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CLINICAL INFORMATION

Anesthesia in pregnant women with HELLP syndrome: case report

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KEYWORDS
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Abstract
Background and objectives: HELLP syndrome, characterized by hemolysis, high levels of liver enzyme, and low platelet count, is an advanced clinical stage of pre-eclampsia, progressing to high maternal (24%) and perinatal (up 40%) mortality, despite childbirth care in a timely manner. The goal is to describe the anesthetic management of a case with indication to emergency cesarean.
Case report: Female patient, 36 years old, gestational age of 24 weeks, with hypertensive crisis (BP 180/100 mmHg) and severe headache, was admitted to the operating room for a cesarean section after diagnosis of HELLP syndrome. Indicated for general anesthesia, we opted for total intravenous with intubation after rapid sequence induction with propofol and remifentanil in continuous target-controlled infusion, and rocuronium at a dose of 1.2 mg/kg. Maintenance was achieved with propofol and remifentanil. The surgical procedure was uneventful, the child was born with APGAR 1/5 and transferred to the NICU. At the end of surgery, the patient was extubated in the operating room and taken to the ICU. The postoperative period was uneventful with no changes worthy of note and the patient was discharged on the sixth postoperative day.
Conclusion: When general anesthesia is the choice in parturient with HELLP syndrome, tracheal intubation with rapid sequence induction due to possible difficult airway, as well as the use of drugs to control the hemodynamic response can minimize the complications associated with the procedure, as occurred in this case.

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PALAVRAS-CHAVE
Síndrome HELLP;
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Propofol;
Remifentanil

Anestesia para gestante com síndrome HELLP: relato de caso

Resumo
Justificativa e objetivos: A síndrome HELLP, caracterizada por hemólise, elevação dos níveis de enzimas hepáticas e plaquetopenia, representa estágio clínico avançado da pré-eclâmpsia, cursando com elevada mortalidade materna (24%) e perinatal (até 40%), apesar da assistência ao parto de forma oportuna. O objetivo é descrever o manejo anestésico de um caso com indicação de cesariana de emergência.
Relato de caso: Paciente do sexo feminino, 36 anos, idade gestacional 24 semanas, com crise hipertensiva (PA 180/100 mmHg) e cefaleia intensa, é encaminhada ao Centro Cirúrgico para operação cesariana após diagnóstico de síndrome HELLP. Indicada anestesia geral, optou-se por venosa total com intubação após indução sequencial rápida, com propofol e remifentanil em infusão contínua alvo-controlada e rocurônio na dose de 1,2 mg/kg. A manutenção foi obtida com propofol e remifentanil. O procedimento cirúrgico transcorreu sem anormalidades, a criança nasceu com APGAR 1/5 e foi encaminhada à UTI Neonatal. Ao final da cirurgia, procedeu-se à extubação na sala cirúrgica e a paciente foi encaminhada ao CTI. O pós-operatório decorreu sem alterações dignas de nota e a paciente teve alta no sexto dia pós-operatório.
Conclusão: Quando se opta pela anestesia geral em parturiente com síndrome HELLP, a intubação traqueal com técnica de indução sequencial rápida em função de possível via aérea difícil, bem como o emprego de drogas que controlem a resposta hemodinâmica, podem minimizar as complicações associadas ao procedimento, como ocorreu no presente caso.

Introduction

Hypertension induced by pregnancy has various clinical forms, sometimes presenting with slight increases in blood pressure and sometimes as a severe enough disease to involve various organs or systems.[1] Preeclampsia is a complex multisystem disorder of unknown etiology, characterized by the combined development of hypertension and proteinuria (>300 mg within 24 h) after the first 20 weeks of pregnancy.[1,2] Edema is no longer a diagnostic criterion. Preeclampsia is a clinical diagnosis. Risk factors include obesity, nulliparity, and advanced maternal age. Preeclampsia has an incidence ranging from 1.5% to 3.8% in all pregnancies in developed countries, while in Brazil it may be as high as 7.5%.[3] The Hemolysis Elevated Liver enzymes Low Platelets (HELLP) syndrome is an advanced stage of preeclampsia. Regarding platelet count, HELLP syndrome is divided into three classes[3]: class I if platelets count less than 50,000 mm^{-3}; class II if between 50,000 and 100,000 mm^{-3}; and class III if greater than 100,000 mm^{-3}. HELLP syndrome etiology is not fully known. Its clinical manifestations result from unknown insult to platelet activation and microvascular endothelial damage. Hemolysis, defined by the presence of microangiopathic hemolytic anemia, is the most important disorder. The following diagnostic criteria are suggested: (1) hemolysis, defined by abnormal peripheral bleeding and increased bilirubin levels (1.2 mg dL^{-1} or more); (2) elevated liver enzymes, defined by glutamic–oxaloacetic transaminase (AST) of 70 U L^{-1} or more and lactate dehydrogenase (LDH) above 600 U L^{-1}; (3) low platelet count (less than 100,000 mm^{-3}).[4] HELLP syndrome may affect 4–12% of patients with severe preeclampsia and may contribute to high maternal (24%) and perinatal (up to 40%) mortality, despite the delivery care in a timely manner.[3,5] Given the importance of this involvement during pregnancy and the frequency in anesthesia, appropriate management of pregnant patients with severe preeclampsia is extremely important.

Case report

Female patient, 36 years old, 24 weeks gestational age, with a history of malaise and headache for a week, presenting a hypertensive crisis (BP 180/100 mmHg) and intensive headache at hospital admission. She had no seizures at home or in the emergency room. Test results on admission are shown in Table 1. After expert evaluation, the patient was taken to the operating room for an emergency cesarean diagnosed with HELLP syndrome. She arrives at the operating room awake, lucid and oriented, with hemodynamic stability, receiving magnesium sulfate scheme as obstetrical management. Given the case severity and the contraindications to neuraxial blockade that will be discussed later, total intravenous anesthesia was indicated. The patient was positioned on the surgical table and monitored as routine, with multiparameter monitors (non-invasive blood pressure, cardioscopy, pulse oximetry). Peripheral venous access was performed with Jelco number 18 G and fluid replacement started with heated Ringer's lactate solution. Anesthesia induction was proposed according to the need for tracheal intubation by the rapid sequence technique, with oxygen under FiO_2 100%, propofol target-controlled continuous infusion (4 ng mL^{-1}), remifentanil target-controlled continuous infusion (3 ng mL^{-1}), and rocuronium (1.2 mg kg^{-1}). Tracheal intubation was uneventfully performed. Anesthetic maintenance was performed with target-controlled infusion of

Table 1 Laboratory tests at hospital admission.

Hemoglobin (mg dL^{-1})	13.2	Sodium (mEq L^{-1})	142	Transglutaminase oxaloacetic (TGO) (U L^{-1})	709
Hematocrit (%)	38	Potassium (mEq L^{-1})	4.4	Glutamic pyruvic transaminase (GPT) (U L^{-1})	391
Platelets (mm^3)	52.000	Magnesium (mEq L^{-1})	2.0	Total Bilirubin (mg dL^{-1})	4.8
Prothrombin time (s)	11.5	Urea (mg/%)	23	Direct bilirubin (mg dL^{-1})	1.1
Activated partial thromboplastin time (s)		Creatinine (mg/%)	0.7	Indirect bilirubin (mg dL^{-1})	3.8
INR	1.0	Alkaline phosphatases (U L^{-1})	184	Lactate dehydrogenase (LDH) (U L^{-1})	3799
		Gamma-GT (U L^{-1})	31		

propofol (3 ng mL^{-1}) and remifentanil (3 ng mL^{-1}). Mechanical ventilation with cycled volume, tidal volume = 8 mL kg^{-1}, and FiO_2 = 50% in air. The surgical procedure was uneventful. The baby was born alive, APGAR 1 (1st min)/5 (10th min), and was sent to the neonatal intensive care unit (NICU) for pediatric rigorous evaluation. At the end of surgery, the patient was extubated still in the operating room, remaining stable and admitted to the intensive care unit (ICU). The patient was discharged on the 6th postoperative day without complications. The child remained in the NICU and was discharged on the 26th day with good health and development.

Discussion

As a severe form of preeclampsia, HELLP syndrome originates from abnormal placental development, followed by the production of factors that promote endothelial injury through the activation of platelets and/or vasoconstrictors.[6,7] Endothelial injury of the hepatic vessels, followed by platelet activation, aggregation and consumption, resulting in hepatocyte ischemia and death, is the main hypothesis to explain the characteristic laboratory findings of HELLP syndrome.[6] This is a complication in approximately 6–8% of pregnancies and together with eclampsia it is responsible for significant morbidity and mortality in obstetric patients. Severe preeclampsia involves organ damage. It is characterized by blood pressure values greater than or equal to 160/110 mmHg at rest, severe proteinuria and oliguria (<400 mL 24 h^{-1}), changes in vision, headache, and other brain changes, epigastric pain, signs of pulmonary edema, cyanosis, and HELLP syndrome.[1] Patients with HELLP are susceptible to high incidence of stroke, heart disease, placental rupture, need for blood transfusion, pleural effusion, and infections. Many HELLP cases occur in the preterm period, but 20% may occur in the post-term period with higher incidence of acute pulmonary edema.[4] Studies reported that of 309 patients who developed HELLP syndrome, 69% had it before and 31% after delivery; when it occurs during the postpartum period, its onset occurs mainly 24–48 h after birth of the fetus, although it is described that its onset may occur within hours or up to 6 days after birth.[1,4] Case reports in the international literature confirm the occurrence of HELLP syndrome up to 8 h after the procedure, with the same nosological features already reported.[8]

The decision on anesthesia for patients with preeclampsia depends mainly on an overall assessment of the benefits and risks that the anesthesia effect can cause both to mothers and babies. When preeclampsia is manifested with HELLP syndrome, coagulopathy, and severe dysfunction of multiple organs and cesarean delivery is indicated, general anesthesia may be a safer method than the neuraxial block, as long as a successful airway management is provided.[9–11] Severe preeclampsia with HELLP syndrome should mean a change in anesthetic planning. The presence of coagulopathy with thrombocytopenia predisposes to an increase in the risk of epidural hematoma with neuraxial anesthesia techniques. Guidelines recommend a platelet count greater than 100,000 mm^{-3} to minimize this risk, although there is no statistical data in the literature on neuraxial blockade complications in patients with HELLP syndrome and platelet count less than the cited value.[10] A retrospective study[3] evaluated a total of 102 cases, including seven patients with HELLP syndrome after delivery and 95 patients with HELLP syndrome before delivery. In the latter, 37 patients were submitted to general anesthesia, 53 underwent combined neuraxial blockade (spinal-epidural), and 12 underwent spinal blockade. In patients undergoing combined neuraxial blockade, preoperative platelet count remained about 113,000, with no difference to spinal block (95,000) and general anesthesia (88,000). Two patients underwent combined blockade, even with platelet count less than 50,000 mm^{-3}. There were no cases of epidural hematoma, demonstrating safety and feasibility in the use of neuraxial blockade in selected cases of HELLP syndrome.[12] Anesthesia for HELLP syndrome in a patient with adequate platelet count and absence of coagulopathy is controversial as, in spite of the foregoing, there are literature reports of post-anesthetic puncture spinal hematomas; thus, its use should be avoided and complications readily recognized.[13]

When general anesthesia is considered, the technique of rapid sequence intubation with likely difficult airway assessment and drugs that control the hemodynamic response can minimize complications associated with the procedure. These include esmolol, fentanyl, remifentanil, alfentanil,

and lidocaine.[10] Remifentanil is often used to promote short-term analgesia with cardiovascular stability in high-risk patients.[14] In the case presented here, remifentanil was the option used for induction and maintenance of anesthesia. A recent study reported three cases of good results for the choice of general anesthesia with sevoflurane instead of epidural spinal block for cesarean section in patients with HELLP syndrome, due to severe thrombocytopenia. None of the complications reported in the literature (renal failure, pulmonary edema, cerebral hemorrhage, and hepatic rupture) was observed in the three postoperative days in the reported cases.[15]

Conclusion

Recent evidence about the anesthetic management of patients with HELLP syndrome who should undergo cesarean section is not clear regarding the best approach indication. However, given the range of possible complications, it appears that the general anesthesia technique with airway control (intubation) after rapid sequence induction is a good choice. The available pharmacological arsenal should be used with discretion, with attention to drugs that bring greater stability during surgery. However, further studies are needed to indicate, based on evidence, the best technique to be used in cases of severe preeclampsia and HEELP syndrome.

Conflicts of interest

The authors declare no conflicts of interest.

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