Review Article

A High Light of Hellp Syndrome-Review

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ABSTRACT
Liver disease may manifest during pregnancy, and the condition of the disease necessitates careful examination and management. HELLP Syndrome is a syndrome with series of symptoms that can affect pregnant women. HELLP syndrome will always occurs in connection with Preeclampsia and 4-12% of women with diagnosed preeclampsia will develop Hellp syndrome. Hellp syndrome is characterized by Haemolysis (H), Elevated Enzyme Level (EL), Low Platelet Count (LP).Affected women shows sign of liver damage and abnormalities in blood clotting. In extreme cases the outcome is fatal. This article high light the Etiology, Complication, Diagnosis, Presentation, Management and Prevention of HELLP Syndrome.

Keywords: Introduction, Classification, Aetiology, Epidemiology, Sign and Symptoms, Diagnosis.

INTRODUCTION

Diseases of pregnancy and pathological condition of placenta are important causes of intrauterine death, congenital malformations intra uterine growth retardation, maternal death and a great deal of morbidity to both mother and child[1, 2].

One among the disorder which manifests during pregnancy is being the Hellp Syndrome .It is characterized by H- haemolysis, Elevated Enzyme Level (EL) and Low Platelet Count (LP).This syndrome usually present in women who have “Preeclampsia” or “Eclampsia”, with varying symptoms of liver damage and the abnormalities in blood clotting. Hellp Syndrome occurs in 0.5 -0.9 % of all pregnancies and in 10 – 20 % of the cases with severe Preeclampsia. It is a serious complication of Pregnancy and occurs in any time at the last half of the pregnancy, Non-specific symptoms include Nausea, Vomiting, Fatigue, Epigastric Pain, with Edema and hypertension. Diagnostic features of Hellp syndrome include the determination of Liver Function Test, Platelet Count, Clotting and Bleeding Time, LDL Level Spotting.

Hellp Syndrome is a life threatening obstelic complication usually considered to be a variant or complication of preeclampsia. Hellp syndrome was identified as a distinct clinical entity coined by Dr. LOUIS WEINSTEIN In 1982.[3]

Till date no common precipitating factor of syndrome was dictated and it is found to be manifestation of some insult leading to micro vascular endothelial damage and intra vascular platelet activation.

Prompt recognition and timely initiation of therapy are vital to ensure best outcome of mother and foetus and the treatment profile was based on estimated gestational age and the condition of mother and foetus.

Treatment profile includes the routine treatment with corticosteroid conservative management of hypertension and fluid management with close observation Prophylactic treatment invites a bolus of 4 – 6 gm of $Mgso_4$ as 20% solution of which should be maintained as dose at 2g/hour to prevent seizures.

This article emphasizes the classification, Risk Factors, Diagnosis, and Complications, Clinical Presentations Treatment Profile of Hellp Syndrome.

Classification

Two Classification systems are used for HELLP Syndrome. The First Classification was based on No of complication present as Partial which shows any one or two abnormalities and full which shows all the complications. Including Disseminated Intra Vascular Coagulopathy (DIC).

The second way of classification was based on the No of Platelet count as

Class I – Platelet count >50000/mm and it is featured with severe thrombocytopenia.

Class II – Platelet count ranges from 50,000 to 1 lakhs with moderate thrombocytopenia.
Class III – Platelet count ranges from 1 – 1.5 lakhs (i.e.) with mild thrombocytopenia.[4, 5]

Aetiology of Hellp Syndrome
The causes of helps syndrome are unknown, the symptoms were found to be the final manifestation of some insult leading to microvascular platelet activation. Platelet activation stimulates Thromboxane A Release and serotonin release which causes vasospasm, Platelet agglutination, Aggregation and endothelial damage.

Epidemiology
This syndrome occurs in 0.5 – 0.9 % of all pregnancies and in 10 -20 % of cases with Preeclampsia. Caucasian race was found to be sensitive in Hellp Syndrome and it was generally present in the third trimester of pregnancy (or) in mother with less than 27 weeks of gestation in 11 % of patient in post-natal cases the onset is within 48 hrs of delivery with fasting symptoms apparent for 7 days.

Sign and Symptoms
Hellp Syndrome is a serious complication of pregnancy and may present at any time in the last half of pregnancy.
70% of cases peak between 27 – 37 weeks of gestation but it occurs earlier or later.[6] 30% of women with Hellp Syndrome exhibit after 48 hrs after delivery.
The Haemolysis of Hellp Syndrome is a Micro Angiopathic Haemolysis. RBC becomes fragmented as they pass through small blood vessels with endothelial damage and fibrin deposits.
Elevated Enzyme Level are thought to be secondary to hepatic blood flow by fibrin deposits in sinusoids. This obstruction leads to per portal necrosis and in severity leads to intrahepatic haemorrhage, Sub capsular hematoma (or) hepatic rupture.
Mild moderate and severe thrombocytopenia was been attributed to increase consumption (or) destruction of platelets.
All patients may have underlying coagulopathy which usually undetectable even though DIC is the primary process in HS but it is not spectated during pregnancy.
Common symptoms includes Headache, Nausea and Vomiting Flu like syndrome upper right quadrant abdominal pain (or) tenderness hepatomegaly fatigue (or) Malaise, high blood pressure, edema, proteinuria symptoms was characterization exacerbation during night and relief during day. Increase risk spectates pulmonary embolism and reactionary haemorrhage 84 % of patients was affected by acute Renal failure[7]. Fluid retention and excess weight gain increased bleeding time leading to nonstop bleeding.
As liver being the main site of this process downstream liver cell suffers ischemia leading to per portal necrosis.

Rare Seizures and Convulsions
Serious complications include. Placental abruption, pulmonary edema,Adult and neonatal respiratory distress syndrome, Ruptured liver hematoma, Acute Renal Failure, Intrauterine growth restriction.

Risk Factors
19-27 % of reoccurrence in patients with previous history of Preeclampsia pregnancy.
PIH (pregnancy induced hypertension).
Multiparous pregnancy or null parity.
Aged Pregnancy.
Caucasian race women.

Anti-Phospholipid Syndrome (APS)
10.5 % of patients with Hellp Syndrome have APS[8].
Plasma Glutathione S Transferase.
a1 (α GST or GST a1) a very sensitive marker for liver damage.
In Hellp syndrome following parameters shows abnormalities.

LHD 600 > Iu/L
Platelets < 100000
AST or ALT > 150 IU/L
Uric acid > 7.8 MG/ 100 ML
(> 460 µ/ mol / L)[9, 13],

Diagnosis
In women with suspected Hellp Syndrome. A batch of blood test was performed.
Blood test to determine full blood count a coagulation panel, liver enzymes, electrolytic and renal function test.
Markers of Hellp Syndrome includes lactate dehydrogenase which elevates above > 600U/L
Finding Serum haptoglobin may confirm ongoing haemolysis abnormal prothrombin time thromboplastin, fibrinogen level result in the case of DIC[11].
Fibrin degradation product mild elevated arterial hypertensions belong to markers to Hellp Syndrome other criterion in diagnosing this symptom includes.
Abnormal peripheral smear
Bilirubin above 1.2 mg/ dl
Above factors represent haemolysis
Increases aspartate amino transferase >70u/l
Decrease low platelet count
CT scan for hepatic imaging may reveal liver bleeding hematoma foetal non stress test and

ultra sound may be done to reveal foetal health
Positive ID Dimer tests sensitive indicators of subclinical coagulopathy. Lung Function of baby if the gestational age is above 34 weeks for safer delivery preparation[9,10].

Treatment of Hellp syndrome

Treatment of helps syndrome was primarily based on gestational age and the condition of mother and foetus.
Corticosteroids was recommended for treatment of ANS with above 34 weeks of gestation and it is instituted in patients with platelets count > 100000 Decadron 10 mg/Every 12 hrs to improve labour abnormalities. Blood and coagulation test confirms the selection for transfusion of red cells, platelets fresh frozen plasma and cryoprecipitate of fibrinogen concentrate[9, 11]
Anti-hypertensive (Labetalol, Hydralazine, Nifedipine was recommended for conservative management of BP.
Patients with helps syndrome needs prophylactic treatments with $Mgso_4$ (4-6 gm in 20% solution) as bolus administration and the dose is maintained as 2 g/hrs to prevent seizures. $Mgso_4$ may be replaced with Calcium Gluconate it toxicity occurs. Oliguria responds to fluid placental abruption and cerebral management[11, 13].
Hypertensive crisis may be treated with infusion of Nitro-Glycerine or Sodium Nitro Prusside[12].

Prognosis

Are often good if the problem is diagnosed and treated easier
Premature labour, pulmonary problem for both mother and foetus and other serious problems may occur if this syndrome was not treated properly.
Maternal mortality was about 1% and perinatal mortality was found to be 7.3 % to 11.9 %[5].
Gestational age determines severity of disease and outcome of the baby placental abruption acute renal failure liver hematoma and damage retinal detachments occurs in 20 % of women an autosomal recessive inherited abnormality[13].
LCHAD (Long chain hydroxyl acyl co A-dehydrogenase) deficiency of child and maternal HELLP of acute fatty liver during pregnancy results in significant morbidity and mortality[8,9.]
No proven prophylactic measures was recommended following measures may reduce morbidity and mortality

Prevention

Earlier diagnosis and treatment
Staying vigilance in diet and exercise
Maintaining blood Pressure
Screening for APS
(Anti phospholipid syndrome)[12].
Screening for fatty liver, hypertrophic vascular disease and inherited diseases.

CONCLUSION

This article, concludes that the maternal mortality and morbidity rates, problems in pregnancy outcomes and labour, exculpated as a result of insults to various organ tissues one of such problem is Hellp syndrome, where there is highlighted haemolysis, Elevated Enzyme Level and low Platelet Count and the etiology may be pathological or abnormal function of organ before gestation
Severity of disease ranges from mild to severe based on platelet count which depends on gestational age and the condition of mother and foetus. Blood counts, BP estimation calls for the prognosis of disease after the response to treatment. Liver enzyme abnormalities and pathology can be treated with corticosteroids followed by conservative treatment with Antihypertensive and fluid management to subside oliguria
Periodical screening and care help to give a test outcome of mother and child with Hellp Syndrome.

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