



THE HELLP SYNDROME- AN ASYMPTOMATIC PRESENTATION- A CASE REPORT WITH LITERATURE REVIEW

(1) DR TABINDA BASHIR, Specialist Family Medicine, PHCC Qatar.
(drtabinda@doctors.net.uk)

(2) DR MOHAMMED ABDUL HASEEB, Specialist Family Medicine, PHCC Qatar.
(drhaseeb786@gmail.com)

(3) Dr MUHAMMAD SHARIF, Consultant Physician, Hazm Mebaireek General Hospital, Qatar.
(drsharif@hotmail.com)

1. ABSTRACT

The acronym HELLP refers to a syndrome in pregnant and postpartum women that is characterised by **H**emolysis, **E**levated **L**iver enzymes and **L**ow **P**latelet count [1]. It is thought to represent a severe form of pre-eclampsia, but the relationship between these disorders remains controversial. HELLP may be a separate entity from pre-eclampsia as in 15-20% of cases no preceding history of hypertension or proteinuria is noted [2].

It is one of the rare but serious disorder of pregnancy associated with high maternal and fetal mortality and morbidity. The mother is at risk of placental abruption, liver rupture, renal failure, shock lungs, disseminated intravascular coagulation (DIC) while neonatal complications are mainly related to gestational age at the time of delivery which is usually preterm. The timely diagnosis and management of HELLP syndrome reduce the maternal and perinatal mortality [3].

The presentation can be of acute onset with rapid progression. There is usually a short history of acute upper abdominal pain with nausea, vomiting or malaise. The initial disease process can be asymptomatic, and the diagnosis can be made on abnormal biochemical markers.

Our case demonstrates the asymptomatic presentation of a 36 years old pregnant patient who attended the health centre at 38 weeks of pregnancy for routine antenatal blood tests. Her initial complete blood picture showed thrombocytopenia. She had further investigations for low platelet count which revealed elevated liver enzymes and lactate dehydrogenase (LDH) levels with abnormal peripheral film indicative of intravascular hemolysis. She was referred to the obstetric/gynaecology emergency department and urgent delivery planned to prevent further progression of this syndrome and fatal outcomes.

KEY WORDS: HELLP syndrome, Pre-eclampsia, hemolysis, DIC



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نبذة مختصرة:

يشير الاختصار HELLP إلى متلازمة في النساء الحوامل وبعد الولادة تتميز بانحلال الدم ، وارتفاع إنزيمات الكبد وانخفاض عدد الصفائح الدموية (1). يُعتقد أنه يمثل شكلاً حاداً من أشكال مقدمات الإصابة بالتهاب الكلى ، لكن العلاقة بين هذه الاضطرابات لا تزال مثيرة للجدل. قد يكون HELLP كياناً منفصلاً عن مرحلة ما قبل الإكلامبسيا حيث أنه في 15-20% من الحالات لم يتم ملاحظة تاريخ سابق لارتفاع ضغط الدم أو بيلة بروتينية (2). وهو أحد اضطرابات الحمل النادرة والخطيرة المرتبطة بارتفاع معدل وفيات الأمهات والجنين واعتلالهم. الأم معرضة لخطر الإصابة بانفجار المشيمة والفشل الكلوي وصدمة الرئتين وتمزق الكبد بينما ترتبط المضاعفات الوليدية بشكل أساسي بعمر الحمل وقت الولادة والذي يكون عادة قبل الأوان. إن التشخيص والعلاج في الوقت المناسب لمتلازمة HELLP يقلل من وفيات الأمهات والفترة المحيطة بالولادة (3). يمكن أن يكون العرض التقديمي حاداً مع تقدم سريع. عادة ما يكون هناك تاريخ قصير للألم الحاد في الجزء العلوي من البطن مع الغثيان والقيء أو الشعور بالضغط. يمكن أن تكون عملية المرض الأولية بدون أعراض ويمكن إجراء التشخيص على علامات بيوكيميائية غير طبيعية. توضح حالتنا عرضاً بدون أعراض لمريضة حامل تبلغ من العمر 36 عاماً ذهبت إلى المركز الصحي في الأسبوع 38 من الحمل لإجراء اختبارات الدم الروتينية قبل الولادة. أظهرت صورتها الأولية للدم الكامل قلة الصفائح. كان لديها مزيد من التحقيقات لانخفاض عدد الصفائح الدموية التي كشفت عن ارتفاع مستويات إنزيمات الكبد ونزعة هيدروجين اللاكتات (LDH) مع فيلم محيطي غير طبيعي يشير إلى انحلال الدم داخل الأوعية. تمت إحالتها إلى قسم طوارئ التوليد / أمراض النساء بحيث يمكن التخطيط للولادة العاجلة لمنع المزيد من تطور هذه المتلازمة مما يؤدي إلى نتائج مميتة.

الكلمات المفتاحية: متلازمة هيلب ، تسمم الحمل ، انحلال الدم

2. INTRODUCTION

HELLP syndrome is a rare but one of the life-threatening complications of pregnancy with high maternal and fetal morbidity and mortality. It occurs in 0.5%-0.9% of all pregnancies and in 10-20% of women with severe pre-eclampsia [4]. Unfortunately, if the pre-eclampsia is not present, the diagnosis of HELLP is often delayed [5]. HELLP syndrome typically occurs between 27 weeks of gestation and delivery, or immediately in the postpartum period in 15%-30% of cases [6]. Various life-threatening complications such as placental abruption, pulmonary oedema, cerebral haemorrhage, hepatorenal failure and disseminated intravascular coagulation (DIC) can occur in these patients and fetal poor outcome could be related to preterm delivery. Early diagnosis and intervention can prevent these serious and life-threatening complications.

We present an unexpected asymptomatic case of HELLP syndrome with abnormal biochemical markers in which an early intervention led to the favourable outcome. Asymptomatic thrombocytopenia is common in pregnancy. The objective of this case report is to highlight the importance of having a low threshold for further urgent investigations if abnormal blood results including low platelet count are found on routine antenatal investigations. Early intervention can lead to early detection and prompt actions prevent the development of a serious medical illness with fatal consequences as in our case.

3. CASE REPORT

A 36 years old lady presented to the health centre for routine antenatal check-up at 38 weeks of gestation. Her pregnancy so far had been uneventful. She had no previous antenatal visits for the current pregnancy. This was her 3rd pregnancy with no previous significant obstetric history.

Her vital signs were normal including blood pressure of 110/76 mm of Hg. There was no evidence of protein in the urinary analysis. Obstetric examination revealed gravid uterus of 36-38 weeks with regular fetal heart sounds and cephalic presentation.

Routine antenatal screening tests were ordered including blood tests, urine tests and ultrasound scan. Her complete blood picture showed thrombocytopenia of 106,000 per mm³. Further investigations were ordered in view of low platelet count which revealed raised bilirubin of 36 micromol/L, Alanine aminotransferase (ALT) of 276 U/L and Aspartate aminotransferases (AST) 254 U/L. The lactate dehydrogenase (LDH) was 289 U/L.

The peripheral film showed few target cells, 1-2 Schistocytes per high power field, thrombocytopenia, and toxic granules in neutrophils.

A diagnosis of HELLP syndrome was made and she was urgently referred to an obstetric emergency department. Delivery was induced the following day, delivering a healthy male child weighing 2.9 kilogramme.

4.DISCUSSION

The Acronym HELLP syndrome was first used by Louis Weinstein in 1982. He reported the laboratory findings of this syndrome in 29 patients with eclampsia and pre-eclampsia [7]. The precise aetiology of this disease entity remains an area of controversy to date. Few well known maternal risk factors are multiparity (>4), women living in rural areas, age > 35 years, pre-pregnancy hypertension and high BMI [8]. Interestingly, age less than 25 years has an inverse relation to the risk of developing HELLP Syndrome.

Many experts consider HELLP syndrome as a part of the spectrum or a variant of eclampsia. It is considered to be a multi-system disease causing a generalised vasospasm, microthrombi formation and coagulation defects [2]. It is a disorder involving placenta that shares the

histopathological, placental morphological features and changes in gene expression with the early onset pre-eclampsia [9].

The trigger for HELLP syndrome is the endothelial damage causing activation of coagulation cascade and platelet aggregation that would explain the thrombocytopenia. Platelet-fibrin deposits in the capillaries are responsible for microangiopathic haemolysis as red blood cells pass through [10]. Similarly, the fibrin deposits within hepatic sinusoids leads to hepatic blood flow obstruction causing periportal necrosis. In more severe obstruction, this can lead to intrahepatic bleeding and or subcapsular haematoma formation.

In two percent of patients with the HELLP syndrome, deficiency of fetal long-chain 3-hydroxyacyl CoA dehydrogenase (LCHAD) deficiency is noted,[11] however, screening with genetic analysis is not warranted as this does not change the management.

The onset of HELLP syndrome is usually rapid [12]. Typical clinical presentation includes upper abdominal pain of acute onset, usually in the right upper quadrant or epigastric region with nausea and vomiting. Many Patients experience malaise few days prior to seeking medical attention [13].

Less common symptoms include headache, visual symptoms, jaundice and ascites. The initial disease process can be asymptomatic and in some patients the diagnosis is made on routine evaluation of unexplained thrombocytopenia as was in our case.

The diagnosis of HELLP syndrome involves the presence of all three components including **Hemolysis, Elevated Liver enzymes and Low Platelet** count. Patients with evidence of all three components are diagnosed with complete/full HELLP syndrome. In incomplete/partial HELLP syndrome, only 1 or 2 components of the triad can be present [12].

There needs to be a high index of suspicion to avoid delay in the diagnosis of the HELLP syndrome and improve outcomes for both the mother and the foetus. This is particularly important in any pregnant patient in the second and third trimester or immediately in the

postpartum period, who presents with symptoms of upper abdominal pain. The women with partial HELLP syndrome may not have typical symptoms and may develop less complications. However, it can progress to the complete form of the disease [12].

Platelet count is the best indicator of HELLP syndrome. Isolated thrombocytopenia may be one of the first clues to the diagnosis as discussed above. As was observed in our patient, thrombocytopenia was the first abnormality noted that led to further investigations and identification of abnormal liver functions, hemolysis and abnormal peripheral film.

Other findings may include drop in haemoglobin, presence of Schistocytes and Burr cells, raised LDH levels, low haptoglobin and increased reticulocyte count-all pointing towards intravascular hemolysis [14,15]. Abnormalities of liver enzymes may indicate hemolytic process as well as liver involvement. Raised total bilirubin indicates hemolysis while elevated levels of ALT and AST mostly reflect liver injury.

The definite treatment of HELLP syndrome is delivery of the fetus. If the pregnancy is less than 34 weeks, delivery may be delayed to administer corticosteroids to improve fetal lung maturity. Blood pressure control is very important. Patients with postpartum HELLP syndrome may need plasma exchange.

5.CONCLUSION

This case report highlights the importance of investigating asymptomatic thrombocytopenia in pregnant females for the possibility of HELLP syndrome. Not all patients with HELLP syndrome tend to be symptomatic and HELLP syndrome should be excluded in any pregnant patient in the second/third trimester or the immediate postpartum period with thrombocytopenia. Prompt identification and early intervention can lead to the best outcome for mother and fetus.

References:

1. Stone, J. H. (1998). HELLP syndrome: hemolysis, elevated liver enzymes, and low platelets. *Jama*, 280(6), 559-562.
2. Sibai, B. M., Taslimi, M. M., El-Nazer, A., Amon, E., Mabie, B. C., & Ryan, G. M. (1986). Maternal-perinatal outcome associated with the syndrome of hemolysis, elevated liver enzymes, and low platelets in severe preeclampsia-eclampsia. *American journal of obstetrics and gynecology*, 155(3), 501-507.
3. Haram, K., Svendsen, E., & Abildgaard, U. (2009). The HELLP syndrome: clinical issues and management. A review. *BMC pregnancy and childbirth*, 9(1), 8.
4. Wolf JL. Liver disease in pregnancy. *Medical Clinics of North America*. 1996 Sep 1;80(5):1167-87.
5. Gleeson, R., Farrell, J., Doyle, M., & Walshe, J. J. (1996). HELLP syndrome: a condition of varied presentation. *Irish journal of medical science*, 165(4), 265.
6. Thomas, T., Jophy, R., Mhaskar, A., & Misquith, D. (2005). Are we increasing serious maternal morbidity by postponing termination of pregnancy in severe pre-eclampsia/eclampsia?. *Journal of obstetrics and gynaecology*, 25(4), 347-351.
7. Weinstein, L. (1982). Syndrome of hemolysis, elevated liver enzymes, and low platelet count: a severe consequence of hypertension in pregnancy. *American journal of obstetrics and gynecology*, 142(2), 159-167.
8. Lisonkova, S., Razaz, N., Sabr, Y., Muraca, G. M., Boutin, A., Mayer, C., ... & Kramer, M. S. (2020). Maternal risk factors and adverse birth outcomes associated with HELLP syndrome: a population-based study. *BJOG: An International Journal of Obstetrics & Gynaecology*.



9. Oliveira, N., Poon, L. C., Nicolaides, K. H., & Baschat, A. A. (2016). First trimester prediction of HELLP syndrome. *Prenatal diagnosis*, 36(1), 29-33.
10. Widmer, M., Villar, J., Benigni, A., Conde-Agudelo, A., Karumanchi, S. A., & Lindheimer, M. (2007). Mapping the theories of preeclampsia and the role of angiogenic factors: a systematic review. *Obstetrics & Gynecology*, 109(1), 168-180.
11. Ibdah, J. A., Bennett, M. J., Rinaldo, P., Zhao, Y., Gibson, B., Sims, H. F., & Strauss, A. W. (1999). A fetal fatty-acid oxidation disorder as a cause of liver disease in pregnant women. *New England Journal of Medicine*, 340(22), 1723-1731.
12. Martin Jr, J. N., Rose, C. H., & Briery, C. M. (2006). Understanding and managing HELLP syndrome: the integral role of aggressive glucocorticoids for mother and child. *American journal of obstetrics and gynecology*, 195(4), 914-934.
13. Sibai, B. M. (2004). Diagnosis, controversies, and management of the syndrome of hemolysis, elevated liver enzymes, and low platelet count. *Obstetrics & Gynecology*, 103(5), 981-991.
14. Marchand, A., Galen, R. S., & Van Lente, F. (1980). The predictive value of serum haptoglobin in hemolytic disease. *Jama*, 243(19), 1909-1911.
15. Wilke G, Rath W, Schutz E, Armstrong VW, Kuhn W. Haptoglobin as a sensitive marker of hemolysis in HELLP-



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