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Case Report

Severe Early Onset Pre-eclampsia (PET) / HELLP Syndrome Before the age of Viability

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Abstract

HELLP syndrome is a life-threatening condition, thought to be a type of severe preeclampsia, characterized by Haemolysis (destruction of red blood cells), Elevated Liver enzymes (which indicate liver damage), and Low Platelet counts. This is a case of a woman who presented at 20+5 weeks gestation with HELLP syndrome, highlighting the occurrence of this condition before the limit of viability. This case report aims to emphasise the need to be vigilant that PET / HELLP syndrome can occur in very early gestation albeit rare. Aggressive management with magnesium sulphate, intravenous antihypertensives and pregnancy termination, in this case, prevented the poor maternal outcome. It also highlights the need to have a difficult conversation around the need for the termination of pregnancy as the only means to avoid a poor maternal outcome in fulminant cases.

ABBREVIATIONS

PET: Pre-eclamptic Toxaemia; HELLP: Haemolysis, Elevated Liver Enzymes; Low Platelets

INTRODUCTION

HELLP syndrome is considered a variant of severe PET and is associated with perinatal and maternal morbidity and mortality. It is an acronym that stands for haemolysis, elevated liver enzymes, low platelets [1]. Only 7-10% of cases occur at a gestational age less than 27 weeks and presentation at less than 21 weeks is rare [2]. Only very few cases have been reported of HELLP prior to 24 weeks gestation. Management of HELLP is multidisciplinary and usually delivery is considered essential to minimise maternal morbidity and bring about early resolution of haematological abnormalities. This case highlights the importance of being vigilant for severe PET/HELLP at earlier gestations.

CASE PRESENTATION

A 35-year-old gravida 2 para 1, at 20+5 weeks gestation presented to the Emergency Department with severe epigastric pain, nausea, vomiting and high blood pressure of 200/120 mm Hg. In her first pregnancy, seven years previously, she was delivered by an emergency section at 32 weeks gestation for severe preeclampsia. She was transferred to the labour ward where she was noted to have 3+ protein on urine dipstick. Her blood pressure remained high at 195/115mm Hg, heart rate of 80/min, respiratory rate of 20/min, O_2 saturation of 99% on air and temperature of 36.6 degrees. She had normal reflexes, no clonus and satisfactory urine output.

Initial blood workup showed low platelets, raised transaminases, blood glucose, raised PCR with normal renal function and troponin levels (see chart for blood results). Fetal heart was normal on auscultation with doptone. On oral labetalol and nifedipine her BP came down to 137/95 mmHg but subsequently increased again, therefore, she was started on intravenous labetalol for blood pressure control and intravenous magnesium sulphate for seizure prophylaxis. She remained symptomatic even on iv labetalol with worsening blood parameters.

INVESTIGATIONS

Subsequent investigations were to rule out differentials for epigastric pain. A liver scan ruled out gallbladder disease and liver haematoma. A normal troponin level and ECG ruled out cardiac causes. As the disease declared itself to be HELLP, a CT scan to rule out aortic dissection was not done (Table 1).

The blood work-up showed raised LDH consistent with haemolysis, elevated liver enzymes and low platelets confirming HELLP syndrome (Table 2).

DIFFERENTIAL DIAGNOSIS

The differential diagnoses considered at presentation were severe early-onset PET/HELLP, pancreatitis/ gallbladder disease, aortic dissection. A surgical opinion was obtained because of the raised amylase and a normal CT scan. The surgeons did not think a mildly raised amylase indicated pancreatitis. An ultrasound of the liver and gallbladder was done and reported as normal. As the renal function remained normal and there was no hypoglycaemia

Table 1: Blood work at Initial presentation.

Haemoglobin	Platelets	Urate	AST	ALT	LDH	Urea	Creatinine	Amylase	PCR
106	91	271	259	229	448	3.7	50	136	0.288

Abbreviations: LDH: Lactate Dehydrogenase; PCR: Protein Creatinine Ratio; AST: Aspartate Amino Transferase; ALT: Alanine Aminotransferase

Table 2: Blood panel done during the course of patient's stay.

	At admission	During TOP	24 hr after TOP	48 hr after TOP	6 weeks post TOP
WCC	8.9	6.3	9.7	11.8	5.5
Hb	106	100	87	96	116
Hct	0.30	0.30	0.26	0.28	0.34
Plat	91	64	117	210	247
CRP	-	-	-	81	7
Na	132	134	137	136	139
K	4.8	4.5	4.2	4.1	3.9
Urea	3.7	3.5	3.3	2.4	6.7
Creat	50	52	55	56	58
Bili	5	5	3	4	3
AST	259	146	54	52	18
ALT	229	205	124	89	10
Albumin	37	34	33	40	48
LDH	448	369	-	-	-
PT	10	10	10	10	11
aPTT	25	24	24	22	28
Fibrinogen	>5	4.84	>5	>5	2.24
Lactate	1.5	-	1.3	-	-
PCR	0.624	-	-	-	0.009
Urate	271	-	386	304	-
Amylase	136	128	-	-	-

Abbreviations: WCC: White Cell Count; Hb: Haemoglobin; Hct: Haematocrit; Plat: Platelets; CRP: C Reactive Protein; Na: Sodium; K: Potassium; Creat: Creatinine; Bili: Bilirubin; AST: Aspartate Amino Transferase; ALT: Alanine Amino Transferase; LDH: Lactate Dehydrogenase; PT: Prothrombin Time; Aptt: Activated Partial Thromboplastin Time; PCR: Protein Creatinine Ratio

or coagulopathy, Haemolytic Uraemic Syndrome and Acute Fatty Liver of Pregnancy were ruled out. The mother's blood pressure increased while awaiting CT scan and her platelets dropped further and a diagnosis of fulminant PET / HELLP syndrome was made.

TREATMENT

When symptoms stabilised, the woman and her partner were counselled about the poor prognosis in such an early-onset disease and consented to termination of pregnancy. This was carried out using mifepristone and misoprostol. She was also seen by the bereavement team. A stillborn baby was delivered 24 hours later. The mother's biochemical and haematological markers improved steadily following delivery. Her intravenous labetalol was changed to oral nifedipine 20 mg twice daily. She spiked a temperature after 48 hours which was thought to be due to breast engorgement and started on antibiotics. She had an echocardiogram on day 5 which was normal and she was discharged home on oral antibiotics.

OUTCOME AND FOLLOW-UP

Once intravenous magnesium sulphate was started and the termination process commenced, the haematological and biochemical abnormalities other than low platelets stabilised. They further improved post-delivery and at her six-week review, her platelets and LFTs were normal. She had stopped the nifedipine at this stage.

She was seen for counselling, a few months post-delivery where her chances of recurrence and long term possible morbidity were discussed. Studies show that there is a 33% chance of recurrence of preeclampsia if the birth occurred between 28 and 34 weeks. As per NICE [3] 'No evidence was identified for women who gave birth at less than 28 weeks, but the committee agreed that the risk was likely to be at least as high, if not higher, than that for women who gave birth between 28 and 34 weeks'. As this was the mother's second pregnancy complicated by severe preeclampsia (despite aspirin 150 mg started on booking) she was counselled that her recurrence risk might be higher than 30% in a subsequent pregnancy.

DISCUSSION

HELLP is a syndrome characterized by thrombocytopenia, haemolysis, and liver dysfunction. The acronym was coined by Louis Weinstein in 1982. It is believed to result from microvascular endothelial activation and cell injury. HELLP syndrome is estimated to complicate 0.1% to 0.8% of pregnancies, while 10% to 20% of HELLP occur with severe pre-eclampsia [4]. HELLP is widely considered to be a severe form of pre-eclampsia yet up to 15–20% of patients with HELLP do not have antecedent signs or symptoms of pre-eclampsia [5].

The pathophysiology of HELLP syndrome is not known. It is theorized that because HELLP is a variant of preeclampsia, the pathophysiology is similar. Aberrant placental implantation and placental oxidative stress leads to insufficient placental function and release of factors into maternal circulation resulting in endothelial dysfunction and activation of leucocyte, complement and coagulation pathways leading to the clinical syndrome of preeclampsia and HELLP [6].

Only a handful of case reports have been published on PET/HELLP at gestations less than 22 weeks [7-9]. The largest case series of women with severe early-onset hypertension at limits of viability over a fourteen-year period from the Netherlands mentions only 22 women at less than 22 weeks [10]. Other cases reported in the literature of severe PET/HELLP prior to 20 weeks gestation are usually associated with fetal triploidy or anti-phospholipid syndrome.

With improved perinatal care of very preterm infants, some authors favour a policy of prolonging pregnancy in selected cases. Ganzevoort [11], and Sibai [12], have both reviewed different management regimes and concluded that before 24 weeks of gestation, because of the absence of perinatal benefits and high maternal complication rate, an expectant management approach should not be offered routinely.

NICE guidelines on hypertension in pregnancy mentions various thresholds for considering planned early birth in severe pre-eclampsia which includes progressive deterioration in liver function, renal function, haemolysis, or platelet count. Given that our patient presented with HELLP at 20+5 weeks gestation and the deterioration in the biochemical picture, termination of pregnancy was advocated to prevent maternal morbidity/mortality.

TAKE HOME MESSAGES

- This case illustrates that obstetricians should be alert about the possibility of the early-onset severe PET / HELLP syndrome, albeit rare and aggressive management via a multidisciplinary approach should be undertaken to prevent maternal morbidity and mortality. Fulminant PET/HELLP syndrome may occur at very early gestations albeit rare.
- Aggressive management must be pursued to prevent maternal morbidity and mortality.
- Termination pregnancy is the appropriate treatment for severe HELLP syndrome and women should be appropriately counselled and offered support to undertake this very difficult decision.

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