ICU Management of HELLP Syndrome

Essay

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In ICU

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بسم الله الرحمن الرحيم

﴿وقُل رَّبِّ زِدْنِي عِلْماً﴾

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Abstract

Despite the development of tertiary care facilities intensive care and advanced blood banking techniques, maternal and neonatal deaths continue to occur in association with HELLP syndrome.

HELLP syndrome Characterized by hemolysis, elevated liver enzyme levels and low platelet count. The syndrome has been considered a variant of preeclampsia, but it can occur on its own or in association with preeclampsia. It is an obstetric complication that is frequently misdiagnosed at initial presentation.

The pathogenesis of HELLP syndrome isn't well understood. It's attributed to abnormal vascular tone, vasospasm and coagulation defects. The syndrome seems to be the final manifestation of some insult that leads to microvascular endothelial damage and intravascular platelet activation that leads to a cascade that is only terminated with delivery.

The vague nature of the presenting complaints can make the diagnosis of HELLP syndrome frustrating to physicians. Approximately 90% of patients present with generalized malaise, 65% with epigastric pain, 30% with nausea and vomiting, and 31% with headache, any pregnant woman who presents with malaise or a viral-type illness in the third trimester should be evaluated with a complete blood cell count and liver function tests.

Two classification systems are used for HELLP syndrome. The first is based on the number of abnormalities that are present. In this system, patients are classified as having partial HELLP syndrome (one or two abnormalities) or full HELLP syndrome. The other classification is based on platelet count: class I, less than 50, 000 per mm^3 ($50x10^9$ per L); class II, 50, 000 to less than 100, 000 per mm^3 (50 to 100 x 10^9 per L); class III, 100, 000 to 150, 000 per mm^3 (100 to 150 x 10^9 per L).

Key warders: ICU: Intensive care unit; HELLP: hemolysis elevated liver enzymes low platelets; CS: corticosteroid. RDS: Respiratory distress syndrome.

Introduction

Despite the development of tertiary care facilities intensive care and advanced blood banking techniques, maternal and neonatal deaths continue to occur in association with HELLP syndrome ***(Schlembach et al., 2003)***.

HELLP syndrome Characterized by hemolysis, elevated liver enzyme levels and low platelet count. The syndrome has been considered a variant of preeclampsia, but it can occur on its own or in association with preeclampsia. It is an obstetric complication that is frequently misdiagnosed at initial presentation ***(Fitzpatrick et al., 2014)***.

The pathogenesis of HELLP syndrome isn't well understood. It's attributed to abnormal vascular tone, vasospasm and coagulation defects. The syndrome seems to be the final manifestation of some insult that leads to microvascular endothelial damage and intravascular platelet activation that leads to a cascade that is only terminated with delivery ***(American College of Obstetricians and Gynecologists, 2013)***.

The vague nature of the presenting complaints can make the diagnosis of HELLP syndrome frustrating to physicians. Approximately 90% of patients present with generalized malaise, 65% with epigastric pain, 30% with nausea and vomiting, and 31% with headache, any pregnant woman who presents with malaise or a viral-type illness in the third trimester should be evaluated with a complete blood cell count and liver function tests ***(Woudstra et al., 2010)***.

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having partial HELLP syndrome (one or two abnormalities) or full HELLP syndrome. The other classification is based on platelet count: class I, less than 50, 000 per mm^3 ($50x10^9$ per L); class II, 50, 000 to less than 100, 000 per mm^3 (50 to 100 x 10^9 per L); class III, 100, 000 to 150, 000 per mm^3 (100 to 150 x 10^9 per L) ***(Hupuczi et al., 2007)***.

Once the diagnosis of HELLP syndrome has been established the best markers to follow are the maternal lactate dehydrogenase level and the maternal platelet count ***(Murrany et al., 2011)***.

Prompt recognition of HELLP syndrome and timely initiation of therapy are vital to ensure the best outcome for mother and fetus when the syndrome was first described prompt delivery was recommended ***(Murrany et al., 2011)***.

Antenatal administration of dexamethasone (Decadron) in a high dosage of 10 mg intravenously every 12 hours has been shown to markedly improve the laboratory abnormalities associated with HELLP syndrome ***(Woudstra et al., 2010)***.

Mortality rate for women with HELLP syndrome is approximately 1.1%. From 1 to 25% of affected women develop serious complications as DIC, placental abruption, ARDs, hepatorenal failure …. also infants affected by HELLP syndrome likely to intrauterine growth retardation, RDS ***(Murrany et al., 2011)***.

Aim of the Essay

To identify early diagnosis and prompt management of HELLP syndrome because the morbidity and mortality rates associated with the syndrome have been reported be as high as 25%.

Chapter (1)

HELLP Syndrome

Definition:

HELLP syndrome is a rare but serious illness in pregnancy. HELLP stands for Hemolysis, Elevated Liver enzymes levels and Low Platelet count, many cases of HELLP syndrome are associated with preeclampsia or eclampsia ***(Strock, 2012).***

It has been known for a long time that preeclampsia may be associated with hemolysis, elevated liver enzymes and thrombocytopenia, Weinstein regarded signs and symptoms to constitute an entity separated from preeclampsia and named the condition HELLP syndrome ***(Weinstein, 2005).***

The exact cause of HELLP syndrome remains unknown, making it difficult to determine which women will be affected ***(Strock, 2012).***

HELLP syndrome is associated with increased maternal morbidity and mortality, it is usually presented after 20^{th} week of gestation, its onset earlier than 28 weeks is rare to occur ***(August and Sibai, 2012).***

Classification:-

HELLP syndrome can be classified into two types according to the presence of the triad signs, complete and partial HELLP:-

Complete form of HELLP requires the presence of all three components, while partial form consists of only two

elements of the triad (Hemolysis, Elevated Liver enzymes or Low platelets) ***(Sibai, 2004).***

HELLP is a serious condition in its complete form and is associated with substantial risk for the mother and her foetus ***(Martin et al., 2006).***

On the other hand, partial form of HELLP has fewer symptoms and develop less complications than the complete form ***(Celik et al., 2003).***

However, a partial or incomplete HELLP may develop to a complete form of the disorder ***(Sibai, 2004).***

Another classification of HELLP according to Mississipi classes which classify HELLP into three classes according to laboratory findings of the triad as follows:

- Class (1): platelets <50, 000 /ul, AST or ALT >70IU/L, LDH >600IU/L
- Class(2): platelets 50, 000-100, 000/ul, AST or ALT >70 IU/L, LDH>600IU/L
- Class (3): platelets 100, 000-150, 000 / ul, AST or ALT >40 IU /L, LDH>600 IU/L ***(Martin et al., 2006).***

Risk Factors For HELLP Syndrome

Include the following factors:
1) Maternal age older than 34 years
2) Multiparity
3) White race or European
4) History of poor pregnancy outcome

(Lichtman et al., 2010).

Also, previous pregnancy with HELLP associated with 19-27% chance of recurrence in each pregnancy. Preeclampsia or pregnancy induced hypertension increase the risk of developing HELLP ***(Oudejans and van Dijk, 2008).***

Table (1): Comparison of Risk Factors for HELLP Syndrome and Preeclampsia:

Pre-eclampsia	HELLP syndrome
Nulliparous	multiparous
Marenal age less than 20 years or greater than 45 years	maternal age older than 25 years
Family history of pre-eclampsia	white race
Minimal prenatal care	history of poor pregnancy outcome
Diabetes mellitus	
Chronic hypertension	
Multiple gestation	

(Ertan et al, 2002)

Epidemiology:

HELLP syndrome occurs in 0.1% - 0.8% of all pregnancies and in 10% - 20% of patients with preeclampsia or eclampsia ***(VanOostwaard and Langenveld, 2015).***

HELLP syndrome typically occurs between 27th of gestation and delivery, or immediately postpartum in 15%-30% of cases ***(Ukomadu et al., 2009).***

The mean age of pregnant women with HELLP syndrome is usually higher than in women with preeclampsia ***(Celik et al., 2003).***

In the post-partum period the HELLP syndrome usually develops within the first 48 hours in women who have had proteinuria and hypertension prior to delivery ***(Karumanchi et al., 2005).***

The majority of women with HELLP syndrome have had hypertension and proteinuria, which may be absent in 10-20% of the cases ***(Barton and Sibai, 2004).***

Excessive weight gain and generalized oedema precede the syndrome in more than 50% of the cases ***(Sibai, 2004).***

Pathogenesis

The pathophysiologic mechanisms potentiating the development of HELLP syndrome are the following:

1) Haemolysis

Is one of the major characteristics of the disorder which is due to a microangiopathic haemolytic anaemia (MAHA). Red cell fragmentation caused by high- velocity passage through damaged endothelium appears to represent the extent of small vessel involvement with intima damage, endothelial dysfunction and fibrin deposition. Presence of fragmented or contracted red cells with specula (Burr cells) in the peripheral blood smear reflects the hemolytic process and strongly suggests the development of microangiopathic haemolytic anaemia ***(Martin et al., 2006).***

Destruction of red blood cells by haemolysis causes increased serum lactate dehydrogenase (LDH) levels and decreased haemoglobin concentrations ***(Matern, 2013).***

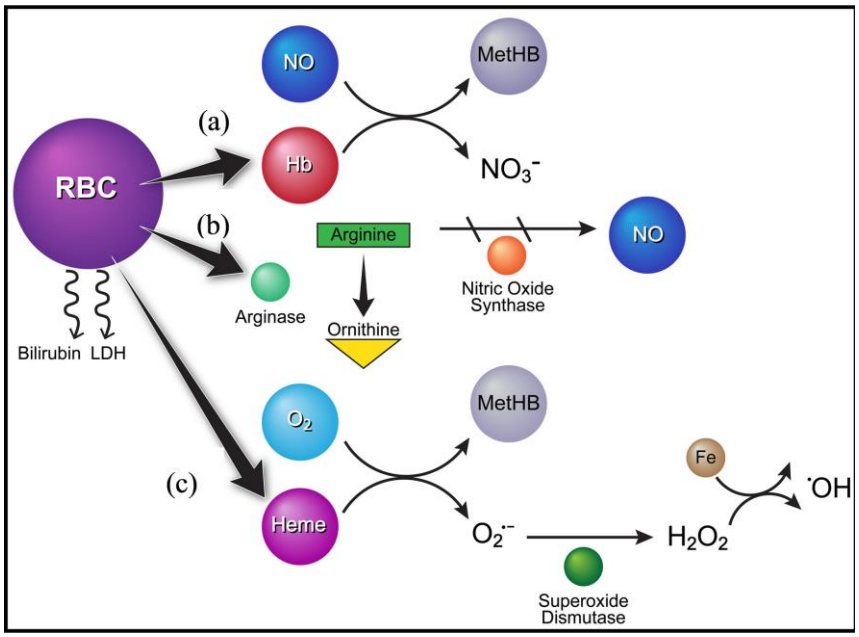

Fig. (1): Haemolysis *(Matern, 2013).*

2) Elevation of liver enzymes:

It may reflect the haemolytic process as well as liver involvement. Plasma glutathione S-transferase-a1 (GST-a1) may provide a more sensitive indicator for acute liver damage than AST and ALT and allow earlier recognition ***(Derulle et al., 2006).***

3) Thrombocytopenia:

Decreased platelet count in HELLP syndrome is due to their increased consumption. Platelets are activated, and adhere to damaged vascular endothelial cells, resulting in increased platelet turnover with shorter lifespan ***(Baxter and Weinstein, 2004).***

Genetic Aspects of Hellp Syndrome:

Many genetic factors involved in development of HELLP syndrome as follows: