HELLP SYNDROME: A CASE REPORT WITH GUIDELINES FOR DIAGNOSIS AND MANAGEMENT

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SUMMARY

A 25-yr-old patient with a twin pregnancy of 34 weeks gestation developed HELLP syndrome and required urgent delivery by Caesarean section. Before operation, a central venous catheter and urinary catheter were inserted, and fresh frozen plasma and platelets were administered to correct hypovolaemia and severe thrombocytopenia. This case demonstrates the critical condition of these patients before operation and that extensive preoperative preparation and invasive monitoring are necessary for successful management. The choice of anaesthetic is governed by the presence of liver and renal dysfunction and severe thrombocytopenia.

KEY WORDS


"Haemolysis (H), elevated liver enzyme activity (EL) and low platelet count (LP)", (HELLP syndrome), named by Louis Weinstein of the University of Arizona, is a rare manifestation of hypertensive diseases of pregnancy and represents the most severe end of the pre-eclampsia spectrum [1]. The syndrome is of interest to the anaesthetist as many of these patients may require urgent delivery by Caesarean section in an effort to halt the disease process. Preganancies complicated by this syndrome are associated with poor maternal and fetal outcome. The mothers are at high risk of developing acute renal failure, shock lung, abruptio placenta and, occasionally, liver rupture with a maternal mortality in the range 2–24% [2]. This poses a considerable challenge to the anaesthetist, who may be presented at short notice with one of these patients for emergency Caesarean section.

CASE REPORT.

A 25-yr-old woman with a twin pregnancy of 34 weeks gestation was admitted to the maternity unit complaining of nausea and vomiting, epigastriic pain and loose bowel motions. She was not in labour on admission and her vital signs were normal. Arterial pressure was 130/80 mm Hg, there was no evidence of peripheral oedema, and urinalysis was negative for protein, glucose and acetone. Her haemoglobin was 11 g% and platelets were 100,000 mm$^3$. Her antenatal history was uneventful before this admission.

Over the next 48 h the patient’s arterial pressure increased to 160/95 mm Hg and she developed proteinuria and peripheral oedema. Laboratory investigations included haemoglobin (10.4 g%) and a platelet count (22,000 mm$^3$). Prothrombin time, activated partial thromboplastin time and serum concentration of fibrinogen were normal; fibrin degradation products were 80 µg litre$^{-1}$ (normal value < 10 µg litre$^{-1}$). Liver function tests included decreased concentrations of total protein (57 g litre$^{-1}$; normal range 60–80 g litre$^{-1}$); albumin 33 g litre$^{-1}$ (normal range 34–48 g litre$^{-1}$); phosphate 0.73 mmol litre$^{-1}$ (normal range 0.8–1.45 mmol litre$^{-1}$); lactic dehydrogenase 295 u litre$^{-1}$ (normal range 60–200 u litre$^{-1}$); alanine aminotransferase 159 u litre$^{-1}$ (normal range 0–45 u litre$^{-1}$); aspartate aminotransferase 200 u litre$^{-1}$ (normal range 0–40 u litre$^{-1}$); bilirubin 18 µmol litre$^{-1}$ (normal range 0–18 µmol litre$^{-1}$). Alkaline phosphatase was normal. A peripheral blood smear showed evidence of a microangiopathic haemolytic anaemia.

At this stage a diagnosis of HELLP syndrome was considered. In view of the rapid progression
of the disease process and the gestational age of the pregnancy, it was decided to proceed to Caesarean section. The anaesthetic team were informed. A central venous catheter was inserted via the right internal jugular approach and a urinary catheter was inserted to assess intravascular volume and monitor urinary output. Central venous pressure and urinary output were low; 3 mm Hg and 10 ml h⁻¹, respectively. Fresh frozen plasma 400 ml was given to increase plasma volume and urinary output. Urinary output increased to 30 ml in the next 1 h. A platelet transfusion (6 units) was also given immediately before surgery to increase the platelet count to more than 50 000 mm⁻³. Before the patient was transferred to theatre, an infusion of hydralazine was commenced, as is the practice in this unit, to reduce mean arterial pressure and thus avoid the risk of eclampsia or intracranial haemorrhage.

On arrival of the patient in the operating theatre, her mean arterial pressure was between 100 and 105 mm Hg. Anaesthesia was induced with thiopentone 5 mg kg⁻¹ and intubation was facilitated by administration of suxamethonium 1.5 mg kg⁻¹ i.v. Fentanyl 5 μg kg⁻¹ was given i.v. at induction to attenuate the hypertensive response to intubation. Atracurium 0.6 mg kg⁻¹ was used to provide neuromuscular block, and the patient’s lungs were ventilated with 50% nitrous oxide and 0.5% isoflurane in oxygen. Two healthy male infants were delivered, weighing 1730 g and 1570 g. The patient’s recovery was uneventful. One unit of fresh whole blood was administered during operation, followed by two further units on the first night after operation. The patient was monitored on a high dependency unit within the obstetric department for the first 24 h after operation. Monitoring included: central venous pressure, urinary output, non-invasive arterial pressure, ECG and oxygen saturation.

The infusion of hydralazine was reduced and stopped when arterial pressure was stable after 6 h. On the second day after operation platelet count was 112 000 mm⁻³. Liver enzyme concentrations returned to normal over the following week.

**DISCUSSION**

Pre-eclampsia is a condition with a spectrum of pathophysiological changes. At the most severe end of this spectrum a syndrome exists comprising haemolysis, increased concentrations of liver enzymes and decreased platelet count (HELLP syndrome). The clinical signs and symptoms of the syndrome include epigastric pain, upper abdominal tenderness, proteinuria, hypertension, jaundice and nausea and vomiting. Progression of the syndrome may lead to haematuria, oliguria, acute tubular necrosis, cortical necrosis and panhypopituitarism. Rarer complications include acute liver rupture and adult respiratory distress syndrome. A non-obstetric diagnosis such as acute cholecystitis, drug reaction or idopathic thrombocytopenia is often considered at presentation [2-4]. The diagnosis is suspected on clinical grounds and confirmed by laboratory investigations. Severe pre-eclampsia is diagnosed if arterial pressure exceeds 160/90 mm Hg or there is proteinuria > 5 g/24 h, oliguria < 400 ml/24 h, cerebral signs or pulmonary oedema [5]. If, in addition, there is evidence of haemolytic anaemia, thrombocytopenia and increased liver enzyme activity, HELLP syndrome should be considered [5]. The degree of thrombocytopenia correlates well with the degree of liver dysfunction [6]. Maternal mortality is high—Weinstein reported a mortality of 3.4% in one of his series [1].

Successful management is aimed at early diagnosis and stabilization, following which prompt delivery is desirable in the presence of the following: evidence of worsening pre-eclampsia such as increasing arterial pressure, cerebral symptoms and signs, worsening hepatic or renal function or severe thrombocytopenia; gestational age at or beyond 32-34 weeks; evidence of fetal distress; evidence of fetal maturity [6]. In the majority of cases, the condition tends to resolve soon after delivery. For obstetric reasons, Caesarean section rates are high: Thiagarajah and colleagues reported an incidence of 61.5% [6], whilst Weinstein reported 76% [1]. Many of these patients present for emergency Caesarean section with little or no warning to the anaesthetist and this may allow only a limited time for investigations.

Preoperative haematological investigations should include platelet count, white blood cell count, PCV, partial thromboplastin time, fibrinogen concentration, fibrin degradation products, peripheral blood smear, liver function tests and serum concentrations of creatinine, urea and uric acid [6]. A chest x-ray should be obtained to exclude early pulmonary oedema and ECG examination performed. Platelet transfusion has been recommended if the platelet count is less
than 50000 mm⁻³ in patients for Caesarean section and less than 20000 mm⁻³ in patients to be delivered vaginally [3]. A transfusion of fresh whole blood is recommended if the haemoglobin concentration is less than 10 g% [3]. A urinary catheter should be passed for hourly monitoring of urinary output. If the output is low and a diagnosis of acute renal failure is considered, measurement of urine concentrations of sodium and osmolality may be useful in confirming a diagnosis and directing treatment. A balloon-tipped pulmonary artery catheter or central venous catheter is indicated in critically ill patients for fluid management. This avoids hypotension at induction of anaesthesia, as many of these patients are relatively hypovolaemic. Measurement of maternal acid-base status is indicated in patients with clinical or radiological evidence of pulmonary complications or with a coagulation disturbance [3]. Maternal blood glucose concentration should be measured during operation, as severe hypoglycaemia has been reported in association with HELLP syndrome [7]. Many of these patients may be receiving therapy to control arterial pressure; i.v. magnesium sulphate or hydralazine have been used successfully for this purpose [6].

The choice of anaesthetic technique depends on the individual anaesthetist. The use of regional techniques may be limited by the coagulation disturbance and the urgency of the situation in the presence of fetal distress. Thiagarajah and colleagues reported successful use of extradural anaesthesia in a patient with HELLP syndrome, although the platelet count at the time of operation was 112000 mm⁻³ [6]. In view of the hepatic and renal involvement, anaesthetic drugs with minimal hepatic or renal metabolism should be chosen. Propofol would seem a logical choice for induction of anaesthesia as it has no active metabolites and a short half-life with rapid recovery. Suxamethonium is useful to ensure early good intubating conditions, but its half-life may be prolonged because of decreased serum cholinesterase concentrations as a result of hepatic dysfunction and pregnancy. Neuromuscular block may be maintained with atracurium, which is independent of renal excretion. The choice of volatile agent depends on the individual anaesthetist but, because of its low biotransformation, isoflurane seems a good choice. Intraoperative arterial pressure may be controlled with an infusion of hydralazine and isoflurane. For the first 12 h after operation, patients should be monitored in a high dependency unit where CVP, urinary output and haemodynamic state may be monitored.

REFERENCES