ZKTB CASE REPORT *(Olgu Sunumu)*

Early Onset Hellp Syndrome with Systemic Lupus Erytematosus and Acute Cholecystitis Attack at 17th Week of Gestation

17. Gebelik Haftasında Akut Kolesistit Atağı ve Sistemik Lupus Eritematozus ile Birlikte Görülen Erken Başlangıçlı Hellp Sendromu

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ABSTRACT

Hellp syndrome is an atypic, severe and life threatening form of preeclapmsia, which is usually associated with hemolysis, elevated liver enzymes and low platelet count. Also it is very important because it causes significantly maternal and perinatal mortality. It is known that hellp syndrome almost always occurs after 20 weeks of gestation. But in the presence of some accompanying diseases, hellp syndrome or severe preeclampsia can occur at earlier gestational weeks. This may also lead to diagnostic difficulties. In this case, we described the diagnosis and recovery period of a systemic lupus erytematosus (SLE) patient who developed hellp syndrome at 17 weeks of gestation. Our experience takes attention that detection of hellp syndome accompanying diseases like systemic lupus erytematosus or acute cholecystitis can be difficult and it can occur distinctly at earlier weeks of gestation. Also the differential diagnosis is very important in such cases, because many diseases can mimic hellp syndrome.

Keywords: hellp syndrome, systemic lupus erytematosus, early onset, liver enzymes

ÖZET

Hellp sendromu, preeklampsinin atipik, şiddetli ve hayatı tehdit eden bir formu olup genellikle hemoliz, yüksek karaciğer enzimleri ve düşük platelet düzeyleri ile ilişkilidir. Ayrıca, belirgin maternal ve perinatal mortaliteye de neden olduğundan önemi büyüktür. Hellp sendromunun hemen her zaman gebeliğin 20. haftadan önce meydana geldiği bilinmektedir. Fakat bazı eşlik eden hastalıkların varlığında hellp sendromu veya şiddetli preeklampsi daha erken gebelik haftalarında da görülebilir. Bu durum aynı zamanda tanısal zorluğa da neden olur. Biz bu olguda, 17. Gebelik haftasında hellp sendromu gelişen bir sistemik lupus eritematozus (SLE) hastasının tanı ve tedavi periodunu anlattık. Çalışmamız; hellp sendromunun SLE ya da akut kolesistit gibi eşlik eden hastalıklar varlığında tespit edilmesinin zorluğuna ve şaşırtıcı olarak erken gebelik haftalarında meydana gelebilmesine dikkat çekmektedir. Aynı zamanda, böyle olgularda, birçok hastalık hellp sendromunu taklit edebileceğinden ayırıcı tanı da çok önemlidir.

Anahtar Kelimeler: hellp sendromu, sistemik lupus eritematozus, erken başlangıç, karaciğer enzimleri

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INTRODUCTION

Hemolysis, elevated liver enzymes and low platelet count forms the known triad of hellp syndrome. The laboratory findings of the syndrome are seconder to this triad; hyperbilirubinemia, increasement of serum AST, ALT and LDH levels, platelet levels under 100.000 microL. (1). Hellp is known as a form of preeclampsia, but sometimes there are no symptoms of classic preeclampsia like hypertansion, edema or proteinuria. Also epigastric pain, abdominal tenderness, headache, vomiting, end organ failures (like renal failure, pulmonary edema, acute liver infarction, dissemine intravascular coagulation etc.) can be present with varying frequency (2). It is known that hellp syndrome almost invariably occurs after 20 weeks of gestation (3-5). On the other hand, atypic forms of hellp syndrome can develop at different patients or some diseases can come to exist with symptoms seen like at hellp syndrome. This leads to diagnostic difficulties. The clinician must evaluate all symptoms and laboratory findings attentively and make a good differential diagnosis for management of hellp syndrome. In our case, we discussed the diagnosis and recovery period of a SLE patient superimposed with hellp syndrome and acute cholecystitis at 17 weeks of gestation. In this case, renal failure and hypertension due to SLE could be confused with hypertension and renal involvement seen in the hellp syndrome. Also increasement of serum liver enzyme levels could both cause from acute cholecystitis and hellp syndrome. The gestation week was an other confusing stuation to diagnose hellp syndrome in this case.

THE CASE

A 24 years old, gravida 1, parity 0 women presented with right upper abdominal pain and vomiting to the emergency department of XXXX university hospital. Her pregnancy dating was 17 weeks and 3 days. She had diagnosis of SLE before her pregnancy and her treatment period was proceeding while she became pregnant. Also renal failure due to SLE was occured before pregnancy and she had chronic hypertantion. She had only one time ultrasound examination previously at 9th weeks of gestation and antihypertensive therapy was rearranged according to the pregnancy at that visit. Her first laboratory findings were elevated liver enzymes (asparate aminotransferase (AST) 192 U/L and alanine aminotransferase (ALT) 149 U/L).

Serum lactate dehydrogenase was 594 U/L, creatine 1,5 mg/dL, total and direct bilirubin were normal range. Hemoglobin(hb) value was 8,7 g/dL, platelet(plt) 113000/µL, proteinuria +2. Her blood pressure was 140/80mmHg. The abdominal ultrasound performed by radiologist showed acute cholecystitis symptomes, pericholecystic and peripancreatic edema, minimal fluid arround pancreas and gall bladder. Also the abdominal MRG imaging interpreted in the manner of acute edematous pancreatitis. As hellp syndrome scarcely ever occures before 20 weeks of gestation, we got away from the diagnosis of hellp syndrome. According to these symptomes and laboratory, the patient was hospitalised to the department of general surgery intensive care unit with a diagnosis of acute edematous pancreatitis and acute cholecystitis eventually. Proper antibiotherapy and follow up period started for thereatment.

18 hours after the hospitalisation, her blood pressures started to increase progressively. New blood analysis showed a rapid increase at liver enzymes, AST was 757 U/L, ALT 424 U/L, LDH 1971 U/L. Serum creatine level was 2,2mg/dL, hemoglobin value was 7,1 g/dL, platelet 16000/µL, hct %20,4. Blood pressure increased to 190/100 mmHg. She was counseleed to gyneacology and obstetrics department of hospital immediately and the diagnose changed to hellp syndrome and we decided to terminate pregnancy immediately. Because of very low platelet levels of the patient, before the termination procedure we prepared platelet suspension by apheresis. We decided to terminate pregnancy by surgycal procedure, because there is not enough time for other methods. After 2 units of apheresis transfusion, platelet level was increased to 123000/µL and patient went to hysterotomy anterior operation. During the operation, 2 units of erytrocite suspension transfusion was performed to the patient.

After the operation, liver enzymes and creatine values decreased progressively (postopertive 12. hours AST 300 U/L, ALT 357U/L, LDH 1113U/L, creatine 1.8mg/dL; postoperative 40. hours AST 108U/L, ALT 232U/L, LDH 663U/L, creatine 1,6mg/dL; postoperative 72. Hours AST 69U/L, ALT 183U/L, LDH 534U/L, creatine 1,2mg/dL). Hemoglobine and platelet values decreased, totally 4 units of erytrocite suspension and 3 units of platelet suspension by apheresis were given to the patient until postoperative 72. Hours, Hb: 9,4 g/dL, plt 106000 /µL. Her blood pressure was in rise at first 24 hours. Magnesium sulphate and glyceryl trinitrate infusion continued for first 24 hours and then we started oral antihypertensive treatment . Postoperative 7th day, we discharged the patient from the hospital with normally ranged liver enzymes and 116000 /µL platellet value with oral antihypertensive drugs.

DISCUSSION

Hellp syndrome commonly occures in third trimestr of pregnancy and before 27 weeks of gestation, it is very rare (6). It is known that hellp syndrome almost always occurs after 20 weeks of gestation (3-5). We scanned all cases for the last 20 years. There was no case of early onset hellp syndrom accompanying with both SLE and acute cholecystitis attack.

Wada et al. reported a case of hellp syndrome in 17th week of gestation who had SLE and antiphospholipid syndrome (APS) (7). In that case, intrauterine fetal death was occured and the characteristic features of HELLP syndrome was determined, but because of early onset, diagnostic difficulty was present. There is a close relationship between antiphospholipid syndrome (APS), SLE and Hellp syndrome. Patients with APS or SLE may be complicated with hellp syndrome at earlier weeks of gestation (8). Our patient was at 17th week of gestation, this is a rarely seen condition for hellp syndrome, so it could cause a diagnostic delay.

Another early onset hellp syndrome was reported by Suzumori et al. (9). That patient was at 22th week of gestation and had SLE diagnosis with lupus nephritis. She complicated with hellp syndrome and because of her renal disorder, early onset hypertansion and proteinuria occured like our patient. In our case, the woman had already chronic hypertansion and proteinuria because of renal disorder, but she had also acute cholecystitis and suspected edematous pancreatitis that could cause the elevation of liver enzymes. Therefore, when the patient first applied to hospital with minimal elevation of liver enzymes, minimal hypertansive blood pressures and proteinuria, she got diagnosis of acute cholecystitis and suspected edematous pancreatitis in terms of developing hellp syndrome. After the formation of absolute clinical findings with severe liver enzyme levels, very low pletelet count and extremely high blood pressures, final diagnosis was established.

Also, Haram et al. reported an 18th week of pregnancy associated with the antiphospholipid–antibody syndrome complicated with hellp syndrome (10). They showed that hellp could occure at early second trimestr accompanied with other medical pathologies like SLE or APS. The patient hospitalised with epigastric pain like our patient. She had elevated liver enzymes but not low platelet count at first. After hospitalisation, her liver enzymes elevated progressively and platelets were decreased. Her blood pressures distinctly were not raised high. At last, her pregnancy terminated and her clinical condition improved progressively like our patient.

As seen in these patients, early onset hellp syndrome has a relationship with SLE. Especially we must consider that hellp sendrome can occure before 20 weeks of gestation as it is very rare. Also some diseases like SLE, molar pregnancies and APS can be superposed with hellp syndrome at early second trimestr, before 20th week of gestation (10). Some diseases can imitate hellp syndrome's findings or laboratory results, so differential diagnosis must be made carryfully. In all cases with elevated liver enzymes, hypertansion, epigastric pain or low platelet; hellp syndrome must be in our minde regardless of the gestational week.

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