

Splenomegaly with Splenic Artery Aneurism Associated with Severe Preeclampsia Complicated by Massive Ascites

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ABSTRACT

A 27 year G1P0 at 31 weeks period of gestation with severe preeclampsia with HELLP Syndrome complicated by massive ascites and hypersplenism and splenic artery aneurism managed with battery of investigation, employing Cesarean Section for fetal distress is presented, emphasizing to overcome low/ timely booking status of woman still prevalent in Nepal and improvisation of preconception counseling and qualitative antenatal care for good maternal and perinatal care.

Keywords: HELLP Syndrome, Hypersplenism with splenic aneurism, severe preeclampsia

INTRODUCTION

Severe Preeclampsia (SPE) may sometimes be complicated by HELLP syndrome, latter showing features of liver dysfunction and thrombocytopenia. Thrombocytopenia may be invariably associated with splenomegaly. Splenomegaly and massive ascites are two out of three components with the exclusion of variceal bleeding forming triad of portal hypertension which was present in this unique case reported herewith, in a non-anemic Primigravida at 31 weeks with splenic artery aneurysm with fetal growth restriction (FGR) and unexplained elevated copper levels outside of Wilson's disease likely contributory factor to SPE /FGR.

CASE

A 27 year, Hindu house maker married for 1.5 years and a primigravida G1P0 from Butwal was admitted 2014 June 14 (2071/2/31) with the history of amenorrhea for eight months, swelling of body for one month, focal twitching movement of hands and face and spasticity on left upper limb for one day. There was past history cholelithiasis.

It was a spontaneously conceived unplanned pregnancy. From 2nd trimester iron and calcium was started. Quickening was perceived from 5th month. Anomaly scan was not done. TT injection was not taken. On 3rd trimester from the completed 7th months of pregnancy there was swelling of body. She was brought to the hospital when family noticed she was delirious with poor vision and twitches on left side of face and left upper limb. On general examination: temperature - (98F), pulse rate - 84/min and respiratory rate was 18/min. Bilateral pitting pedal edema was present. BP- 160/100 mm Hg (Rt. Upper limb), 130/100 mm Hg (Lt.

Upper limb). GCS - 15/15. On systemic examination: Upper and lower limbs motor 4+/5. Cerebellar signs were absent, there was no signs of meningeal irritation. On per abdominal examination, uterine fundal height corresponded to 28 weeks [POG= 31 (Fetal growth restriction)], longitudinal lie, cephalic presentation. Fetal heart rate (FHR) -152/min. Spleen felt 14 cm below left subcostal margin amidst abundance of ascites. (Fig1). Vulval edema was present on inspection of perineum. Urine showed 3+ albumin.

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Figure 1. pregnancy with splenomegaly and ascitis

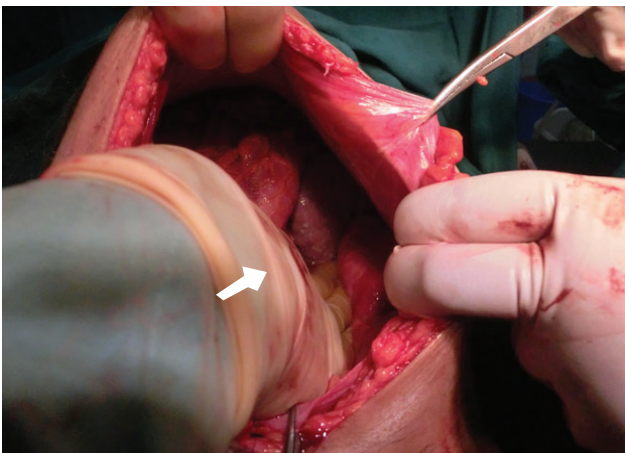


Figure 2. arrow points exposed spleen, after delivery of baby and repair of the cut ends of uterus

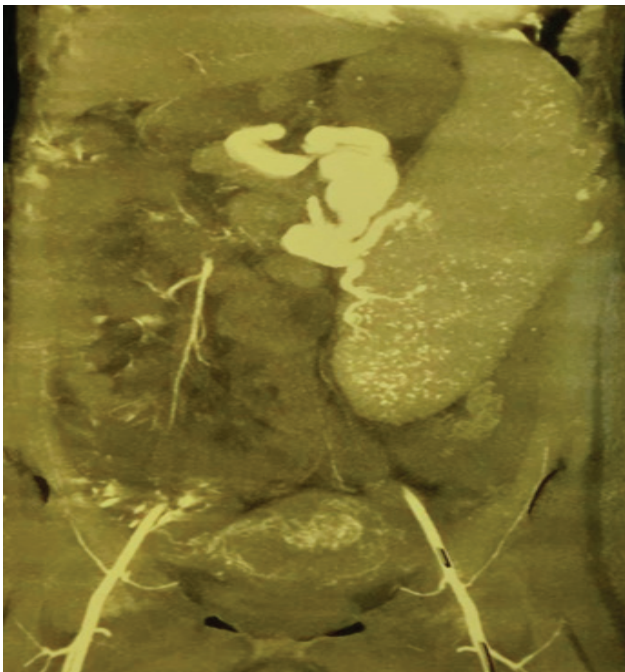


Figure 3. splenic artery aneurism

Bed side Ophthalmological findings (portable) showed evidence of severe preeclampsia without signs of retinal detachment.

Abnormal hematological investigation were: platelets - 27000, Total bilirubin - 83 $\mu\text{mol/L}$ (1.71 to 20.5 $\mu\text{mol/L}$), Direct bilirubin -10 $\mu\text{mol/L}$ (< 5.1 $\mu\text{mol/L}$); Total Protein- 56 g/L (60 to 80 g/L), Albumin- 24 g/L (35 to 52 g/L), LDH -778 IU/L (105 - 333 IU/L), d -dimer - 0.8mg/dl (< 0.50 mg/dl), serum copper -191 $\mu\text{g/dl}$ (70 - 145 $\mu\text{g/dl}$) and total calcium- 1.9 mmol/L (2.1-2.6 mmol/L)

On USG Abdomen and Pelvis, massive splenomegaly measuring 18.1 cm was noted with ascites and a live baby with AFI - 12.8 cm. Normal venous doppler study of B/L lower limbs was confirmed by Doppler USG with normal flow but dilated hepatic artery / dilated portal vein measuring 14.5mm. Multiple dilated vessels were seen in the splenic hilum of hypersplenism (20 cm). Pulmonary angiogram was normal. MRI Brain showed T1- high and T2- low signal intensity in B/L basal ganglia. Echocardiography showed mild TR/ mild PS with suspicious clot in pulmonary artery bifurcation.

She was transferred to ICU. Subsequently DBP >110 mmHg, so per oral antihypertensive, nifedipine 5mg was given. Prophylactic Mgso4 (Pritchard's regime) was started, so was Inj. Dexamethasone - 8 mg IM 8 hourly for lung maturity, and antibiotic Inj Ceftriaxone 1gm iv 12 hourly along with Inj RL and DNS.

On 2nd day of ICU admission, under general anesthesia Emergency LSCS was done for fetal distress. Per-operative findings: Straw colored ascitic fluid-1700ml, lower uterine segment well formed, adequate mild meconium-stained liquor. Baby presenting as cephalic in ROT was delivered. CPR was done for the baby and was intubated - 3mm ET tube and upon return of spontaneous circulation (ROSC) rushed to NICU. Meanwhile anterior fundic placenta was removed. Bilateral tubes and ovaries were normal. After closure of uterus, abdominal wall was exposed to view enlarged spleen with white patches on surface (Fig 2) . To avoid PPH, platelet level was maintained above 50,000 (mcL) by per-operative transfusion of 3 units of platelet rich plasma (PRP). Abdominal drain was kept thereby completing abdominal closure.

Postoperatively she was continued on inj. ceftriaxone-1gm iv 12 hourly and inj. Metronidazole 500 mg iv 8 hourly. Inj. Fentanyl - 25 microgm iv, inj. ranitidine 50 mg iv TDS, in j. RL 100 ml/hr, N-acetylcysteine 600mg PO BD, syrup lactulose 30 ml PO 8 hourly was added. Post operative Hb was 13 gm %. Platelets level increased from 27000 - 53000 per microliter. Serum creatinine was slightly increased to 113 mg/dL (0.59 to 1.04 mg/dL).

Twitching disappeared. She was stabilized. Abdominal drain was removed after three days and rest of the post operative period was uneventful. Case was discussed with GI surgeon for the further management of hypersplenism and splenic artery aneurism that was demonstrated on CTA abdomen (Fig 3). Apparently, from surgical side, was advised for follow up.

DISCUSSION

This is a case of SPE with systolic blood pressure (SBP) ≥ 160 mmHg or diastolic blood pressure (DBP) ≥ 110 mm Hg and proteinuria (urinary Alb 3+) that was noted for the first time in a previously normotensive and nonproteinuric woman (after the 20th week POG). This could have probably developed around 7th month of gestation coinciding with the occurrence of swelling of body. And was associated with HELLP syndrome {described by Dr. Louis Weinstein in 1982 [H (hemolysis, the breaking down of red blood cells) EL (elevated liver enzymes), LP (low platelet count $<1,00,000$)]} and with severe thrombocytopenia platelet of 27,000 falling in Class I HELLP according to Mississippi classification which denotes platelets under 50,000/mm.

At the outset, it's difficult to assume whether pre-eclampsia and FGR correlated to the unexplained excess copper levels outside of the Wilson's disease.¹

Massive ascites has been known to be a complication of SPE with the documentation of removal of 2L of ascitic fluid at CS section and 12 L more thereafter via abdominal drain over the next three consecutive days.²

Ascites in association of splenomegaly, two of the three components in the triad of portal hypertension (with an exception of variceal bleeding) makes the diagnosis plausible, favorably in the absence of anemia. Splenomegaly in conjunction with thrombocytopenia do result from non-cirrhotic portal hypertension.³

Splenic artery aneurysms besides being congenital may be acquired during third trimester of pregnancy as in our case, but seldom has been described coexisting with massive splenomegaly resultant to extrahepatic portal hypertension in pregnancy, giving this case a uniqueness.⁴

Splenic artery aneurysm are cited as a threat to rupture during pregnancy or post-partum, more so, with prolonged hypertension (of PE/SPE) in the background, therefore imparting a need for early diagnosis and management. During pregnancy metal embolization coil to embolize splenic artery aneurysm has been practiced with fair chance of slippage fear in the absence of splenomegaly.⁵ With splenomegaly associated with SAA as in this case, splenectomy and aneurysmectomy with a proximal splenorenal shunt seems to be best surgical option.⁴

CONCLUSIONS

In view of emergent encountering of a primigravida with multiple complications as SPE-HELLP Syndrome, massive ascites and a giant splenomegaly in association with SAA, a timely qualitative antenatal care is advisable with improvisation of preconception counseling to correctly handle medical - surgical conditions emerging before and during pregnancy for better maternal perinatal outcome.

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