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Acute liver necrosis in the HELLP syndrome: successful outcome after orthotopic liver transplantation. A case report

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Abstract. We discuss the case of a 30-year-old primipara woman who developed a liver rupture as a complication of the HELLP syndrome. A liver necrosis and bleeding made a hepatectomy necessary. A portocaval shunt was able to maintain the patient until she underwent urgent liver transplantation. In an excellent state of recovery, the woman and her baby were discharged from the hospital 66 days after having been admitted.

Key words: Liver transplantation, HELLP syndrome – HELLP syndrome, liver transplantation – Preeclampsia, liver rupture

Introduction

The HELLP syndrome, which comprises the symptoms of hemolysis (H), elevated liver enzymes (EL), and low platelets (LP) during preeclampsia, was first described by Weinstein in 1982 [22]. It is important to recognize the laboratory and clinical signs of this syndrome since urgent delivery of the child is often necessary in order to save the lives of both mother and child. The following case report focuses on the severe complication of liver rupture in a woman with the HELLP syndrome during pregnancy who developed a liver necrosis, making emergency liver transplantation necessary.

Case report

A 30-year-old, 27-week-pregnant woman was admitted to a local hospital for the treatment of hypertension and edema. Her antenatal history had been uneventful before this admission. Upon admission a severe gestosis with hypertension and fetal distress was diagnosed. Over the next 3 weeks, the woman's platelet count dropped to 90,000/mm^3 and her liver enzymes rose to 560 IU/l for SGOT and 590 IU/l for SGPT. The diagnosis of HELLP syndrome was considered and a cesarean section was performed immediately. A healthy baby weighing six pounds was delivered.

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On the 2nd postoperative day the woman developed hemolysis, followed by progressive renal failure and symptoms of shock. Abdominal ultrasound showed a fluid collection in the right paracolic gutter, which led to an emergency laparotomy. A rupture of the right lobe of the liver was found, together with diffuse parenchymal bleeding. Although surgical control of the hemorrhage was possible with abdominal packs, the patient's condition worsened the following day, which made transferral to our hospital necessary. We diagnosed disseminated intravascular coagulation (DIC), acute renal failure, and progressive shock with coma. Liver enzymes were highly elevated and platelet count had dropped to 60,000/mm^3 (Fig. 1).

Cardiopulmonary resuscitation had to be performed during admission and again in the intensive care unit (ICU).

After adequate fluid resuscitation, the woman was reoperated. A large hematoma of the right liver lobe with severe bleeding was found, while the left lateral segment of the ruptured liver showed only slight edematous changes. A right-sided, extended hemihepatectomy was performed. Over the next 20 h the patient developed a metabolic acidosis and became hemodynamically unstable. Another laparotomy revealed a total necrosis of the remaining segments of the liver. The decision was made to remove the remaining segments of the liver in order to stop the bleeding.

A direct shunt was created between the portal vein and the vena cava. After readmission to the ICU, the patient was kept alive with plasmapheresis for the removal of toxic metabolites. Within the next 20 h it was possible to find a suitable, but not blood group-compatible, liver (recipient B, donor AB) and an orthotopic liver transplantation (OLT) was successfully performed.

As shown in Fig. 1, fibrinogen normalized within 48 h after transplantation. Platelet count rose to normal values within 10 days. On the 3rd postoperative day, a moderate rejection episode was treated with 500 mg of prednisone. Liver function recovered and the woman was given triple drug immunosuppressive therapy consisting of low-dose prednisone, azathioprine, and cyclosporin.

Over the next 9 days the patient awoke from her coma and recovered renal function. Ten days later, the woman was transferred to a regular ward. After 66 days in the hospital, the woman and her baby were discharged in excellent condition.

Discussion

The case presented here meets the diagnostic criteria for a severe form of HELLP syndrome, the etiology of which still remains unknown. A number of theories are under investigation [1, 5, 7, 13]. Weinstein [22] considered the syndrome to be a unique form of severe preeclampsia and called it the HELLP syndrome in reference to the

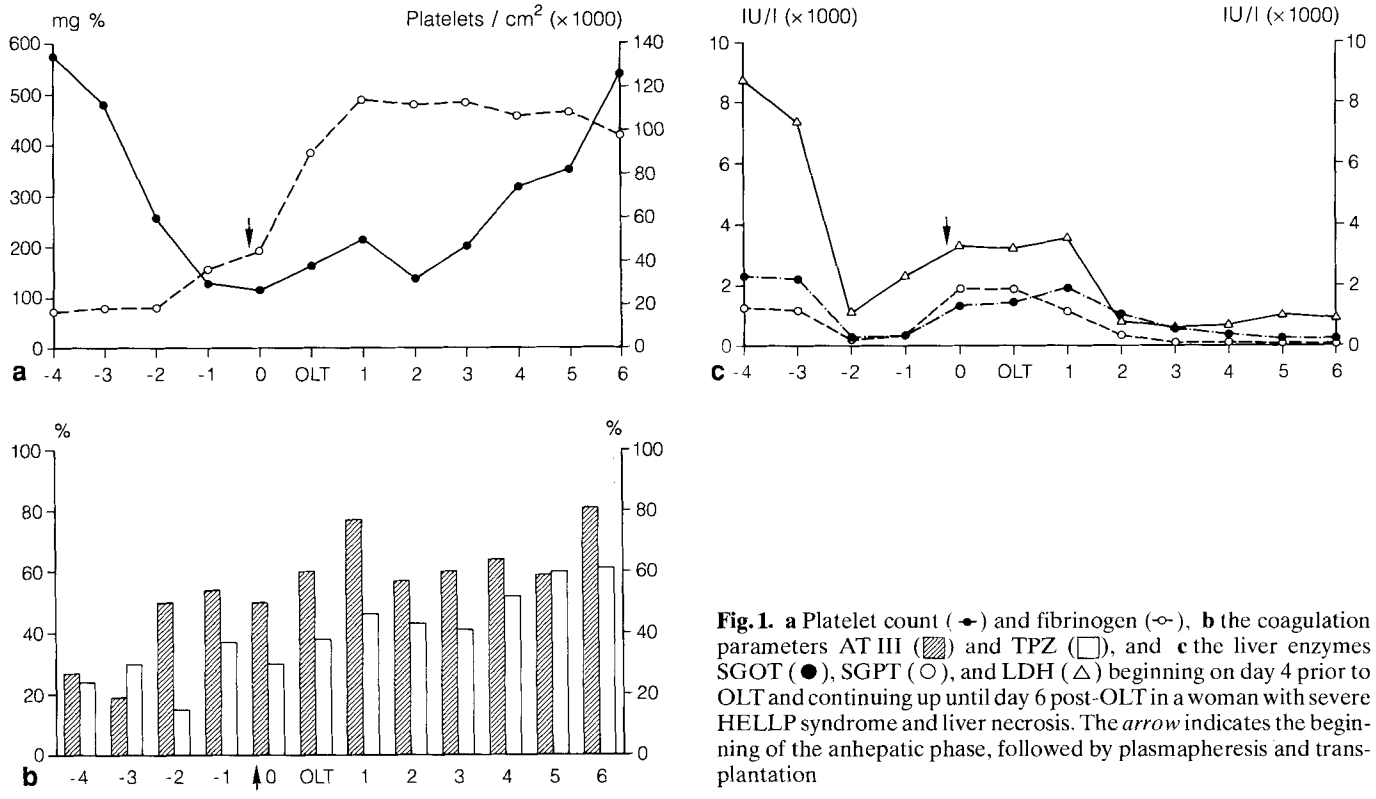

Fig. 1. **a** Platelet count (-•-) and fibrinogen (-○-), **b** the coagulation parameters AT III (▨) and TPZ (□), and **c** the liver enzymes SGOT (●), SGPT (○), and LDH (△) beginning on day 4 prior to OLT and continuing up until day 6 post-OLT in a woman with severe HELLP syndrome and liver necrosis. The *arrow* indicates the beginning of the anhepatic phase, followed by plasmapheresis and transplantation

laboratory data. Other authors have classified the syndrome according to the severity of the gestosis [13] or to maternal obstetric factors, such as hypertension, vomiting, etc. [10, 15]. Still others have suggested toxic factors or immunological factors, such as complement activation [3, 7, 18].

However one wishes to classify it, management of the syndrome is still controversial [10, 12, 13, 15, 19], with some authors recommending immediate delivery of the child in order to alleviate symptoms and others preferring a more conservative approach that involves allowing the pregnancy to go to term.

Depending on the age and race of the women involved, the incidence of eclampsia is 1 in 150–200 pregnancies. In about 10% of all cases of eclampsia, the HELLP syndrome may arise. The mortality rate is reported to be up to 3% [13, 15, 16, 22]. It is as yet unknown why some patients develop liver rupture and hepatic hemorrhage in a more severe form of the syndrome. When this complication occurs, the mortality rate increases to 60% [3, 9, 18]. Besides liver rupture, the development of adult respiratory distress syndrome (ARDS) has also been observed [13].

Diagnosis of the HELLP syndrome is based on clinical symptoms [3, 13, 21] and can only be ascertained by laboratory investigations [13]. Important are the coagulation parameters such as AT III, changes in liver enzymes, and platelet count. Most of the severe cases require an urgent cesarean section [3, 4, 9, 10, 13, 15, 18, 19]. Rapid, progressive liver and kidney dysfunction associated with coma are complications in the early phase of the severe form. The risk for severe complications continues after delivery of the child [13, 15].

Abdominal ultrasound seems to be the method of choice to diagnose alterations in liver structure at the early stage [2, 20]. However, only abdominal CT scan is a reliable diagnostic tool in cases of suspected liver rupture and hepatic hemorrhage [6]. In these cases the patient should be transferred to a liver center. If an operation becomes inevitable, a step-by-step approach should be followed according to the clinical and histological progression of the liver necrosis. Depending on these findings, an aggressive surgical approach may have to be taken at an early stage.

The decision to perform a hepatectomy can be life-saving, as in the case reported here [17]. However, removal of the whole liver should not be considered the method of choice; packing or extended resection of the necrotic liver tissue would be a preferable first step of any surgical maneuver. Alternative methods, such as ligation of the hepatic artery [14] or transcatheter embolization [11] to stop the hepatic hemorrhage, should be avoided in cases of extended liver rupture [9].

After hepatectomy we used the technique described by Ringe et al. [17] to leave the vena cava in situ and to create a direct, sutured, end-to-side portocaval shunt. This method of performing a hepatectomy and then inserting a shunt was used first and foremost in cases of acute hepatic failure or primary nonfunctioning livers after transplantation. The rationale was to remove the necrotic tissue as the cause of ongoing septic complications.

An alternative technique has been suggested by Husberg et al. [8], who devascularized the necrotic liver only and left the organ in situ. In our view, one should bear in mind the possibility that in severe HELLP syndrome, an urgent liver transplantation may be indicated.

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