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Spontaneous hepatic rupture in pregnancy with initial intrapartum cholestasis of pregnancy – a case report

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ABSTRACT

Spontaneous hepatic rupture in pregnancy or the puerperium is an extremely rare and life-threatening condition. We present a case in which the patient initially presented with intrahepatic cholestasis of pregnancy and shortly after delivery developed acute spontaneous hepatic rupture. Unlike most other cases, the patient never developed a fulminant syndrome of hemolysis, elevated liver enzymes and low platelet count (HELLP). Early recognition of the syndrome, an aggressive surgical approach with cesarean section and liver packing and blood component therapy contributed to a successful outcome and admission of 27 days.

Keywords: Liver rupture, Liver hematoma, Pregnancy, Puerperium, Preeclampsia, Cesarean delivery, Intrahepatic Cholestasis of Pregnancy, Spontaneous rupture.

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INTRODUCTION

Spontaneous hepatic rupture in pregnancy or the puerperium is an extremely rare and life-threatening condition (1). Incidence estimates vary from 1:45 000 to 1:225 000 pregnancies (1) with fetal mortality at 37.2% and maternal mortality at 22.1% (2). Early recognition and diagnosis are vital in the treatment and management of the condition.

We present a case in which the patient initially presented with intrahepatic cholestasis of pregnancy and later developed acute spontaneous hepatic rupture.

CASE

A 32-year-old woman, gravida 3, para 0, was examined due to itching in gestational week 31. Laboratory tests showed elevated bile acid levels (40 µmol/L) in agreement with intrahepatic cholestasis of pregnancy (3) and treatment with ursodeoxycholic acid was initiated. Fetal ultrasound examination showed intrauterine growth restriction (-38%) and umbilical artery waveform flow class 1. Therefore, the patient was admitted to the hospital and betamethasone was administered. Aside from itching, the patient had no clinical symptoms, and blood pressure and pulse were normal.

After five days, the patient developed hypertension (135/96 mmHg) and antihypertensive medication (methyldopa 250 mg x 3 daily) was administered. No further clinical symptoms occurred. Blood tests were stable except for a progressive rise in alanine transaminase (120 U/L), which could be explained by intrahepatic cholestasis of pregnancy.

Seven days after admission, the patient presented with sudden shoulder pain. Blood tests showed low haptoglobin (0.21g/L), alanine transaminase had risen steeply (245 U/L), lactate dehydrogenase was high (235 U/L), and thrombocytes were low (141 x 10^9/L). Bilirubin was within normal values (7 µmol/L). These values approached the definition of syndrome of hemolysis, elevated liver enzymes and low platelet count (HELLP), but never exceeded the thresholds (4).

Later the same day, the patient experienced a worsening of shoulder pain as well as upper abdominal pain. Bedside abdominal ultrasound (performed by an obstetrician) showed no hepatic hematoma or intraabdominal fluid. An uncomplicated cesarean section was performed shortly afterwards on indication preeclampsia and clinical suspicion of liver hematoma. Cesarean section revealed no intraperitoneal blood, and a boy (1465 g, Apgar 8 at 5 minutes) was delivered.

In the recovery room, the condition was initially stable. The pain from the upper right quadrant had disappeared. Seven hours after the cesarean section, the patient became hemodynamically unstable with low blood pressure (88/45 mmHg). Bedside abdominal ultrasound showed intraabdominal free fluid around the liver.

Abdominal CT scan revealed a large subcapsular hematoma of 9.2 x 13.8 x 21.3 cm in the right liver lobe (Figure 1). A multidisciplinary team consisting of obstetricians, surgeons, radiologists, and anesthesiologists decided on conservative treatment with blood transfusion and observation in the intensive care unit.

Twelve hours after the cesarean section, the patient became hemodynamically unstable again. Balanced blood transfusions including erythrocyte suspension, plasma, and thrombocytes were initiated. An explorative laparotomy was performed, revealing 2.5 liters of intraabdominal blood originating from a lesion in segment 6-7 of the liver. No bleeding from the uterine incision was identified. The bleeding was managed surgically with liver packing and the abdomen was prepared for Pringles maneuver (surgical occlusion of inflow of blood to the liver from the hepatic artery and portal vein)(5).

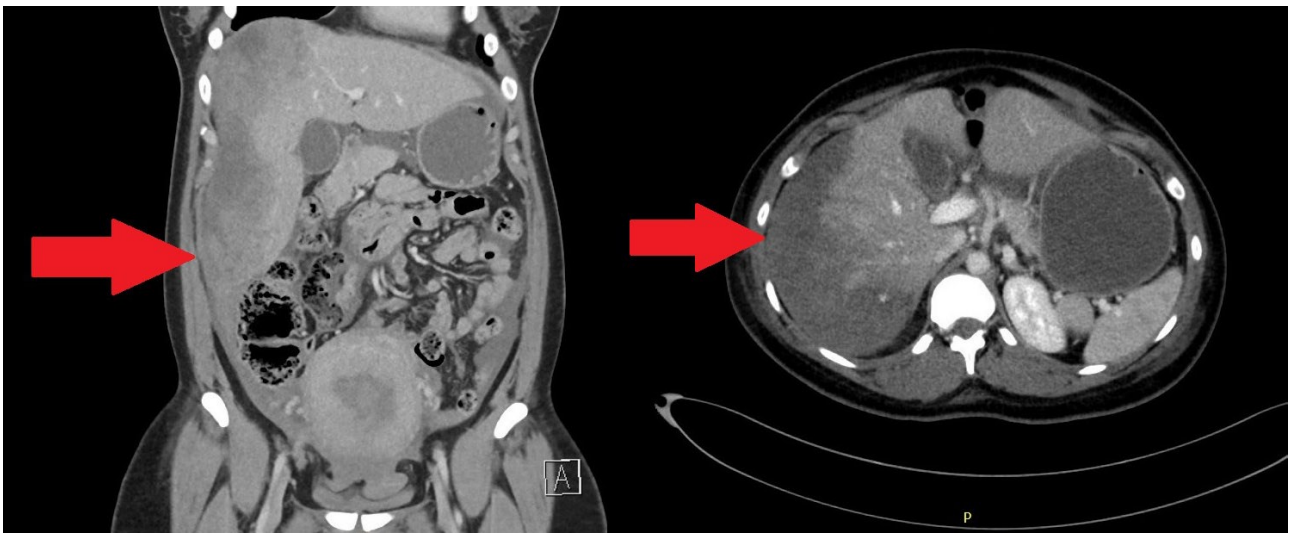

Figure 1: CT radiography of the patient's abdomen, arrow pointing to hepatic hematoma.

The patient was transferred intubated to The Department of Liver Surgery at Rigshospitalet University Hospital of Copenhagen. Coiling of three small arteries in the liver was performed, and the liver bleeding was managed with repacking, and use of TachoSil and Surgicel Fibrillar. Twelve hours later the patient was extubated in the Intensive Care Unit. Forty-eight hours later, the bleeding stopped, and the incision was closed leaving only a drainage tube. Liver enzymes had normalized two days postoperative. The patient developed right sided pleural effusion, which is a known complication to upper abdominal surgery (6). The pleural space was drained for 2.1 liters of fluid and the patient had an uncomplicated recovery afterwards. During the admission the patient received transfusion with six units of thrombocytes, twelve units of erythrocytes, eighteen units of plasma and one unit of pooled cryoprecipitate. The patient was discharged 27 days after delivery.

DISCUSSION

This case describes in detail the clinical picture of a patient with severe spontaneous hepatic rupture in pregnancy, which initially presented with intrahepatic cholestasis of pregnancy and intrauterine growth restriction. Risk factors, clinical presentation, diagnostic and treatment approaches, maternal and fetal outcomes, and their interrelations with spontaneous hepatic rupture in pregnancy or the puerperium, were recently investigated in an extensive review by Augustin et al. (2). Most common symptoms included upper abdominal pain, hemodynamic instability, nausea/vomiting, and shoulder pain. The most important risk factors were preeclampsia and HELLP (81% of patients with spontaneous hepatic rupture in pregnancy or the puerperium had HELLP) (2). Our patient presented with classical symptoms of upper abdominal and shoulder pain and developed spontaneous hepatic rupture in the right liver lobe in the third trimester at age 32, which is in accordance with the findings of Augustin et al. (2) who found that most cases occurred in third trimester, with a maternal median age of 31, almost all cases involved right liver lobe and 36.6% were nulliparous.

Unlike most other cases, the patient never developed fulminant HELLP (2).

The patient was initially examined due to intrahepatic cholestasis of pregnancy and a fetal ultrasound revealed intrauterine growth restriction which led to admission. Intrahepatic cholestasis of pregnancy is not known to have a relation to development of spontaneous hepatic rupture in pregnancy or the puerperium, and no other case reports has been identified with spontaneous hepatic rupture in pregnancy or the puerperium and intrahepatic cholestasis of pregnancy in the same patient.

Early recognition of the syndrome, an aggressive surgical approach, and blood component therapy contributed to the outcome. Clinicians should consider the diagnosis in patients with pre-eclampsia or HELLP and hemodynamic instability, upper abdominal pain, shoulder pain, nausea or vomiting.

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