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A Rare Case in 17 Weeks of Gestation: Hemolysis, Elevated Liver Enzymes and Low Platelet Count Syndrome.

17. Gebelik Haftasında Nadir Bir Vaka: Hemoliz, Yükselmiş Karaciğer Enzimleri, Düşük Platelet Sayısı Sendromu.

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Abstract

The HELLP syndrome is a condition characterized by Hemolysis, Elevated Liver enzymes and Low Platelet numbers. It occurs in 0.5-0.9% of all pregnancies and 10-20% of patients with severe preeclampsia. It often occurs as part of severe preeclampsia after 27th week of gestation. We hereby present an unusual case of HELLP syndrome, which was diagnosed at 17 weeks of gestation. If definitive diagnosis of HELLP syndrome is made, prompt termination of pregnancy should be considered, particularly in pregnancies, which are remote from term, to prevent potentially fatal complications both for the mother and the fetus..

Keywords: HELLP syndrome, early pregnancy, Pre-eclampsia

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Özet

HELLP (Haemolysis, Elevated Liver enzymes and Low Platelet) sendromu hemoliz, yükselmiş karaciğer enzimleri ve düşük platelet sayısı ile karakterize bir sendromdur. Tüm gebelerin %0.5-0.9'unda ve şiddetli preeklampsili gebelerin %10-20'sinde görülür. Hellp sendromu genellikle 27. gebelik haftasından sonra görülen bir durumdur. Biz burada 17 haftalık gebede görülen bir Hellp sendromu vakasını sunduk. Özellikle termden uzak gebeliklerde Hellp sendromundan kesin emin olunursa ileriki zamanlarda gelişebilecek anne ve bebeğin hayatını riske edebilecek komplikasyonları önlemek için gebelik hızla sonlandırılmalıdır.

Anahtar Kelimeler: HELLP sendromu, erken gebelik, preeklampsi

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Introduction

The HELLP syndrome is a condition characterized by Hemolysis, Elevated Liver enzymes and Low Platelet numbers[1]. It occurs in 0,5-0,9% of all pregnancies[2]. While 70% of cases are diagnosed between 27 and 34 weeks of gestation, HELLP syndrome has been reported very rarely before 20 weeks of gestation[2]. The syndrome is associated with serious maternal and fetal complications, and thus, management and optimization of maternal and fetal well-being is challenging.

Case report

A 24 year-old woman (gravida 3, para 2) was admitted to Cardiology clinic with the diagnosis of 3rd degree mitral insufficiency. On the day of her admission, since the patient was pregnant and reported epigastric pain,

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she was consulted to our clinic. In her history, she reported epigastric pain for one day. Physical examination revealed no abnormal finding. Obstetric ultrasonography detected single, viable intrauterine fetus of 17 weeks. Since admission, maximum arterial blood pressure was 130/80mmHg. The initial laboratory values were reported as the following: spot urine protein: +/- (trace), Hb: 11.7g/dl, platelet count: 242 103/mm3 , AST:22 IU/L, ALT:18 IU/L and LDH: 245 IU/L. Follow-up of arterial blood pressure, daily work-up of laboratory routines and collection of a 24-hour urine sample for protein measurement was ordered. Patient was reevaluated the next day. Her epigastric pain was not relieved despite antiacid medications. The highest blood pressure on patient chart was 140/90mmHg. Laboratory values were as follows: spot urine protein: +2, Hb: 11.9g/dl, platelet count: 145 103/mm, AST:39 IU/L, ALT:45 IU/L and LDH: 375 IU/L. 24-hour urine protein was reported as 0,4g/24h. In the 3rd day of follow-up, the highest blood pressure was measured as 160/100mmHg and laboratory values were as follows: spot urine protein +3, Hb: 12.2g/dl, platelet count: 142 103/mm, AST:72 IU/L, ALT:78 IU/L and LDH: 573 IU/L. The next day, patient reported increase in severity of her epigastric pain, and a new-onset headache. Control ultrasonography revealed that the fetus of 17 weeks and 4 days of gestation was viable. Arterial blood pressure increased to 180/110mmHg. Laboratory values in the 4th day of follow-up was: spot urine protein +3, Hb: 12.4g/dl, platelet count: 62 103/mm, AST: 556 IU/L, ALT: 623 IU/L ve LDH: 1137 IU/L, total bilirubin: 2.9 mg/dL, direct bilirubin: 1.2 mg/dL , creatine:1.1 mg/dL. Recollected 24-hour urine protein was 2.4g/24h. Diagnosis of HELLP syndrome was made. Intravenous magnesium sulphate treatment for the prevention of eclamptic seizures was started immediately, just before oral and vaginal administration of prostaglandin analogue, misoprostol (200µg) for termination of the pregnancy. The patient had complete abortion 7 hours after misoprostol administration. Magnesium sulphate administration was continued for 36 hours following abortion. Patient's symptoms of epigastric pain and headache disappeared and improvements in laboratory values and arterial blood pressure measurements began 1 day after the termination of pregnancy. In the 2nd day after abortion, the highest measured arterial blood pressure was 120/70mmHg and her laboratory values were: Hb: 11.1g/dl, platelet count: 214 103/mm, AST: 58 IU/L, ALT:49 IU/L and LDH:463 IU/L. After satisfactory symptomatic and clinical resolution, the patient was discharged with instructions, four days after the induced abortion.

Discussion

HELLP syndrome is a life-threatening condition both for the mother and the fetus. It occurs in 0.5-0.9% of all pregnancies and 10-20% of patients with severe preeclampsia[2]. Maternal mortality rate associated with HELLP syndrome was reported as 1.1%[3]. It often occurs as part of severe preeclampsia after 27th week of gestation. While occurring less frequently before 27th week of gestation, reports of patients with HELLP syndrome before 20th gestational week are extremely rare in the literature[2]. In this paper, we report an unusual case of HELLP syndrome which was diagnosed at 17 weeks of gestation.

According to the Tennessee classification system, diagnostic criteria for HELLP syndrome include: LDH: > 600 IU/L, AST: ≥ 70 IU/L and platelet count: < 100 103/L (4). In our patient, while normal at admission, those parameters were deteriorated quickly by the 4th day of admission as: LDH: 1137 IU/L, AST: 556 IU/L and platelet count: 62 103/mm. The most commonly reported symptoms of HELLP syndrome are epigastric and right upper quadrant pain[3]. Only complaint of our patient was epigastric pain. At initial laboratory work-up, only positive finding was trace (+/-) amount of spot urinary protein which can usually be considered as normal in pregnant women. Since proteinuria may occur late in pregnancy, patients with negative urinary protein but who are suspected to develop preeclampsia and HELLP syndrome should be followed closely for proteinuria development. In our case, while initial proteinuria in spot urine and 24-hour urine sample was +/- and 0,4g, respectively, during follow-up, it increased up to +3 and 2,4g, respectively. HELLP syndrome is a progressive condition. Diagnosis of preeclampsia and HELLP syndrome can be made more precisely as severity of the proteinuria and hyper-

tension increases. Additional presence of epigastric pain and headache helps the definitive diagnosis (5). During follow-up, our patient's proteinuria increased from 0.4g to 2.4g, her blood pressure increased from 130/80mmHg to 180/110mmHg, her liver enzymes increased (i.e. AST increased from 22 IU/L to 556 IU/L) and she complained of new-onset headache so that a definitive diagnosis has been able to be established.

Delivery remains the ultimate treatment for severe preeclampsia and HELLP syndrome. Should the delivery delayed, especially when remote from term, fatal complications such as consumptive coagulopathy, intracerebral hemorrhage and sub capsular liver hematoma with rupture may occur[6]. Therefore, pregnancies with HELLP syndrome, which are remote from term, must be terminated as soon as possible since it may result in severe maternal morbidity and mortality. In our case, immediately after we reached the accurate diagnosis of HELLP syndrome and informed the patient about her condition and risks, we terminated the pregnancy by inducing abortion with misoprostol (CytotecR, Pfizer) (400mcg vaginally or sublingually 3-hrly, max x5)[7].

Acute fatty liver of pregnancy (AFLP) should be considered in the differential diagnosis of early-onset HELLP syndrome. Acute fatty liver of pregnancy almost always manifests late in pregnancy. Malaise, persistent nausea and vomiting are major symptoms in AFLP while headache, abdominal or epigastric pain and hematuria are the most common symptoms of patients with HELLP syndrome[8]. Abdominal pain and jaundice are also common in AFLP. Major hypoglycemia, hypocholesterolemia, hypotriglyceridemia, serum transaminase activity and low antithrombin III are common in women with AFLP[8]. Disseminated intravascular coagulation, acute renal insufficiency, ascites, seroma and encephalophaty are more common with AFLP[8]. Thorough investigation of clinical presentation, biochemical findings and complications aids in distinguishing AFLP from HELLP syndrome.

The patient was counseled about a significantly increased risk of preeclampsia and HELLP syndrome in subsequent pregnancies[4]

In conclusion, early detection, cautious maternal monitoring, and early intervention are critical in preventing maternal and fetal mortality related to this disorder. Therefore, diagnosis of HELLP syndrome should be considered in pregnant women presenting with symptoms, signs or laboratory abnormalities compatible with this diagnosis, even before 20th gestational week.

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