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CASE REPORT

Postpartum HELLP syndrome—the case of lost battle

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Abstract

Unexpected rapid maternal death after delivery due to HELLP syndrome is rarely encountered and may become the subject of forensic expertise. Unexpectedness, suddenness, and fulminant course of this syndrome as well as absence of classical signs of pre-eclampsia can confuse physicians and lead to diagnostic delay. A definitive post-mortem diagnosis of HELLP syndrome in questionable cases of maternal death should be based on accepted laboratory criteria and characteristic histopathological alterations. We present a case of acute postpartum HELLP syndrome complicated by disseminated intravascular coagulation and acute renal failure which caused rapid maternal death only 20 hours after a caesarean section following an uncomplicated pregnancy.

Key words: *Acute renal failure, DIC, HELLP syndrome, maternal death*

Introduction

The HELLP syndrome is a serious complication in pregnancy characterized by haemolysis, elevated liver enzymes, and low platelet count, occurring in 0.5%–0.9% of all pregnancies. About 70% of the cases develop before delivery, the majority between the 27th and 37th gestational weeks; the remainder within 48 hours after delivery (1). There are no differences in laboratory findings between HELLP syndrome before and after delivery; however, women with postpartum HELLP syndrome have significantly higher incidences of complications such as: pulmonary oedema, renal failure, disseminated intravascular coagulation (DIC), and subcapsular liver haematoma (2). HELLP syndrome is associated with serious maternal morbidity, especially when it arises in the postpartum period. Since Weinstein's publications in 1982 maternal death from HELLP syndrome is rarely encountered in forensic pathology (3). We report a case of an unexpected rapid maternal death caused by postpartum HELLP syndrome complicated by DIC and multiorgan dysfunction syndrome (MODS).

Case report

The 31-year-old white primipara with twin pregnancy was admitted to hospital in the 38th week of gestation with elevated blood pressure (150/100 mmHg). After receiving antihypertensive treatment blood pressure was 120/80. All laboratory variables, including plasma proteins, were within their respective reference intervals. Few hours after admission to the hospital the contractions started, and the caesarean section was performed because of vertex-transverse presentation of twins. Liveborn female and male were delivered. The postoperative course was initially inconspicuous. Four hours postpartum she experienced sudden epigastric pain. The blood pressure rose to 190/130. Laboratory findings showed haemolysis, thrombocytopenia, and an increase in serum creatinine and aminotransferases (Table I). Intravenous magnesium sulphate was administered. Abdominal ultrasound disclosed an empty uterine cavity without placenta residue. Five hours postpartum, the patient was transferred to the intensive care unit because of poor urine output, drowsiness, and suspicion of DIC. On examination

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Table I. Laboratory findings on the first day postpartum.

Laboratory findings	Time postpartum		
	4 h	10 h	19 h
Haemoglobin (g/L)	76	88	104
Platelet count ($\times 10^9$/L)	69	38	43
Serum urea (mmol/L)	6.1	7.1	14
Serum creatinine (μmol/L)	98.2	170.9	190.5
Total bilirubin (μmol/L)	21.4	39.3	53.7
AST (IU/L)	456	772	966
ALT (IU/L)	185	359	651
LDH (IU/L)	1000	734	8478
Fibrinogen (g/L)	7.1	1.61	0.55
D-dimer (μg/L)	–	894	985
APTT (s)	47.8	79.1	105
PT (s)	30.1	392	44.3
INR	1.8	2.68	4.12

ALT = alanine aminotransferase; APTT = activated partial thromboplastin time (in seconds); AST = aspartate aminotransferase; INR = international normalized ratio (prothrombin complex); LDH = lactate dehydrogenase; PT = prothrombin time (in seconds).

she was sleepy and disoriented. The combination of haemolysis, thrombocytopenia, and elevated liver enzymes suggested a postpartum HELLP syndrome, complicated by DIC. The patient was rehydrated, and treatment was instituted with fresh frozen plasma, red cell transfusion, fresh platelets, and kybernin P (antithrombin III). High serum urea, creatinine, and persistent anuria were compatible with acute renal failure. A high dose of furosemide failed to increase diuresis. Sixteen hours postpartum her blood pressure was 60/40 and oxygen saturation 70%. Few minutes later the patient had acute cardiac arrest, and resuscitation started. Resuscitation was successful, and normal heart action was re-established; blood pressure rose to 150/110, and oxygen saturation was 90%. Laboratory findings were deteriorated, and development of acute renal failure indicated the necessity of urgent dialysis. Following dialysis, the patient was stable, oxygen saturation 98%. Twenty hours postpartum patient developed again cardiopulmonary arrest. Despite resuscitation attempts the outcome was lethal.

Due to the fact that death occurred in the hospital a short time after a surgical intervention, a medico-legal autopsy was ordered. An autopsy revealed oedema of the brain and lungs as well as dilatation of right and left ventricles. Hydrothorax (900 mL), hydroperitoneum (2500 mL), and hydropericardium (200 mL) were present. The field of operation of the caesarean section was unremarkable. Petechial and suffusional haemorrhages were observed under the pleura, endocardium, in the mucosae of the renal pelvis, and peritoneum of the small and large bowel. Histology revealed periportal hepatocellular necrosis, bloodless glomeruli with swollen and vacuolated intracapillary cells, as well as confluent haemorrhages in kidneys, liver, and spleen. Death was attributed to multiple organ failure due to DIC as a consequence of HELLP syndrome.

Discussion

HELLP is a multisystem disease, resulting in generalized vasospasm, microthrombi formation, and coagulation defects. The syndrome seems to be the final manifestation of insult that leads to microvascular endothelial damage and intravascular platelet aggregation (4).

Up to 30% of all patients who develop HELLP syndrome will develop this syndrome after parturition, typically within 48 hours. Unexpectedness, suddenness, and fulminant course of this syndrome are essential. The usual short period of observation after an uncomplicated delivery and uneventful medical history contributes to the risk of missing a life-threatening complication. The first, often the sole but always the most important symptom of this syndrome is epigastric pain which is assumed to be caused by stretching of Glisson's capsule due to sinusoidal obstruction of blood-flow. However, such non-specific abdominal symptom may lead to diagnostic delay (5).

The HELLP syndrome usually occurs with pre-eclampsia. However, in 20% of cases there may be no evidence of pre-eclampsia before or during labour, as in this pregnancy which was regularly controlled. The patient had only one isolated episode of hypertension, and all laboratory findings were normal. With regard to the forensic aspects of a timely diagnosis, an absence of obvious signs of pre-eclampsia significantly affected the clinicians who could not predict the development of HELLP syndrome (6,7).

Clinically evident DIC as a secondary pathophysiological phenomenon to the primary process is seen in 4%–38% of the patients, suggesting an important role of coagulopathy in the aetiology of HELLP syndrome. The development of a decompensation of coagulation correlates with the appearance of severe maternal complications such as renal failure (8). Acute renal failure (ARF) complicating HELLP is unusual but a life-threatening complication. It occurs in only 2%–3% cases of HELLP syndrome, but it can be the cause of maternal death in 56%–61% of all cases. In most cases ARF is caused by acute tubular necrosis with favourable renal outcome, but it

can also be caused by cortical necrosis which leads to irreversible renal damage, as in this case (9,10). Although severe maternal complications as a sequel to HELLP syndrome in the postpartum period are reported to be rare events, in this study the maternal course was complicated by deterioration of DIC with subsequent multiple organ failure and death. The fact that development of complications cannot usually be predicted is of special forensic significance in considering timely medical interventions.

The predominant morphological post-mortem findings in the case of maternal death from HELLP syndrome were petechial haemorrhages and suffusions in the skin, mucous surfaces, and serous coats of internal organs that can be attributed to DIC and an almost identical feature of histopathological alterations in the liver and kidneys. These histopathological alterations can also be found in the literature as a characteristic of HELLP syndrome (11,12).

When examining fatalities that have occurred during pregnancy or postpartum, the forensic pathologist should take HELLP syndrome with fatal outcome as a potential differential diagnosis into consideration. A definitive post-mortem diagnosis of HELLP syndrome must be based on laboratory criteria and characteristic histopathological alterations. The histopathological alterations in liver and kidneys can be considered characteristic for the disease, and their presence may enable the forensic pathologist to establish the definite post-mortem diagnosis of HELLP syndrome.

Declaration of interest: The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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