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Case Report

The Impact of Intracerebral Hemorrhage on Maternal Outcome in Hemolysis, Elevated Liver Enzymes and Low Platelet Count Syndrome

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Abstract

Hemolysis, Elevated Liver Enzymes and Low Platelet Count (HELLP) syndrome can be complicated by intracranial hemorrhage. The incidence is 0.6-6.1 per 100.000 women delivering and is associated with a high mortality rate and long-term morbidity.

We present the case of a 29-year-old woman with pregnancy-induced hypertension who developed HELLP syndrome and uncontrollable hypertension at the beginning of induction of labor. A cesarean section was performed. Shortly after, she had an eclamptic seizure. CT showed multiple intracranial hemorrhages. The patient was transferred to a tertiary neurological center, after which she recovered well. The only complaint she had was a mild aphasia. After six months, neurological testing showed no signs of aphasia anymore and she achieved her normal level of daily functioning.

Intracranial hemorrhage is a severe complication in HELLP syndrome. Early awareness and rapid management can improve outcome.

Keywords: Hemolysis; Elevated liver enzymes and low platelet count syndrome; HELLP; Intracranial hemorrhage; Eclampsia; Hypertension; Pregnancy

Abbreviations

CT: Computerized Tomography; DIC: Disseminated Vascular Coagulation; GCS: Glasgow Coma Scale; HELLP: Hemolysis, Elevated Liver Enzymes and Low Platelet Count; ICH: Intracerebral Hemorrhage; ICU: Intensive Care Unit; PACU: Post Anesthesia Care Unit

Introduction

HELLP syndrome is characterized by hemolysis, elevated liver enzymes and a low platelet count and its incidence is approximately 0.1-0.8% of all pregnancies and 10-20% of pregnant women with severe preeclampsia of eclampsia [1]. HELLP syndrome is associated with systemic complications such as respiratory distress, acute renal kidney injury, liver rupture, Disseminated Vascular Coagulation (DIC) and Intracerebral Hemorrhage (ICH). A case is reported in which a patient developed multiple intracerebral hemorrhages resulting from HELLP. The impact of HELLP associated intracerebral hemorrhages on maternal outcome is discussed.

Case Presentation

A 29-year-old primigravida, without prior medical history and no use of medication, was admitted to the department of Obstetrics and Gynaecology of our hospital at 37 2/7 weeks of gestation for induction of labor. During her pregnancy, there had been two admissions to the hospital at 30 and 31 weeks of gestation because of gestational hypertension, without symptoms of preeclampsia, normal blood tests and absence of proteinuria. At 31 weeks labetalol 200 mg orally 2 times a day was administered to control her blood pressure with good result. Ultrasonography of the fetus showed a small gestational age (<10^{th} percentile) without any other abnormalities. Induction of labor was planned between 37 and 38 weeks and the patient was admitted to the hospital at 37 2/7 weeks of gestation in order to induce labor the next day. During the night the patient suddenly complained of headache, nausea and vomiting. The blood pressure was 207/115 mmHg and patellar reflexes were normal. Continuous fetal cardiac monitoring showed a good fetal condition. Intravenous labetalol was started to stabilize blood pressure. Laboratory tests showed HELLP syndrome. Despite increased doses of labetalol (up to 80mg/h) hypertension persisted (190-220/110-125 mmHg). Subsequently, intravenous s magnesium sulphate (1gr/h) was started. In the meantime, the patient started having spontaneous contractions and showed 5cm dilatation. After amniotomy, ultrasonography revealed an unexpected breech presentation. After counseling, it was decided to continue a vaginal delivery, because of a fast progression of the dilatation and unstable blood pressure at that moment. After initial decrease, the blood pressure increased again and methyldopa 250mg orally was started. Repeated blood test showed signs of progressive HELLP syndrome. Fetal cardiac monitoring was normal. As the patient increasingly complained about headache and vomiting and vaginal delivery was progressing slowly, the decision was made to perform a caesarean section under spinal anesthesia. Shortly before surgery, the blood test showed a platelet count of 74x10^9/L, sufficient to perform spinal anesthesia. While performing the time out procedure, the patient was unable to remember her date of birth. However, spinal anesthesia and cesarean section went uncomplicated. A healthy girl was born

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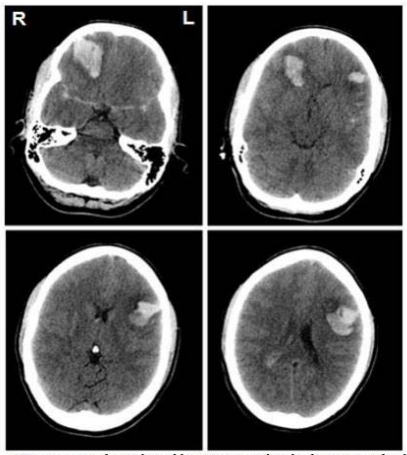

CT imaging: subarachnoid hematoma and multiple intracerebral hemorrhages in the frontal lobe, right and left hemisphere with expansion to the right lateral ventricle.

Figure 1: Subarachnoid hematoma and multiple intracerebral hemorrhages, amongst others in the frontal lobe and right and left hemisphere with expansion to the right lateral ventricle.

(weight 2095gr, 3rd percentile) with APGAR scores of 7 and 9 (after 1 and 5 min) respectively. During surgery, the blood pressure was regulated with additional inhalational nitrous oxide and kept below 140 systolic. Post-operatively the patient went to the Post Anesthesia Care Unit (PACU). Ten minutes after arrival she suddenly developed a generalized tonic-clonic insult for which midazolam 1mg and an additional bolus of magnesium sulphate was given, where after her seizure stopped, the Glasgow Coma Scale (GCS) was 1-5-1. She was transferred to Intensive Care Unit (ICU) for further evaluation. The GCS increased (3-6-3) and Computerized Tomography (CT) of the cerebrum showed subarachnoid hematoma and multiple intracerebral hemorrhages, amongst others in the frontal lobe and right and left hemisphere with expansion to the right lateral ventricle (Figure 1). As neurologic monitoring showed, no deterioration observation was performed only. Treatment of HELLP consisted of transfusion of platelets and fresh frozen plasma and levetiracetam was administered to prevent new seizures. Shortly after, she was transferred to a neurological ICU in a tertiary care center. Repeat CT showed minor edema and no increase of intracranial hemorrhage. The HELLP syndrome slowly recovered and antihypertensive medication could be decreased and finally stopped after 18 days. The main neurological symptom she had was aphasia, but she did not suffer from sensory or motor symptoms. After 4 days on the neurological ICU the patient was transferred to the gynaecology and obstetrics ward; and after another 6 days she was transferred to the neurology ward for further rehabilitation.

Fourteen days after surgery she went to a rehabilitation center and her symptoms consisted of global aphasia and mild frontal disinhibition. The patient recovered very well: nine weeks postpartum the only symptom that was left was a mild aphasia, without any other neurological deficits and she was able to take care of her baby daughter. She was discharged from the rehabilitation center and rehabilitation was continued in an ambulant setting. A control MRI of the cerebrum nine weeks postpartum showed a decrease in hematomas without underlying pathology. After six months, neurological testing showed no signs of aphasia anymore and she achieved her normal level of daily functioning.

Discussion

Intracranial hemorrhage is a rare complication of HELLP syndrome and its incidence is 0.6-6.1 per 100.000 women delivering. However, this complication is associated with a high mortality rate of around 20-25% and is the most common cause of death in patients with HELLP syndrome (45%) [2-5]. Furthermore, intracranial hemorrhage in HELLP syndrome is associated with long-term morbidity.

Early awareness of the possibility of intracranial hemorrhage as result of HELLP syndrome is the key in decision-making strategies. However, before HELLP syndrome may develop, eclampsia is also associated with brain lesions such as hemorrhage, infarction or cerebral edema [6]. A recent report in which 39 patients were admitted to the ICU for eclampsia, 19 patients had brain lesions such as ischemic (10), intraparenchymal (3) and subarachnoid hemorrhage (1) [6]. Nine patients suffered cerebral edema with neurological symptoms. Less than one third of all patients had HELLP syndrome and of these patients three patients died directly to intracranial hemorrhage.

Independent risk factors for intracranial hemorrhage in pregnant women are advanced maternal age, African-American origin, coagulopathy, smoking, preeclampsia/eclampsia and preexisting or gestational hypertension [4]. Another case series of 27 patients showed that severe systolic hypertension (>160mmHg) increased the risk of developing intracranial hemorrhage in patients with severe preeclampsia and eclampsia [7]. Moreover, cerebral autoregulation is also impaired in patients with preeclampsia, which also contributes to the vulnerability to develop intracranial hemorrhage [8]. Our patient had only two risk factors: hypertension and coagulopathy.

Aggressive treatment of hypertension in pregnancy is of major importance to decrease the risk of complications. However, adequate regulation of blood pressure may be cumbersome as shown our patient who suffered severe hypertension for almost six hours despite treatment with labetalol, methyldopa and magnesium sulphate. The rapid decrease in platelet count and subsequent coagulopathy in combination with severe hypertension might have contributed to the development of intracranial hemorrhage. The strategy of delivery should be decided case by case. However, vaginal delivery might be of higher risk as intracranial pressure is increased during the last phase of delivery and cesarean section might be the best option. However, coagulopathy is a contraindication for spinal anesthesia (platelet count $<50x10^9$/L) and when general anesthesia is performed, a stress free intubation is essential to prevent more hypertension and additional risk of intracranial hemorrhage. A multidisciplinary approach and a protocol driven decision should be made how to approach pregnant patients with HELLP syndrome and to prevent intracranial hemorrhage.

This case report shows how sudden the onset of HELLP syndrome can be in a previously stable gestational hypertension. In only a few hours, she developed HELLP syndrome, uncontrollable blood pressures, eclamptic seizures and multiple intracranial hemorrhages. Despite the severe brain lesions on multiple locations, the patient's neurological outcome was relatively well. Awareness of risk factors concerning intracranial hemorrhage, aggressive regulation of blood

pressure and a multidisciplinary approach is essential to prevent HELLP syndrome, which is associated intracranial hemorrhage. In case of any neurological symptoms early imaging of the brain and close neurological monitoring should be performed.

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