CASE REPORT

Atypical HELLP syndrome secondary to uteroplacental insufficiency

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Very little is known about atypical HELLP syndrome in relation to uteroplacental insufficiency. A 41-year-old primigravida with fetal growth restriction due to uteroplacental insufficiency suddenly experienced severe right upper quadrant pain at 28 weeks gestation without preeclampsia. Although laboratory abnormalities were not evident, the symptoms persisted for 2 hours. Elective cesarean section was performed because atypical HELLP syndrome was highly suspected. On postpartum day 1, a marked elevation of liver enzymes, low platelet count, and anemia without hypertension and proteinuria were noted, but these parameters spontaneously improved within a week. Our experience highlights instances where atypical HELLP syndrome might occur in uteroplacental insufficiency cases.

Introduction

HELLP syndrome, which generally occurs in the presence of preeclampsia, is a serious complication of pregnancy. Stella et al. described cases where presenting symptoms similar to those of HELLP syndrome developed before 20 weeks gestation or without preeclampsia (i.e., atypical HELLP syndrome). However, little is known about the relationship between atypical HELLP syndrome and uteroplacental insufficiency, which is one of the causes of preeclampsia. We describe herein the clinical features of atypical HELLP syndrome in a pregnancy complicated by uteroplacental insufficiency.

Case report

A 41-year-old gravida 0, para 0 woman was referred to our hospital at 28 weeks gestation because of fetal growth restriction (FGR). No maternal risk factors affecting the pregnancy were found, except for advanced maternal age. A fetal ultrasound examination revealed severe FGR (estimated fetal body weight 606 g, − 3.5 S.D.). Doppler studies demonstrated bilateral early-diastolic notches in the uterine artery, end-diastolic reversed flow in the umbilical artery, elevation of the pulsatility index in the ductus venous, and reduction of the pulsatility index in the middle cerebral artery. These findings indicated that the FGR was attributed to uteroplacental insufficiency. Patient blood pressure (119/74 mmHg) and all laboratory findings were normal. Two days later, the patient suddenly experienced severe right upper quadrant pain that radiated into the right shoulder and was associated with vomiting. Blood pressure was normal (126/84 mmHg), and laboratory abnormalities were not noted, except for mildly elevated aspartate aminotransferase levels (Figure 1). An abdominal ultrasound examination did not show any abnormal findings related to the severe right upper quadrant pain such as hematoma, hepatic infarction, fatty liver changes, or gallstone attack. The symptoms persisted for two hours, and finally, elective cesarean section was performed because atypical HELLP syndrome was highly suspected. A female infant weighing 603 g was delivered with Apgar scores of 6 and 7 at one and five minutes, respectively. The patient’s right upper
quadrant pain was relieved soon after delivery. Six hours after delivery (on postpartum day 1), a marked elevation of liver enzymes, low platelet count, and anemia without hypertension and proteinuria were noted (Figure 1). A whole-body computed tomography scan performed on postpartum day 1 did not show any abnormalities. The laboratory parameters returned to normal without any treatment within one week (Figure 1). Based on these findings, the patient was diagnosed with atypical HELLP syndrome. The placenta weighed 120 g and ischemic changes were noted on microscopic examination. The patient was discharged without complications on postpartum day 8.

**Discussion**

In the present case, sudden onset of severe pain radiating from the right upper quadrant into the right shoulder was the only symptom of atypical HELLP syndrome. The clinical symptoms and abnormalities in laboratory findings spontaneously improved after delivery. Right upper quadrant pain results from stretching of Glisson’s capsule, and is caused by hepatocyte injury, which could be caused by bioactive substances derived from the placenta. Several reports suggest that Fas ligand is a strong candidate for hepatocyte injury. Higher production of Fas ligand in a hypoxic villous trophoblast might occur in the maternal circulation in HELLP syndrome than in preeclampsia. Although bioactive substances were not detected in the present case, placental ischemic changes secondary to uteroplacental insufficiency might increase the levels of bioactive substances.

Our experience suggests that atypical HELLP syndrome can occur in cases of uteroplacental insufficiency. In the management of HELLP syndrome, it is important to be aware of clinical symptoms including right upper quadrant pain as well as hypertension, proteinuria, and abnormal blood examination results.

**Conflict of interest**

The authors have no conflicts of interest to declare.

**References**