Reminder of important clinical lesson

Do not forget about HELLP!

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Summary

A 32-year-old female para 4 gravi 3, who was 21 weeks pregnant, presented to the emergency department (ED) with a 2-day history of abdominal pain, headache, blurred vision and vomiting. On arrival, she was agitated and confused with a blood pressure 162/106 mm Hg, pulse rate 107, respiratory rate 18, temperature 37 degrees Celsius, point of care blood glucose 6.2 and her Glasgow coma scale was 13/15 M6V4E3. Paramedics witnessed seizure-like activity lasting <1 min during transport. A diagnosis of eclampsia complicated by the HELLP syndrome (haemolysis, elevated liver enzymes, low platelets count) was made. She was commenced on magnesium and labetalol intravenously for blood pressure control. Initial blood test results were consistent with the HELLP syndrome. Recognition of the HELLP syndrome with prompt management of blood pressure and clotting abnormalities is essential in the ED setting. An aggressive multidisciplinary approach is a key to optimise the prognosis for mother and fetus.

BACKGROUND

HELLP (haemolysis, elevated liver enzymes, low platelets count) syndrome is a multisystem disease that is characterised by microangiopathic haemolytic anaemia, hepatic dysfunction and thrombocytopenia. It was first described by Weinstein in 1982.¹ It has a high maternal and perinatal morbidity and mortality. Its incidence is reported as 0.2%–0.6% of all pregnancies, and 10%–20% of women with co-morbid pre-eclampsia. HELLP usually begins during the third trimester, and usually in Caucasian women over the age of 25.² Prompt recognition of HELLP syndrome and timely initiation of therapy are vital to ensure the best outcome for mother and fetus.

CASE PRESENTATION

A 32-year-old female para 4 gravi 3, who was 21 weeks pregnant, presented to the emergency department (ED) with a 2-day history of abdominal pain, headache, blurred vision and vomiting. Her mother stated that earlier in the day she had an episode consistent with seizure-like activity. Medical and family history was unremarkable.

On arrival, she was agitated and confused with a blood pressure 162/106, pulse rate 107, respiratory rate 18, temperature 37 degrees Celsius, point of care blood glucose 6.2 and her Glasgow coma scale was 13/15 M6V4E3. Paramedics witnessed seizure-like activity lasting <1 min during transport. Primary survey revealed that her chest was clear, heart sounds normal and abdomen was soft and non-tender with a palpable uterus rising just below the umbilicus. She had no per vaginal bleeding. She was

icteric and had evidence of tongue biting. A urine dipstick revealed protein 4+.

An initial working diagnosis of eclampsia complicated by the HELLP syndrome was made. She was commenced on magnesium and labetalol intravenously for blood pressure control. Initial blood test results were consistent with the HELLP syndrome (table 1).

She was transfused two pools of platelets and transferred to an obstetric centre. She was treated conservatively for 24 h; unfortunately she miscarried the following day. Day 1 post spontaneous abortion, the labetalol and magnesium were discontinued.

DISCUSSION

HELLP syndrome is a multisystem disease that is characterised by microangiopathic haemolytic anaemia, hepatic dysfunction and thrombocytopenia. It was first described by Weinstein in 1982.¹ It has a high maternal and perinatal morbidity and mortality. Its incidence is reported as 0.2%–0.6% of all pregnancies, and 10%–20% of women with co-morbid pre-eclampsia. HELLP usually begins during the third trimester, and usually in Caucasian women over the age of 25.²

The aetiology and pathogenesis of HELLP syndrome remains unclear. Van Beek *et al* postulate that abnormal placentation results in placental ischaemia and the production of a circulating toxin that causes endothelial cell injury.³ The injury is believed to cause vascular constriction within multiple organ systems, activation of the coagulation system, increased capillary permeability and platelet

Table 1 Initial blood test results

Haemoglobin	Platelets	Urea	Creatinine	Albumin	Bilirubin	ALT	ALP	GGT	Calcium	Urate	PT	APTT
11.3	34	11.3	131	31	169	586	109	31	2.38	470	11.4	35.2

normal values: haemoglobin 13–18.5 g/dl, platelets 150–400 × 10⁹/l, urea 2.5–6.7 mmol/L, creatinine 62–106 mmol/l, albumin 35–50 g/l, bilirubin 3–17 µmol/l, ALT 3–35 IU/l, ALP 30–300 IU/l, GGT 0–30 IU/l, calcium 2.12–2.65 mmol/l, urate 200–400 µmol/l, PT 10.9–12.5 s, APTT 35–45 s.

ALT, alanine transaminase; ALP, alkaline phosphatase; APTT, activated partial thromboplastin time; GGT, γ-glutamyl transpeptidase; PT, prothrombin time.

activation with platelet consumption in the microvasculature, all resulting in hypertension, proteinuria, oedema and thrombocytopenia.

Prompt recognition of HELLP syndrome and timely initiation of therapy are vital to ensure the best outcome for mother and fetus. When the syndrome was first described, prompt delivery was recommended.⁴ Recent research suggests that morbidity and mortality do not increase when patients with HELLP are treated conservatively. Patients with HELLP syndrome may be eligible for conservative management if hypertension is controlled at less than 160/110 mm Hg, oliguria responds to fluid management and elevated liver function values are not associated with right upper quadrant or epigastric pain. One study found that pregnancy was prolonged by an average of 15 days when conservative management (i.e., bed rest, fluids and close observation) was used in patients who were at less than 32 weeks of gestation.⁴ Maternal morbidity was not increased. For infants, the prolongation of pregnancy translated into less time in the neonatal intensive care unit, a decreased incidence of necrotising enterocolitis and a decreased incidence of respiratory distress syndrome.⁴ Females treated conservatively should be managed in a tertiary care centre that has a neonatal intensive care unit and a perinatologist available for consultation. Patients with HELLP syndrome should be treated prophylactically with magnesium sulphate to prevent seizures, whether hypertension is present or not. A bolus of 4 to 6 g of magnesium sulphate as a 20 percent solution is given initially. This dose is followed by a maintenance infusion of 2 g per h. The infusion should be titrated to urine output and magnesium level. Patients should be observed for signs and symptoms of magnesium toxicity. If toxicity occurs, 10 to 20 ml of 10 percent calcium gluconate should be given intravenously.

Antihypertensive therapy should be initiated if blood pressure is consistently greater than 160/110 mm Hg despite the use of magnesium sulphate. This reduces the risk of maternal cerebral haemorrhage, placental abruption and seizure. The goal is to maintain diastolic blood pressure between 90 and 100 mm Hg. Patients who have had

HELLP syndrome should be counselled that they have a 19 to 27 per cent risk of developing the syndrome in subsequent pregnancies. They also have up to a 43 per cent risk of developing preeclampsia in another pregnancy.⁵

Recognition of the HELLP syndrome with prompt management of blood pressure and clotting abnormalities is essential in the ED setting. An aggressive multidisciplinary approach is a key to optimise the prognosis for mother and fetus. This case report highlights the need for vigilance regarding HELLP recognition in the ED setting.

Learning points

- ▶ HELLP syndrome is a multisystem disease that is characterised by microangiopathic haemolytic anaemia, hepatic dysfunction and thrombocytopenia.
- ▶ An aggressive multidisciplinary approach is a key to optimise the prognosis for mother and fetus.
- ▶ Recognition of the HELLP syndrome with prompt management of blood pressure and clotting abnormalities is essential in the ED setting.

Competing interests None.

Patient consent Obtained.

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