major causes of cirrhosis of liver. Wilsons disease was the cause of cirrhosis of liver in this patient. Excess copper levels are associated with the disorder of the menstrual cycle and infertility [6]. Increased copper levels are also associated with the preeclampsia, intrauterine growth restriction and neurologic damages in the fetus [6] However Pregnancy is not contraindicated in well treated Wilson's disease [6].

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