HELLP Syndrome Detected in Remission of an ITP Patient in Management of HELLP syndrome: A Case Report

Remisyondaki ITP Hastasında Görülen HELLP SendromuYönetimi: Olgu Sunumu

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Özet

HELLP sendromu preeklampsinin hayati tehdit eden bir formudur. Bu çalışmada biz immün trombositopeni ve şiddetli preeklampsi ile komplike HELLP sendromu olan bir olguyu sunuyoruz. Otuz sekiz yaşında HELLP sendromlu 39 gebelik haftasında olgu bulantı ve kusma nedeniyle başvurdu. Hastanın öyküsüne göre 23 yıl önce ITP tanısı almış ve 2 yıldır herhangi bir tedavi almamaktadır. Bu olgu öncelikle, hemoliz, artmış karaciğer enzim düzeyleri ve düşük platelet sayısı gibi bulgularla HELLP sendromu kabul edildi. Aynı zamanda olguda hipertansiyon ve idrarda 3+ protein saptandı. Gerekli medikal müdahalelerden sonra, fetal distress ve olgunun klinik duruma bağlı sezeryan operasyonu yapıldı. Postoperatif 7. gününde destekleyici steroid tedavisi ile beraber laboratuar testleri ve klinik durumu stabil idi. HELLP sendromunun erken tanısı ve tedavisi maternal-perinatal mortalite ve morbiditeyi engelleyebilmektedir.

Anahtar kelimeler: HELLP sendromu; preeklampsi; trombositopeni

Abstract

HELLP syndrome is a life- threatening form of pre-eclampsia. In this study we present a case of HELLP syndrome which was complicated with severe pre-eclampsia and immune thrombocytopenia. A 38-year-old pregnant woman with HELLP syndrome at 39 weeks of gestation was admitted to our clinic with vomiting and nausea. Her medical history revealed was diagnosed with ITP twenty-three years ago and she has had no treatment for 2 years. This case firstly, HELLP syndrome were accepted with these laboratory findings such as haemolysis, elevated liver enzymes and low platelet counts. In additon, she had severe pre-eclampsia with hypertension and 3+ protein in urine. After the necessary medical agents were administered, a cesarean section was performed due to her clinic status and fetal distress. By the 7th day, the clinical course and laboratory values of the patient normalized within supportive steroid treatment. Early diagnosis and treatment of HELLP syndrome may prevent maternal and perinatal mortality and morbidity.

Key words: : HELLP syndrome; pre-eclampsia; thrombocyopenia

INTRODUCTION

HELLP syndrome (H = Haemolysis, EL = Elevated Liver enzymes, LP = Low Platelets) as a life- threatening form of pre-eclampsia includes hemolysis, elevated liver enzymes and low platelets and has a negative effect on both maternal and perinatal outcomes (1). HELLP syndrome usually occurs related to pre-eclampsia. However, about one fifth of HELLP syndrome cases may not be related to pre-eclampsia. Clinical findings such as severe epigastric pain, low platelet counts with high ALT and AST levels may be considered suspicious for HELLP syndrome (2). Thrombocytopenia, which is defined as platelet count below 150x109/L is the second most common cause of hematological abnormality during pregnancy (3). The first and second common causes of thrombocytopenia, respectively are gestational thrombocytopenia for 70%-80% of cases and immune thrombocytopenic purpura in approximately 3% of women defined as thrombocytopenic at delivery (4). This study reports the successful management of a primiparous case presenting with severe pre-eclampsia, HELLP syndrome with term pregnancy and immune thrombocytopenic purpura diagnosed at the age of 15, who underwent cesarean section under general anesthesia.

CASE REPORT

A 38-year-old pregnant woman, gravida 1, was admitted to our clinic in the 392/7 week gestation, with vomiting and nausea, severe pre-eclampsia and HELLP syndrome. In the medical history, immune thrombocytopenia (ITP) had been diagnosed at the age of 15. At the time of presentation, she had not been taking any medication for ITP for 2 years. On examination, she was afebrile with blood pressure of 180/100 mm Hg, pulse of 92 beats per minute and pretibial edema (3+) was detected. Nifedipine 60 mg/day was administered as a slow-release calcium channel inhibitor, after which blood pressure reduced to 140/80 mmHg. The laboratory tests revealed 3+ proteinuria, elevated liver enzymes (AST 1000 and ALT 1241 U/L, normal 7-45 U/L), elevated LDH (1651 U/L, normal 140-280 U/L), low platelets (18 K//uL, normal 142-424 K//uL) and decreased fibrinogen level (164 mg/dL, normal 175-400 mg/dL). The coagulation profile and other biochemical parameters such as electrolyte values, ANA screen and viral hepatitis panel were normal (Table 1). Obstetric ultrasonography showed normal fetal growth and anatomy with estimated fetal weight of 3186 g with normal amniotic fluid. Prednisone was administered at 1mg/kg per day for ITP, and magnesium sulphate for seizure prophylaxis (6 g in 20 minutes). Although low platelet count was determined from the laboratory test results, the peripheral blood smear showed a platelet count of 50 K//uL. Initial treatment was preoperative blood transfusion (2 units of packed red blood cells, 1 unit of packed fresh plasma and 1 unit of packed plateletpheresis). The non-stress test (NST) showed decreased variability and non- reactivity, so an emergency cesarean section

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Kabul Tar: 22.06.2015

Table 1. Preoperative and postoperative follow-up laboratory results.

Laboratory tests	Preoperative tests	Postoperative follow-up tests			
		6 h	9 h	18 h	27 h
Hb (gr/dl)	13.3	11	8.4	7,5	6.5
Plt (x103/mm3)	18	15	31	25	20
Wbc (x103/mm3)	10.5	7.8	10	13,4	15.9
ALT (IU/L)	1241	1128	832	477	377
AST (IU/L)	1000	781	583	515	254
INR	1.12	1.45	1.17		
BUN (mg/dL)	13	15	16	16	15
Cr mg/dL	0.6	0.7	0.7	0.6	0.5
LDH (U/L)	1651	1542	1350	1061	723
Fibrinogen (mg/dL)	164	-	-	-	-

ALT: Alanine aminotransferase; AST: Aspartate aminotransferase; BUN: Urea nitrogen; Cr: Creatinine; d: day h: hour; Hb: Hemoglobin; INR: International normalized ratio of prothrombin time; LDH: Lactate dehydrogenase; Plt: Platelet count; Wbc: White cell count.

was performed under general anesthesia. A 3210-g female was delivered with Apgar 8/9 at 1 and 5 minutes. Two units of packed haemocomplettan 1 g and transexamic acid 1000 mg x 2 were administered peroperatively. Magnesium sulphate infusion (2g/hour/day) was also administered. The patient was admitted to the intensive care unit for further observation and management. Due to findings of anemia (hemoglobin 6.5 g/dL, normal 12-18 g/dL) and thrombocytopenia (platelet count 18.5 K//uL) on postoperative Day 1, 3 units of packed red blood cells and 1 unit of packed plateletpheresis were transfused. The patient recovered without any maternal complications and all laboratory tests normalized at the 168th hour (7th day). She was discharged on the 7th day without any complications.

DISCUSSION

HELLP syndrome is seen in about 0.5% to 0.9% of all pregnancies and in 10%-20% of pregnancies with severe pre-eclampsia(5, 6). HELLP syndrome occurs before delivery in most cases with a maximum frequency between the 27th and 37th week of gestation; 10% develop before the 27th week-, and 20% beyond the 37th weeks of gestation (7, 8). The majority of women with HELLP are multiparous (9). Although there are differences in development, the onset of HELLP syndrome is generally rapid (10). Most women diagnosed with HELLP syndrome also have hypertension and proteinuria, which may not be detected in 10%–20% of cases (11). Specific clinical symptoms include right upper abdominal quadrant or epigastric pain, nausea and vomiting, with upper abdominal pain fluctuating, as in colic (11, 12). The first step for the patient diagnosed with HELLP syndrome is to examine and evaluate. It is necessary to determine the clinical maternal status and should revise obstetric evaluation, the method of birth, ultrasonograpic evaluation, labaratory tests such as complete blood cell count, PLT count, coagulation parameters,AST, LDH and also fetal assessment tests (cardiotocography and doppler examination) (13). Secondly, the aims of treatment are to maintain the maternal clinical condition via supporting intravenous fluids, antihypertensive agents (e.g. labetalol or nifedipine) for hypertension and magnesium sulphate to prevent convulsions (5, 10, 11, 13). HELLP syndrome can be diagnosed during pregnancy or in the postpartum period and is associated with increased maternal risks such as liver hematoma, failure or

rupture, pulmonary edema, renal failure, hemorrhagic complications and death. Perinatal prognosis is poor because of preterm birth and growth restriction (8, 14, 15). As an important form of severe pre-eclampsia, the definitive treatment for HELLP syndrome is delivery of the fetus and removal of the placental tissue (8). There is no specific treatment at present because the full physiopathology remains unclear (16, 17, 18). To date, the methods of treatment are limited to blood pressure regulation, prophylaxis to prevent convulsions, and termination of the pregnancy (19). It is essential to have close monitoring of the maternal vital signs and fluid balance in these patients (13). Thrombocytopenia (platelets (PLTs) < 150x109/L) in pregnancy may be associated with gestational thrombocytopenia (59%), ITP (11%), pre-eclampsia (10%), and HELLP syndrome (12%) (20). The method of delivery for a pregnant woman diagnosed with ITP depends on the obstetric indications. Therefore, it should be considered the risk of cesarean-section for an every labour and general recommendations should be suggested below 50x109/L (21). To succeed in that aim, different combinations of IVIG, platelet transfusions, and corticosteroids can be administered. The administration of platelet transfusions before delivery in pregnant patients diagnosed with ITP has been reported in 5%-18.9% of cases from different patient populations and different practices (22, 23). In cases where the maternal status worsens, immediate caesarean section must be performed (24, 25).The largest clinical study in 2005 reported no difference between the two groups in respect of duration of hospitalization, improvement time for laboratory or clinical values, complications or requirement for blood transfusion. The findings were similar, even with following analysis whether the patients were still pregnant or in the postpartum period (26). A systematic review in the Cochrane Library consisted of 11 randomized controlled studies which compared corticosteroids (dexamethasone, betamethasone, or prednisolone) administered during pregnancy, just after delivery or in the postnatal period, or both before and after birth, with placebo or no treatment. There was no clear evidence of any effect of corticosteroids on significant clinical outcomes. It was concluded that the evidence was insufficient to show better clinical outcomes after the administration of steroids in the management of HELLP syndrome. However, the usage of corticosteroids was recommended because of clinicly confirmation of rapidly increased platelet count in the recovery period (27).

In the current case, the medical history revealed immune thrombocytopenic purpura diagnosed at the age of 15 and all symptoms had a rapid onset two days prior to presentation at hospital. The patient was admitted to our clinic with the complaints of vomiting, nausea. Increased enzymes levels (ALT, AST, LDH), low platelet count, high hypertension and 3+ protein in the urine were found. All of these findings considered HELLP syndrome. Ultrasound examination revealed a normal fetal growth, but the fetal non-stress test had decreased variability. The patient were transfused red blood cells, fresh plasma, platelet pheresis and administered antihipertension drugs and prednison. Although the patient had a term pregnancy, decreased variability, primigravida and HELLP syndrome clinic, a cesarean-section was performed immediately. Postoperatively, the clinical course of patient improved gradually, normalized on 168th hours and was discharged on the 7th day without any complications.

In conclusion, when pregnant women present with vomiting, nausea and hypertension and history of ITP, it should not be forgotten that is one of the first findings of HELLP syndrome and accompanying other hematological disorders. As women with ITP had good pregnancy outcomes, women with HELLP syndrome had poor pregnancy outcomes. Early diagnosis and treatment will be able to prevent maternal and perinatal mortality and morbidity. If the clinical status of the patient worsens, the appropriate mode of delivery will be immediate caesarean section.

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