

Spontaneous hepatic rupture during emergency cesarean section in a patient with HELLP syndrome: a case report and literature review

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Summary

Spontaneous hepatic rupture is a rare but life-threatening complication of pregnancy. *Case Report:* A 30-year-old woman with multi-para pregnancy was admitted to the hospital at 31+3 weeks of gestation who complained of abdominal pain localized in the right upper quadrant on the third day after admission. HELLP syndrome was diagnosed, and emergency cesarean section was performed. Within several hours, blood biochemistry revealed significant elevated liver enzymes and reduced platelet (PLT) count. HELLP syndrome was diagnosed and emergency cesarean section was performed. The authors also made a literature review on the reported cases with spontaneous hepatic rupture during pregnancy in mainland China. A total 39 cases were found. Maternal survival was 78.9% (30/38) and perinatal survival was 50% (21/42). Deaths occurred mainly due to disseminated intravascular coagulation (DIC), acute renal failure, respiratory failure, and so on. An emergency physician should be aware that early evaluation, appropriate diagnosis, and prompt surgical intervention are crucial to deal with such a life-threatening emergency. Multidisciplinary cooperation between obstetricians, surgeons, and pediatricians is required for treating spontaneous hepatic rupture during pregnancy.

Key words: Spontaneous hepatic rupture; Pregnancy.

Introduction

Spontaneous hepatic rupture is a rare but life-threatening complication of pregnancy. The incidence of spontaneous hepatic rupture during pregnancy has been reported to be one in 22,500 and one in 4,500 overall deliveries [1]. Spontaneous hepatic rupture is commonly secondary to the liver diseases, such as hepatitis, liver cirrhosis, and severe gestational hypertension [2]. This rare condition is primarily associated with HELLP syndrome during pregnancy. It occurs in less than 2% of HELLP syndrome cases, and very rarely occurs in patients with normal liver functions due to other diseases, such as acute infection and excessive muscle movements [3]. The maternal and fetal mortality rates due to spontaneous hepatic rupture remain high despite advanced diagnosis and treatment [4].

Herein, the authors present a patient with hepatic rupture occurring during emergency cesarean section and a literature review of the cases since 1990 was also made.

Case Report

A 30-year-old woman with multi-para pregnancy was admitted to the hospital at 31+3 weeks of gestation due to elevated blood pressure (160/110 mm Hg) and severe headache for 24 hours. She complained of abdominal pain localized in the right upper quadrant on the third day after admission. Within several hours, blood biochemistry revealed elevated liver enzymes increased remarkably [serum glutamic pyruvic transaminase (SGPT) of 465 U/L, serum glutamic oxaloacetic transaminase [SGOT] of 373 U/L,

reduced platelet (PLT) count of 28,000/mm³, hemoglobin level of 154 g/L, and LDH 1243 U/L. HELLP syndrome was diagnosed, and emergency cesarean section was performed. A female newborn, 1900 grams, had an Apgar score 7 at first minute and 9 at the fifth minute.

During surgery, the authors found profuse bleeding from the right upper quadrant of the abdomen with unknown origins. The abdomen was explored through a bilateral subcostal incision, and extensive rupture was found in the segments 6 and 7 of the liver. The liver was found to be enlarged with focal areas of necrosis as well as extensive and diffuse bleeding (Figure 1). Several compress drapes and hemostatic sponges were used to pack the liver for hemostasis. During surgery, the patient was transfused with a total of 14 units of packed red blood cells, 1,000 mL fresh frozen plasma, two units of platelets, and factor VII. Post surgery, the patient was transferred to the Intensive Care Unit (ICU) due to the severe clinical condition. Throughout recovery in ICU, she underwent anti-hypertensive medical therapy and parenteral nutrition. The day after first surgical operation, laboratory findings showed increase of total bilirubin (4.56 mg/dL), of liver enzymes (AST, 850 IU/L; ALT, 762 IU/L), low levels of Hb (8.2 g/dL), of Ht (22.4%), and of cholinesterase (2,890 IU/L; normal value 4,650–12,220). About five to six hours afterwards, a second laparotomy was necessary because hemoglobin levels were progressively reducing. Four days after the second laparotomy, hemocoagulatory and hepatic (liver enzymes, total bilirubin) laboratory findings were normal. A double drainage was left and then removed in the third postoperative day. She was discharged in good health and with normal laboratory parameters 14 days later. On follow-up, both the patient and baby were all healthy.

This study was approved by the Ethical Committee of the hospital, and a written consent for reporting was obtained from the patient.

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Figure 1. — Rupture is found in segments 6 and 7 of the liver. The liver is seen enlarged with focal areas of necrosis as well as with extensive and diffuse bleeding.

Discussion

The incidence of spontaneous hepatic rupture is reported to be one in 225,000 and one in 45,000 overall deliveries. The exact pathogenesis remains unclear yet. Researches have suggested that the complication may be associated with gestational hypertension and original liver diseases, accounting for 50% of the complications of HELLP syndrome, and DIC induced by preeclampsia and trauma has a great influence on the occurrence of hepatic rupture.[2]

In addition to the present case, a total of 38 cases have been reported in China (Table 1) [5-32]. The cases had a median age of 27 (range 22 to 43) years, and a median gestational age at delivery of 36 (range 28-41) weeks. Primipara accounted for 53.8% (21/39). Twin pregnancy was 7.7% (3/39). Pregnancy-induced hypertension syndrome is 76.9% (31/39), 7.7% (3/39) eclampsia, and HELLP syndrome is 25.6% (10/39). Regarding the history, hepatitis cholelith disease was found in one case, cholelithiasis in one, polycystic kidney in one, liver schistosomiasis in one, and liver hemangioma in one. Cesarean delivery accounted for 76.9% (31/39). Of the 39 cases, the most common was upper abdominal pain to the right shoulder radiation (23.1%), followed by abdominal pain (17.9%), upper abdominal pain with nausea and vomiting (17.9%), abdominal pain (12.8%), upper abdominal pain (10.3%), lower back pain (7.7%), chest tightness, abdominal distension (5.1%), and below xiphoid process under the pain to the right shoulder radiation (2.6%), and eclampsia coma (2.6%).

Among the reviewed cases, pregnancy-induced hypertension syndrome accounted for 76.9%, 7.7% of eclampsia and HELLP syndrome in the hepatic rupture occurred at 25.6%. One case in 39 patients had polycystic kidney with high blood pressure. The substantial liver lesions in pregnant women potentially increased the risk of hepatic rupture, such as hepatitis, hepatocellular carcinoma (HCC), liver schistosomiasis, liver hemangioma, and local liver

anomalies caused by cholelithiasis. Hepatitis was found in two cases, cholelithiasis in one, liver schistosomiasis in one, and liver hemangioma in one, HCC in one; other hepatic diseases include liver tissue blood clot, embolization of hepatic sinus expansion, liver cell degeneration, and hemorrhage around the lobule. Only two patients had normal liver. Hepatic rupture always occur in late pregnancy and postpartum within 48 hours. Among the 39 cases, 23.1% (9/39) occurred during the prenatal period, 51.3% (20/39) during cesarean section and 25.6% (10/39) at postpartum; in ten patients it occurred in five to 24 hour after delivery. Therefore, postpartum patients still should be monitored closely for occurrence of this disease.

The pathological classification of hepatic rupture includes three types: (1) hepatic rupture, (2) the subcapsular hematoma, and (3) the central type laceration. Rinehart *et al.* [1] reported that the most frequent signs and symptoms of hepatic rupture and hemorrhage was epigastric pain in 70%, followed by hypertension (66%), shock (56%), nausea and vomiting (25%), shoulder pain (21%), and headache (11%).

The diagnosis of hepatic rupture is difficult because symptoms are common and nonspecific. In the present study, hepatic rupture diagnosed in prenatal period accounted for only 23.1%. Ultrasound and CT is helpful to early diagnosis. Pregnant women with abdomen or the right epigastric discomfort with nausea and vomiting, should be checked closely to avoid delaying diagnosis. With ultrasound is difficult to find focal necrosis and wounds, but it can render diagnosis timely according to the location of coagulation clot and hemoperitoneum. Frequent contractions after labor, forcibly breathlessness, and intra-abdominal pressure change may induce hepatic rupture. Therefore, once hepatic subcapsular hematoma is prenatally diagnosed clearly, cesarean delivery should be performed immediately. In this series, 51.3% of hepatic ruptures were found during cesarean section. Hepatic rupture should be considered as a diagnosis in pregnancy complicated with the following conditions: (1) pregnancy hypertension, pre-eclampsia—abdominal pain—shock, or cirrhosis—abdominal pain—shock, (2) abdominal ultrasound found positive, and (3) abdomen puncture blood with no clotting.

In 1844, Abercrombie was the first to describe the complication, and surgery becomes the primary treatment [33]. In 1991, Smith *et al.* [34] advocated adjuvant therapies in the treatment of invalid persistent liver bleeding in patients with low blood pressure, which included removal of hematoma and drainage, when handling invalid, hepatic artery ligation and lobe resection were required. These opinions were of pioneering significance, placing the treatment of this disease into the new era of conservative surgery. In 1999, Barton and Sibai [35] proposed that hemodynamics and blood coagulation condition should be monitored closely for patients with conservative treatment, while at the same time using ultrasound and CT to evaluate liver in-

Table 1. — A literature review of the cases with spontaneous hepatic rupture during pregnancy.

Author, year	Age (yrs)	Parity	Gestational age (wks)	Preeclampsia	Treatment	Delivery	Maternal death (+)	Fetal death (+)
Zhong <i>et al.</i> , 1989 [5]	25	Multiparous	Term	+	Packing	Vaginal	-	+
	25	Primiparous	36+2	+	NR	Vaginal	NR	+
Yang <i>et al.</i> , 2006 [6]	32	Multiparous	34+5	HELLP	Hepatic artery ligation + packing	Cesarean	+	+
Cheng <i>et al.</i> , 1988 [7]	23	Primiparous	36+3	+	Packing	Vaginal	-	-
Liu <i>et al.</i> , 2002 [8]	27	Primiparous	31+3	+	Lobe resection	Cesarean	+	+
Zhang <i>et al.</i> , 2002 [9]	30	Multiparous	NR	+	Packing	Vaginal	-	-
	26	Primiparous	Term	-	Surgical stitching	Vaginal	-	-
Liu <i>et al.</i> , 2000 [10]	42	Multiparous	36	+	Surgical stitching + packing	Cesarean	-	-
	23	Primiparous	34	+	Packing	Cesarean	-	-
Lv <i>et al.</i> , 1995 [11]	25	Primiparous	37+	+	Packing	Cesarean	-	-
	30	Multiparous	40	+	Packing	Cesarean	-	+
Li <i>et al.</i> , 1992 [12]	24	Primiparous	34+5	+	Packing	Cesarean	+	+
Dong <i>et al.</i> , 2012 [13]	24	Primiparous	38+6	HELLP	Surgical stitching	Cesarean	+	-
Yue <i>et al.</i> , 2001 [14]	43	Multiparous	35	+	Surgical stitching	Cesarean	-	-
Huo <i>et al.</i> , 1995 [15]	32	Multiparous	34	+	Surgical stitching + packing	Cesarean	+	+
	42	Multiparous	41	+	Packing	Cesarean	-	-
Zhang <i>et al.</i> , 2001 [16]	23	Primiparous	34+2	+	Packing	Cesarean	-	-
	25	Primiparous	35+5	+	Packing	Cesarean	-	-
Gao <i>et al.</i> , 1998 [17]	38	Multiparous	38	+	Surgical stitching	Vaginal	-	-
	25	Primiparous	37+1	+	Packing	Cesarean	-	+
Chen H <i>et al.</i> , 2007 [19]	30	Primiparous	35+4	HELLP	Surgical stitching + packing	Cesarean	-	+
Wang, 2008 [20]	27	Primiparous	38+1	+	Surgical stitching	Cesarean	-	-
Lou <i>et al.</i> , 2008 [21]	34	Primiparous	36+2 (twins)	-	Surgical stitching	Cesarean	-	-,+
Hong <i>et al.</i> , 2001 [22]	26	Multiparous	35	-	Packing	Vaginal	-	-
He, 1992 [23]	22	Primiparous	36	-	Surgical stitching + packing	Cesarean	-	+
	22	Primiparous	30 (twins)	-	Hepatic artery ligation + packing	Cesarean	-	+
Sha <i>et al.</i> , 1994 [25]	27	Primiparous	39	-	Packing	Cesarean	-	-
	22	Primiparous	38	-	Surgical stitching	Cesarean	-	+
	39	Multiparous	37	-	Surgical stitching + Packing	Cesarean	-	-
	27	Multiparous	38	HELLP	Observation	Cesarean	-	-
	34	Multiparous	36	HELLP	Observation	Cesarean	-	-
	35	Multiparous	36+6	HELLP	Packing	Cesarean	+	+
Gao <i>et al.</i> , 2006 [27]	32	Multiparous	34+3	HELLP, (+)	Hepatic artery ligation + packing	Cesarean	+	+
Wang, 2006 [28]	41	Multiparous	28	HELLP	Hepatic artery ligation + packing	Cesarean	+	+
Tang, 2002 [29]	29	Primiparous	32 (twins)	-	Lobe resection	Cesarean	-	+
Cheng <i>et al.</i> , 2000 [30]	29	Primiparous	34	HELLP	Packing	Cesarean	-	+
Yu <i>et al.</i> , 1989 [31]	38	Multiparous	Term	Postpartum eclampsia	Packing	Vaginal	-	-
Mei <i>et al.</i> , 1991 [32]	25	Multiparous	32	+	Packing	Vaginal	-	+

jury situation, treating preeclampsia and HELLP syndrome, supplying blood product, in order to avoid direct or indirect liver surgery operation. Patients should undergo a radiological follow-up postpartum. All these strategies further improved the survival rate. In the reviewed cases, peri-he-

patic packing in 46.2% (18/39), liver repair in 30.8% (12/39), proper hepatic artery ligation 10.3% (4/39), liver resection 5.1% (2/39), 5.1% (2/39) were included as conservative treatment. The maternal mortality rate was reported to be 17%, and the mortality rate was 21.1% among

the reviewed cases. The median days of hospital stay was 15 (range 8-46) days. The reasons of death were mainly DIC, acute renal failure, respiratory failure, septic shock, acute respiratory distress syndrome (ARDS), cerebral edema, and cerebral hernia. Perinatal survival was 50% (21/42). Liver recovery of patients with hepatic rupture is slow, but all can recover completely, usually after four to six months when CT scan revealed a normal liver. Hepatic rupture in the next pregnancy in patients are rare, but incidence of obstetric complications of pregnancy are high, so the following pregnancies in these women still need to be monitored carefully.

In conclusion, hepatic rupture during pregnancy is associated with significant maternal and perinatal mortality and morbidity. Early evaluation, appropriate diagnosis, and prompt surgical intervention are crucial to deal with a life-threatening emergency. Management of such a situation requires close interdisciplinary cooperation between obstetricians and surgeons, and aggressive intensive care and surgical treatment.

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