

Case Report

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Surgical Management of Spontaneous Hepatic Rupture Associated with HELLP Syndrome: Case Presentation and Literature Review

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Abstract

Objective: HELLP syndrome is a serious complication in pregnancy characterized by hemolysis, elevated liver enzymes and low platelet count occurring in 0.2 to 0.6% of all pregnancies and in 10–20% of cases with severe preeclampsia. Spontaneous hepatic capsular rupture is a rare (0,015% of all pregnancies), but a life-threatening complication of HELLP syndrome with a mortality rate of up to 50% for both mother and infant.

Methods: We present the case of a 34-year-old nulliparous woman with a second trimester gestational hepatic rupture associated with HELLP syndrome.

A systematic literature search was conducted on database of Pubmed, Scopus, Medline and Embase.

Results: Spontaneous hepatic rupture is an infrequent but life-threatening condition of pregnancy associated with severe pre-eclampsia and HELLP syndrome.

Diagnosis is based on clinical suspicion. Abdominal CT with intravenous contrast is considered to be the best diagnostic method.

The management of this pathology requires a multidisciplinary approach between gynecologists, general surgeons and intensive care physicians. The main therapeutic objectives are: eliminate the causal factor, end the pregnancy and control hemorrhage. If there is not hepatic rupture and the patient is hemodynamically stable, conservative nonoperative management for selected patients has been recommended. However, if the patient presents hemodynamic instability despite adequate resuscitation or hemoperitoneum, it has been seen that surgery significantly reduces the mortality associated with this pathology.

Conclusions: Hepatic subcapsular hematoma and hepatic rupture are infrequent but life-threatening complications that can occur during pregnancy and are fundamentally related to pathologies such as preeclampsia and HELLP syndrome. The first step is to end the pregnancy and subsequently control the acute liver problem. Different surgical techniques have been described that should be individualized for each patient according to damage control principles. A high index of suspicion and prompt recognition allows a multidisciplinary approach and mortality can be reduced.

Keywords: Hepatic rupture; Hematoma; Preeclampsia; HELLP syndrome.

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Introduction

HELLP syndrome (hemolysis, elevated liver blood tests and low platelets) is a serious complication in pregnancy characterized by hemolysis, elevated liver enzymes and low platelet count occurring in 0.2 to 0.6% of all pregnancies and in 10-20% of cases with severe preeclampsia. Spontaneous hepatic capsular rupture is a rare (0,015% of all pregnancies), but a life-threatening complication of HELLP syndrome with a mortality rate of up 50% for both mother and infant. The clinical presentation of hepatic hematoma is generally non-specific and may include right upper quadrant or epigastric pain and vomiting. A high index of suspicion and prompt recognition allows a multidisciplinary approach and mortality can be reduced.

We present the case of a 34-year-old nulliparous woman with a second trimester gestational hepatic rupture associated with HELLP syndrome. We discuss the aetiology and treatment options involving this rare presentation.

Case description

A 34-year-old nulliparous woman, with no history of relevant medical conditions, was referred to the delivery room at 28 weeks of gestation complaining of 3 hour severe abdominal pain localized to the right upper quadrant. Her blood pressure was 160/115 mmhg. No data on arterial hypertension during pregnancy. Proteinuria 1000 mg/dL in urine test. Pre-eclampsia was diagnosed. The tracing on the fetal heart monitor indicated pathological fetal distress. In light of these findings she underwent an emergency caesarean section.

When entering in abdominal cavity a 1000 mL hemoperitoneum was detected. Hysterotomy was done and a 28 weeks alive fetus was obtained. It was subsequently converted to median laparotomy. A subcapsular hepatic hematoma involving the entire right lobe was found with hepatic rupture of segments V and VI and active hemorrhage (Figure 1). Subdiaphragmatic and hepatic packing was performed with eleven gauze pads placed above and around the liver in a routine manner. Finally a negative pressure open abdomen was performed. During surgery, the diagnosis of HELLP syndrome was confirmed with laboratory findings (hemoglobin 8.3 g/dL, platelets 66,000 mm³, AST 1404, ALT 770).

After hemodynamic stabilization of the patient an abdominal CT angiography was done confirming hepatic subcapsular and parenchymal hematoma, with multiple foci of active venous bleeding in the area of the hepatic dome and right hepatic lobe (Figure B).

The patient was admitted to ICU where aggressive management with fluid therapy was continued. Renal and respiratory function were preserved and treatment with broad-spectrum antibiotics was initiated due of febrile peaks.

A second-look surgery was performed 72 hours later, in which the packing was removed and a large organized subcapsular hematoma was observed involving the right hepatic lobe, with the left lobe appearing normal. Cholecystectomy was performed due to the presence of cholelithiasis to avoid ischemic/infectious complications during the postoperative period. There were no signs of active bleeding, so primary abdominal wall closure was performed.

The patient presented a satisfactory postoperative evolution with progressive improvement of the distributive shock. During her admission she required a total of two units of packed red blood cells and three units of fresh frozen plasma. In the immediate postoperative she presented amnesia and pregnancy denial. A cranial CT scan was performed with normal findings. Progressive recovery of platelet and liver enzyme levels (Table A), until normalization and resolution of her confusional syndrome. The patient was discharged on 18 postoperative day (POD).

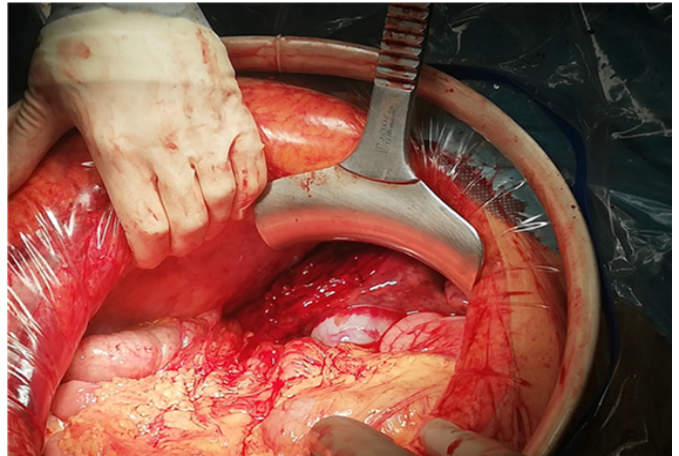


Figure 1



Figure 2

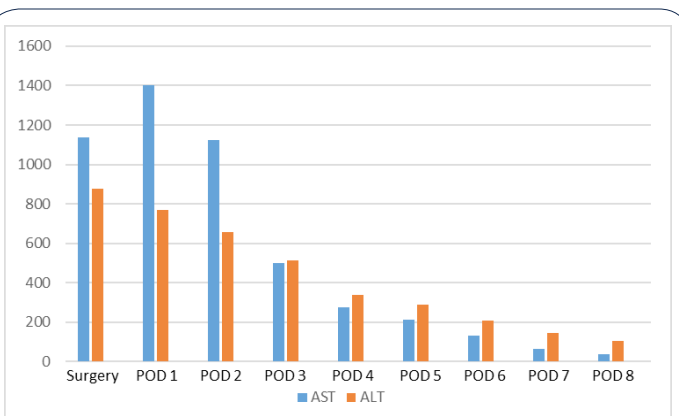


Table 1

Discussion

Spontaneous hepatic rupture is an infrequent but life-threatening condition of pregnancy associated with severe pre-eclampsia. Pre-eclampsia is defined as elevated blood pressure (>160/90 mmHg) with proteinuria (dipstick reading >1+ or >0,3 g of protein in a 24-hour urine) after 20 weeks of pregnancy or after delivery. It occurs in 8% of all pregnancies, and 10-15% of cases may be complicated by Hemolysis, Elevated Liver Enzymes and Low Platelet Count (HELLP) syndrome [1]. HELLP syndrome was first defined by Weinstein in 1982 [2]. It is a multisystemic disorder that carries significant maternal and perinatal morbidity and mortality. This syndrome has an incidence of 0,17 to 0,85% of all pregnancies, whereas the risk of recurrence in a subsequent pregnancy is about 19 to 27%. The incidence of spontaneous hepatic rupture is reported to be between 1 case in 45,000-225,000 pregnancies and only 1-2% of all of them in the context of HELLP syndrome [3].

The pathophysiology of HELLP syndrome is not well understood. The origin of preeclampsia/HELLP can be attributed to defective trophoblastic invasion that leads in a deficient placentation. All this induces a generalized vasoconstriction state secondary to dysfunction in the vascular epithelium, rather than the vasodilation typical of normal pregnancy. All this is manifested by an imbalance between vasodilator prostaglandins (prostacyclins) versus vasoconstrictors (thromboxanes) that generates an excess production of proinflammatory cytokines (IL-2, TNF) causing endothelial damage. Vasospasm produces a reduction in plasma volume leading to hemoconcentration and hypercoagulability. All this leads to multi-organ hypoperfusion that worsens the maternal and fetal condition [4]. A periportal hemorrhage and intravascular fibrin aggregation in the liver cause sinusoidal obstruction, intrahepatic vascular congestion, increased liver pressure, hepatic necrosis and intraparenchymal and subcapsular hemorrhage, which may result in capsular rupture [5].

Henny et al. described a series of 75 patients where hematomas are more frequent in the right hepatic lobe (75% of cases), compared to the left hepatic lobe (11%) or both (14%). In our patient the hematoma involved the right hepatic lobe, like the majority described [6].

The most common clinical signs of hepatic hematoma are right upper quadrant pain and/or epigastric pain (70-90%), right shoulder pain, right lung base hypoventilation, nausea and vomiting. These symptoms are supposed to be caused by the stretching of Glisson's capsule. Liver rupture is manifested as massive hypovolemic shock and abdominal distention. Most liver related complications of HELLP syndrome occur during the third trimester as in our case, late in the second trimester and also a few cases have been reported in literature during the immediate postpartum period [3].

Diagnosis is based on clinical suspicion and laboratory evidence of severe anemia, elevated liver enzymes and thrombocytopenia [7]. Abdominal CT with intravenous contrast is considered to be the best diagnostic method, being more sensitive than abdominal ultrasound. Nuclear magnetic resonance is also useful for surveillance of chronic hematomas [8]. In our case no imaging test was needed because the blood in abdominal cavity was observed during the emergency caesarean section.

The management of this pathology requires a multidisciplinary approach between gynecologists, general surgeons and intensive care physicians. The main therapeutic objectives are: eliminate the causal factor, end the pregnancy (emergency caesarian section) and control hemorrhage (exploratory laparotomy). Hemodynamic stabilization with aggressive volume resuscitation and blood products represents a fundamental point of early support [9]. If there is not hepatic rupture and the patient is hemodynamically stable, conservative nonoperative management for selected patients has been recommended consisting on close monitoring of the patient and imaging test controls [9,10]. However, if the patient presents hemodynamic instability despite adequate resuscitation or hemoperitoneum, it has been seen that surgery significantly reduces the mortality associated with this pathology [7,10].

In the cases of hepatic rupture, achieving liver hemostasis though surgery is difficult because of the presence of multiple areas that are affected by infraction and hematomas and especially because of coagulopathy. Surgical management of these patients must follow the same principles as damage control surgery. Multiple surgical options have been described: hemostasis with specific devices, hepatorrhaphy, vessels ligation, hepatic artery ligation, collagen sponges, argon laser and perihepatic packing followed by a second-look surgery [3]. Transcatheter Arterial Embolization (TAE) is a well-recognized treatment method that can be performed as an alternative or as a complement to surgery. In-Chul Nam et al. described that TAE is useful and less invasive than other techniques and recommended it as a bridge between perihepatic packing and hepatectomy if necessary [5]. The use of hemostatic meshes has also been described, which have demonstrated their effectiveness in managing ruptured hematomas, with less need for reinterventions [11]. In case of massive hematoma or necrosis of the hepatic parenchyma, partial hepatectomy or liver transplantation may be necessary.

In our reported case, the diagnosis was intraoperative during cesarean section and liver packing was performed due to the extensive capsular rupture in the liver, making it impossible to achieve hemostasis in any other effective way. A CT angiography was performed to assess possible arterial bleeding that could be embolized, but this was not confirmed. A second-look laparotomy was performed 72 hours later, verifying resolution of the bleeding with the initial packing.

Conclusion

Hepatic subcapsular hematoma and hepatic rupture are infrequent but life-threatening complications that can occur during pregnancy and are fundamentally related to pathologies such as preeclampsia and HELLP syndrome. The first step is to end the pregnancy and subsequently control the acute liver problem. Different surgical techniques have been described that should be individualized for each patient according to damage control principles. A high index of suspicion and prompt recognition allows a multidisciplinary approach and mortality can be reduced.

Conflict of interest: The authors declare no conflict of interest.

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