

Management of Spontaneous Hepatic Rupture on Top of HELLP Syndrome: Case Report and Review of the Literature

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Summary

Introduction: We report the case of a patient with antepartum HELLP syndrome and simultaneous rupture of the right liver lobe. An emergency caesarean section was performed and the liver rupture was managed surgically via perihepatic packing. The mother and her child recovered well and were discharged 19 days after admission. **Case Report:** We describe a case report and review the literature. Based on our own experience and the most common clinical presentations of such patients, we were able to establish an algorithm for managing such cases. **Conclusion:** An association between liver rupture and HELLP syndrome is rare but was previously described in several case reports. In pregnant women with HELLP syndrome and acute onset abdominal pain, a potential spontaneous hepatic rupture should be taken into consideration.

Introduction

The HELLP syndrome is a serious variant of preeclampsia which is characterized by a triad of haemolysis (H), elevated liver enzymes (EL), and low platelet count (LP). It has a low incidence of 0.5–0.9% of all pregnancies. The majority of cases occur before delivery, mostly between the 27th and 37th week of gestation, while the rest of

cases develop within 48 h after delivery [1]. HELLP syndrome occurring postpartum is usually associated with a higher rate of maternal morbidity and mortality, with a higher risk of developing complications such as pulmonary oedema, renal failure, disseminated intravascular coagulation (DIC), and subcapsular liver haematoma [2]. Because of that a fast diagnosis and initiating therapy is the most important principle in the treatment of those patients. The induction of labour or even an emergency caesarean section has to be done immediately. Common symptoms associated with HELLP syndrome include right upper quadrant (RUQ) abdominal pain, epigastric pain, nausea, vomiting, and headache. These non-specific symptoms always lead to a delay of the diagnosis; however, the epigastric and/or RUQ pain are considered to be the most alarming symptoms of this syndrome which are assumed to be caused by stretching of Glisson's capsule due to sinusoidal obstruction of blood flow [3]. Diagnosis of HELLP syndrome requires the presence of all of its three major components, while partial or incomplete HELLP syndrome may include only one or two elements of the triad (H or EL or LP) [4–7]. Spontaneous rupture of a subcapsular liver haematoma in pregnancy is a rare incident occurring in 1/40,000–1/250,000 deliveries and in about 1 to <2% of the cases with HELLP syndrome. However, it is a potentially fatal complication. The right liver lobe is the most common site of spontaneous rupture [8–12]. The symptoms are sudden onset severe pain in the epigastric and abdominal RUQ radiating to the back, right shoulder pain, anaemia, and hypotension. The condition may be diagnosed by ultrasound, computed tomography, or magnetic resonance imaging examination [8, 10, 11, 13]. Hepatic rupture may also occur postpartum [14]. We would like to present our management of a case of a spontaneous hepatic rupture with formation of a subcapsular haematoma that was encountered during an emergency caesarean section in a patient diagnosed with antepartum HELLP syndrome.



Fig. 1. Intraoperative view after removal of the liver packs.

Case Report

A 29-year-old nulliparous primigravida (G1P0) in the 34th gestational week was referred from the secondary care to the Department of Gynaecology and Obstetrics at our facility because of a concurrently diagnosed HELLP syndrome. The clinical examination on admission revealed abdominal pain without tenderness or rigidity and a foetal bradycardia of 80 beats/min which was refractory to treatment with i.v. administration of 25 µg Fenoterol-HBr (Partusisten®; Boehringer Ingelheim, Ingelheim am Rhein, Germany). Thus, an emergency caesarean section was indicated and performed immediately. On opening the peritoneum we encountered an unexpected gush of blood which raised the suspicion about a concomitant hepatic rupture on top of HELLP syndrome. We found a premature detachment of placenta which explained the foetal bradycardia. After performing the quick delivery followed by suturing of the uterus, we started the palpation of the solid abdominal organs as a potential source of bleeding. The surface of the liver was irregular on palpation. In order to gain an adequate access to the abdominal cavity, we extended the Pfannenstiel incision pararectally at the right side and started the intra-abdominal exploration to identify the source of bleeding. On inspection, a localized rupture of the right lobe of the liver with formation of a subcapsular haematoma was noted (stage Moore II). With absence of signs of hepatic necrosis we performed the perihepatic packing as a primary management of the bleeding. A vicryl mesh was first laid in direct contact with the bleeding surface of the liver; then the packs were applied above it in order to prevent the direct contact between the bleeding liver surface and the packs. Hence, we could avoid shearing of the newly formed fibrin mesh on their removal. During the planned second-look laparotomy, which was performed 48 h later, we could safely remove the packs with absence of signs of any potential bleeding (fig. 1). Postoperative abdominal ultrasound showed an irregular hepatic surface with remnants of the evacuated subcapsular haematoma.

The patient was initially admitted to the surgical intensive care unit and then transferred to the maternity ward after stabilization of her condition. Her baby daughter was admitted to the neonatal intensive care unit for monitoring as well. After a hospital stay of 19 days in total the mother and her daughter could be discharged in a good condition.

Admission Laboratory

White blood cells 13,000/µl; haemoglobin 11 g/dl; thrombocytes 46,000/µl; aspartate aminotransferase (AST) 324 U/l; alanine aminotransferase (ALT) 341 U/l; γ-glutamyl transpeptidase (GGT) 50 U/l; alkaline phosphatase (AP) 162 U/l; lactate dehydrogenase (LDH) 503 U/l; C-reactive protein (CRP) 3.8 mg/dl; haptoglobin <10 mg/dl. Other laboratory values were unremarkable.

The trend of the parameters is shown in figures 2–5.

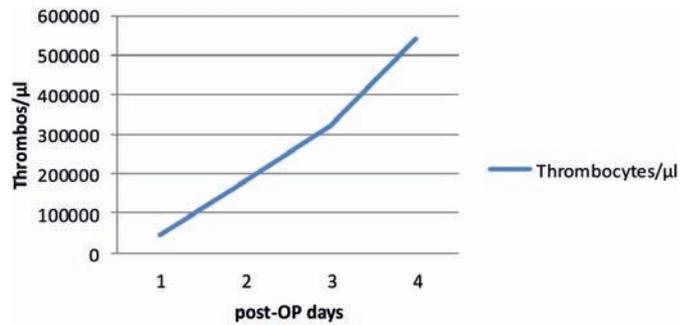


Fig. 2. Laboratory values for thrombocytes.

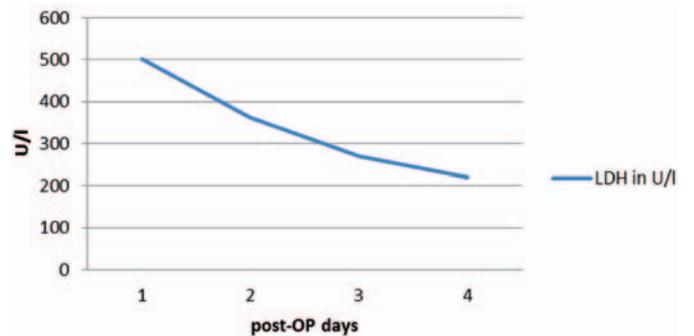


Fig. 3. Laboratory values for LDH.

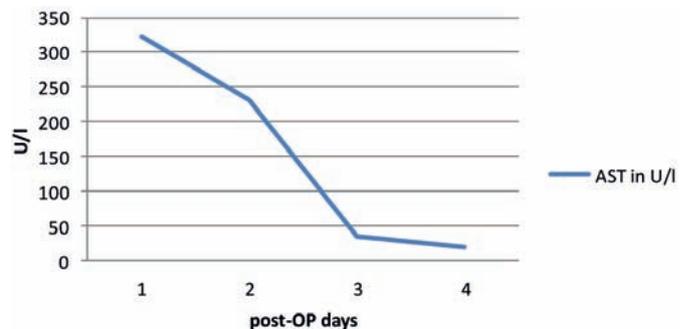


Fig. 4. Laboratory values for AST.

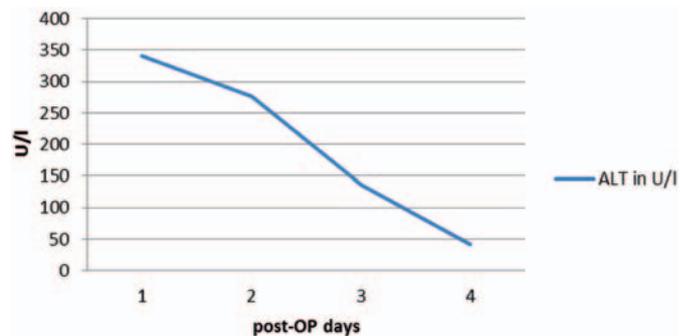


Fig. 5. Laboratory values for ALT.

Discussion

Spontaneous hepatic rupture occurring during pregnancy, though rare, is a potentially fatal incident. It is almost always associated with preeclampsia, eclampsia, and/or HELLP syndrome which always carry an increased risk of both maternal and foetal morbidity and mortality that was reported to be as high as 40–80% [8]. Most of the cases of HELLP syndrome develop before delivery, typically between the 27th and the 34th week of gestation, as in our case presentation here. In spite of the fact that the mechanism leading to hepatic rupture is not exactly known, the syndrome seems to be the final manifestation of an insult that leads to microvascular endothelial damage and intravascular platelet aggregation [15]. These hepatic ruptures are almost always diagnosed intraoperatively during a caesarean section or in the early postpartum period in patients suffering from severe abdominal pain typically affecting the epigastrium and the right hypochondrium. Ultrasound can detect these liver insults preoperatively; however, most of the patients present to the hospital in an acute maternal and/or foetal condition that necessitates an urgent intervention in the form of an emergency caesarean section and allows only a narrow time frame for the completion of preoperative investigations. The standard management in cases diagnosed with HELLP syndrome with an anticipated hepatic rupture is an emergency caesarean section followed by immediate exploratory laparotomy. The transfusion of blood components is mandatory. Some authors recommend conservative non-operative treatment in some highly selected cases [8]. Nevertheless, the standard procedure has to be the induction of labour – even by caesarean section, if necessary.

Therapeutic options in the case of a subcapsular liver haematoma or even liver rupture range from conservative treatment to surgery including hepatectomy or even transplantation. Besides the preservation in an intensive care unit, the aim of the therapy is to control the bleeding or even to stop it.

Whereas the concept of conservative management of patients with isolated subcapsular liver haematoma is well established in the literature, we consider it to be inapplicable in patients suffering from the same incident occurring during pregnancy on top of HELLP syndrome. The reason for this is the increased risk of bleeding on top of DIC arising as a result of thrombocytopenia. Thus, we recommend surgical intervention in the form of an emergency caesarean section with exploratory laparotomy to be the first

line of treatment in such cases. Operative management of these hepatic ruptures can vary from a simple haematoma evacuation with insertion of drains, passing by liver packing, up to performance of an extended hepatic resection, and ending with liver transplantation [16, 17]. In cases of preeclampsia and eclampsia it may be taken into consideration to apply a combination of conservative and interventional managements [13]. Vigil-De Gracia and Ortega-Paz [18] evaluated all reported cases with HELLP syndrome and hepatic rupture between 1990 and 2010. N = 13 patients were treated by transplantation. Among these patients, the overall survival was over 90% and therefore better than in the other therapy groups [18]. Liver transplantation seems to be a good option but remains an exceptional decision [8].

Conclusion

While preeclampsia and eclampsia are more common among primigravidas, spontaneous hepatic rupture is more commonly encountered in multigravidas. The combination of spontaneous hepatic rupture on top of HELLP syndrome is much more common among multigravidas [19].

According to the evaluation of the risk factors for developing spontaneous hepatic rupture during pregnancy by Vigil-De Gracia and Ortega-Paz [18], preeclampsia and eclampsia do not seem to constitute a real risk. Nevertheless, the risk is markedly increased once the diagnosis of HELLP syndrome has been made [18].

In conclusion, a spontaneous hepatic rupture should always come into consideration as a potential diagnosis in pregnant patients presenting to the hospital with a sudden onset of epigastric and/or right hypochondrial pain accompanied with manifestations of early haemodynamic shock, e.g. tachycardia, hypotension, etc. The involvement of a multidisciplinary team including experienced obstetricians, hepatobiliary surgeons, and neonatologists is the cornerstone of the management of these cases to enhance the survival chances of both the mother and the newborn.

Disclosure Statement

The authors report no declarations of interest.

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