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Case Report

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HELLP syndrome complicated by hepatic rupture

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Abstract

Objective: Although spontaneous hepatic rupture associated with HELLP (hemolysis, elevated liver enzymes, and low platelets) syndrome is rare, it is a life-threatening complication of pregnancy. The cornerstone of prognosis for successful outcome is its prompt diagnosis.

Case: We report a case of 34-year-old female at 29 weeks of gestation with spontaneous hepatic rupture caused by HELLP syndrome as a presenting symptom of weakness without right upper quadrant abdominal pain.

Conclusion: If there is a pregnant woman who is complaining of weakness accompanying with hypotension, obstetrician should check the patient for the presence of hepatic rupture. Hepatic rupture in HELLP syndrome should be considered as a differential diagnosis in pregnant patients with the complaint of weakness. It usually causes sudden onset of abdominal pain and accompanying hypotension. But as in this case, it may present without abdominal pain. An interdisciplinary surgical approach including supportive measures with the use of temporary packing of the liver to control the bleeding can result in successful outcome.

Keywords: Atypical presentation, HELLP syndrome, hepatic rupture.

Özet: Hepatik rüptür ile komplike olan HELLP sendromu

Amaç: HELLP (hemoliz, yükselmiş karaciğer enzimleri ve düşük trombosit) sendromu ile ilişkili spontan hepatik rüptür nadir görülse de, gebeliğin hayatı tehdit eden bir komplikasyondur. Başarılı bir sonuç için prognozun köşe taşı, hızlı tanı konulmasıdır.

Olgu: Gebeliğinin 29. haftasındaki 34 yaşında kadın hastada, sağ üst kadran ağrısı olmaksızın halsizlik semptomu olup HELLP sendromunun neden olduğu spontan hepatik rüptürü saptanan bir olgu sunuyoruz.

Sonuç: Hipotansiyona bağlı halsizlik şikayetinde olan bir gebe varsa, doğum uzmanı hastada hepatik rüptür olup olmadığını kontrol etmelidir. HELLP sendromundaki hepatik rüptür, halsizlikten şikayet eden gebelerde ayırıcı tanı olarak düşünülmelidir. Bu, genellikle ani başlangıçlı karın ağrısı ve buna eşlik eden hipotansiyona neden olur. Ancak bu olguda olduğu gibi, karın ağrısı olmadan da ortaya çıkabilir. Kanamayı kontrol altına almak için karaciğerin geçici tampon uygulaması ile destekleyici önlemleri de içeren disiplinlerarası bir cerrahi yaklaşım başarılı sonuçlar sağlayabilir.

Anahtar sözcükler: HELLP sendromu, sıradışı bulgu, hepatik rüptür.

Introduction

Subcapsular liver hematoma has been reported in less than 2% of pregnancies complicated by HELLP syndrome (hemolysis, elevated liver enzymes, and low platelets).[1] Clinicians should be aware of this rare but life-threatening complication with a presenting symptom of right upper quadrant abdominal pain.[2,3] In this paper, we report a patient with spontaneous hepatic rupture without a history of sudden-onset upper abdominal pain.

Case Report

A 34-year-old woman, gravida 2, para 1, was admitted to our emergency room at 29 weeks of gestation. She had history of gestational diabetes in current pregnancy and

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mild preeclampsia in her previous pregnancy, and vaginal delivery at term. She denied any history of trauma. When she admitted to the hospital, she was acutely ill with a pale face developed within an hour. Her arterial blood pressure was 50/35 mmHg with a pulse rate of 144 beats/min, and her body temperature was 36.4 °C. The clinical examination on admission revealed no abdominal pain, no tenderness or rigidity. Her laboratory findings revealed that white blood cell count (WBC) was 16,010/mm^3, hemoglobin was 12.3 g/dL, thrombocytes was 46,000/µL, aspartate aminotransferase (AST) was 419 U/L, alanine aminotransferase (ALT) was 321 U/L, and lactate dehydrogenase (LDH) was 688 U/L. Other laboratory values were unremarkable. On ultrasonography, the liver showed heterogeneous echogenicity, and a hyperechogenic material was noted in the perihepatic space, suggesting the possibility of perihepatic hematoma and fetal heart beats could not be found. We established the clinical diagnosis for suspected hemorrhage or rupture of liver with a circulatory collapse. Therefore, emergency cesarean section was performed.

On opening the peritoneum, we encountered a massive intraperitoneal hemorrhage from a large ruptured subcapsular hepatic hematoma (**Fig. 1**). The color of the amniotic fluid was clear. After performing the quick delivery of the fetus with no life sign, uterine incision was promptly closed and, we examined solid abdominal organs as a potential source of bleeding. At the same time, general surgery was called for an intraoperative consultation. Due to the actively bleeding large liver hematoma, direct pressurization was applied for five minutes with cotton gauze tampons to the perihepatic region after the aspiration of free blood from peritoneal cavity. The operation was terminated when the evident bleeding stopped. The patient received intravenous fluid resuscitation with two units of packed red blood cells, three units of fresh frozen plasma, and was admitted to the intensive care unit for additional support. Postoperatively, the blood pressure increased to 170/115 mmHg. Postoperative intravenous magnesium sulfate was given to the patient for eclampsia prophylaxis. On the first postoperative day, WBC was 9310/mm^3, hemoglobin was 7.8 g/dL, thrombocytes was 87,000/µL, serum creatinine was 0.47 mg/dL, serum AST level was 411 U/L, serum ALT level was 373 U/L, and LDH was 493 U/L. Urine output was adequate. By the 7th postoperative day, ALT and AST levels declined to 49 and 25 U/L, respectively. It was confirmed by clinical signs and serial ultrasound examinations that the patient had no other complications and no evidence of further intra-abdominal bleeding. She was transferred from intensive care unit to normal service floor on postoperative 5th day and discharged from the hospital on the 23rd postoperative day with scheduled outpatient follow-up.

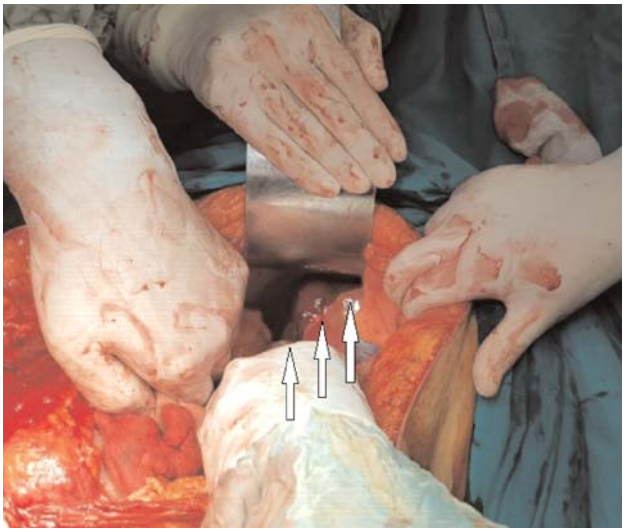

Fig. 1. Appearance of hepatic rupture before packing applied. The white arrows show the hepatic rupture.

Discussion

The clinician should keep in mind the spontaneous hepatic rupture as a rare complication of HELLP, which results in fetal and maternal mortality. The incidence of this complication is estimated to be about 1 case in 45,000–220,000 births.[4,5] Hepatic rupture associated with pregnancy is accompanied by preeclampsia in 80% of the cases.[6] Rupture of hepatic hematoma should be suspected in HELLP syndrome in the presence of sudden hypotension, tachycardia, abdominal pain and pain on the right shoulder.[7] Haram et al. reviewed the complications of HELLP syndrome, and reported that symptoms of spontaneous rupture of subcapsular liver hematoma are sudden-onset severe pain in the epigastric and right upper abdominal quadrant radiating to the back, right shoulder pain, anemia and hypotension.[2] Mascarenhas et al. reported a series of five pregnant women with spontaneous hepatic rup-

ture, and found that pregnant women with HELLP syndrome are more prone to hepatic rupture, but it can also occur with other liver pathology such as hepatic adenomas, primary and secondary malignancies and hemangiomas. In their case reports, all patients had history of sudden-onset upper abdominal pain.[3] It should not be forgotten that although hepatic rupture with pregnancy is accompanying with severe abdominal pain, it is possible to present without pain complaint as in our case.

Although there is still not an agreement on the best approach to treat hepatic rupture, in any case, when it is suspected of hepatic rupture, exploratory laparotomy should be performed.[8] Hepatic packing has the lowest mortality (25–30%) and that is why it is used as a first option surgical treatment. Other treatment options, such as hepatic lobectomy, hepatic artery ligation, and hepatic embolization, have high mortality of 75%, 40%, and 35%, respectively, can be used if packing does not work.[8]

Conclusion

Consequently, early surgical intervention plays a crucial role in HELLP with hepatic rupture. Furthermore, we indicate that hepatic rupture in pregnancy requires a multidisciplinary teamwork including supportive measures with the use of temporary packing of the liver to control the bleeding to obtain a successful outcome. Additionally, it should be realized that, pregnant women with complaints of weakness accompanying with hypotension may be considered to have HELLP syndrome with hepatic rupture even if there is no abdominal pain.

Conflicts of Interest: No conflicts declared.

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