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Case Report

Anesthesia Management in Patient with Plasminogen Deficiency for Cesarean Section (C/S): A Case Report

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ABSTRACT

Plasminogen (PLG) is a serum protein, a precursor of plasmin, in addition to its functions in the coagulation pathway, it also plays an important role in wound healing. PLG deficiency is a rare condition and has been associated with pseudomembranous (ligneous) lesions on mucous membranes, for example: ligneous gingivitis, endometritis and mostly conjunctivitis in the literature. In our case, the patient was a 30-years-old pregnant woman who was diagnosed with PLG deficiency and was at 39+4 week of pregnancy. We chose the neuraxial anesthesia (NA) to avoid possible airway obstruction due to difficult intubation and mucosal damage of upper airways. So, we administered single shot atraumatic spinal anesthesia (SA) for emergency cesarean section (C/S). Herein, preoperative optimization, a good preliminary assessment and selection of the type of anesthesia are important in terms of preventing complications in these patients.

Plasminogen is a serum protein, a precursor of plasmin, a serine protease, that is involved in fibrinolysis, wound healing and inflammation modulation on the wound site. PLG is primarily produced by the liver, but also is produced by the brain, kidney, heart, lungs, intestines, uterus, spleen and thymus in smaller amounts [1].

PLG is synthesized in the liver and converted to plasmin by tissue PLG activator or urokinase type PLG activator. Although plasmin is involved in the coagulation pathway, it degrades fibrin and glycoproteins and plays an important role in wound healing by stimulating the release of transforming growth factor beta (TGF-β) [2].

PLG deficiency presents itself as an inability of organism for proper wound healing and therefore forming of thick fibrinoid pseudomembranes ('woody'-ligneous), composed of epithelial debris, fibrin, hyaline - like substance on conjunctiva, ears, mouth, respiratory, gastrointestinal and urogenital mucous membranes. PLG deficiency can be quantitative (hypoplasminogenemia) or functional (dysplanminogenemia); inherited (autosomal recessive, PLG gene mutations) or acquired [1-3]. Severely impaired skin wound healing has been reported in a few patients with PLG deficiency following most surgical procedures [4].

Condition has no definitive treatment currently, but symptoms can be limited by PLG-containing eye drops, fresh frozen plasma (FFP), immunosupressive agents, sex hormones. Recently the Food and Drug Administration (FDA) approved Ryplazim® in 2021, plasma-derived human PLG indicated for the treatment of patients with PLG deficiency type 1.

Although the incidence is not known, it is estimated at 1.6 people per 1.000.000 in the population. There are no reports regarding neuraxial anesthesia in these patients.

In this case, we aimed to present our perioperative anesthetic management and NA experience in the C/S of a patient with PLG deficiency, performed in December 2021.

The authors declare no conflicts of interest.

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Case Report

A 30-years-old female (81 kg, 165 cm) who had a history of infertility had no other known systemic disease. Local ethics committee approval and written consent of the patient were obtained. PLG deficiency was diagnosed after fibrinous pseudomembranes were spotted in the uterine cavity during hysteroscopy, later diagnosed as ligneous endometritis. PLG activity was 43 % (normal range is 75 to 140 %) and ligneous endometritis was treated with intrauterine tissue plasminogen activator (tPA) administration, and intravenous (i.v.) administration of FFP co-administration as a source of plasmin. Diagnosis of PLG deficiency was made by the hematology department based on PLG activity, and genetic analysis is being carried on at the moment. There were no other symptoms related to PLG deficiency. The pregnant woman who was at 39+4 week, had no pathological findings detected in cardiopulmonary evaluation, preoperative laboratory tests and ECG (electrocardiogram). The patient, who was a smoker, was evaluated as American Society of Anesthesia (ASA) II-E and had a Mallampati score of IV.

Normal vaginal delivery was planned at first, but the patient had labor arrest and due to fetal meconium aspiration suspicion, emergency C/S was planned. Approximately 10 ml/kg of FFP (3 Units) and 1000 ml of balanced crystalloid solution were pre administered during transport and preparation period to increase the PLG activity levels and to replenish intravascular volume, lost during fasting. Patient was admitted to operation room immediately because of amniotic fluid discharge. SA was planned for the patient concerns in regards to the frequency of difficult airway risk in pregnant women, the patient's consumption of solid food two hours before the surgery, and the reported cases of airway obstruction due to mucosal damage in patients with PLG deficiency.

After standard monitorization (ECG, HR, NIBP, SpO2), heart rate (HR) was 120/min and blood pressure (BP) was 143/72 mmHg, other vital parameters were within normal range. Antibiotic prophylaxis was administered and then patient received a single shot SA in a sitting position using a midline approach, using 26G Quincke needle, and 3 ml of 0.5 % hyperbaric bupivacaine® was injected intrathecally through L4-L5 intervertebral space without any complication. Sympathetic sensory and motor block were tested. Sensory block at T4 level was observed with pin prick test. We also made general anesthesia preparation in case of insufficient sensory block, and a videolaryngoscope was ready to prevent possible difficult intubation. The patient was positioned to prevent aortocaval compression and nasal oxygen was administered.

Surgery started after aseptic conditions were established and the surgical area was draped. The newborn was born 7 minutes after intrathecal local anesthesia and she had an Activity-Pulse-Grimace-Appearance-Respiration (APGAR) score 9/10, birth weight was 3800 grams. Amniotic fluid was stained with meconium. After clamps were placed on the umbilical cord, 100 mcg of carbetocin® were administered to the mother. Additionally, 15 ml/kg FFP were administered within 24 postoperative hours divided into 4 doses to sustain PLG levels.

Patient was observed for any complication until discharge from hospital. Sensory and motor block resolved within 3 hours almost completely, except mild numbness and weakness on ankles and feet bilaterally. Complete resolution took 6 hours, and then she was mobilized. The patient's vital signs were stable, no complications developed within the perioperative period and the patient was discharged from hospital in 48 hours succeeding the operation. Newborn was followed up in the neonatal intensive care unit (ICU) for 48 hours and discharged with no complications.

Discussion

Understanding the physiological changes that occur during pregnancy is essential when planning C/S anesthesia. Maternal hemodynamic changes occur during pregnancy in which cardiac output (CO) rises, HR and stroke volume (SV) increase. BP drops slowly with a 20% reduction in systemic vascular resistance (SVR) in mid-pregnancy [5]. Hypotension due to aortocaval compression in pregnant women becomes more evident with the addition of general anesthesia or NA.

Neuraxial anesthesia provides advantages such as the mother being awake at the birth of her child, avoiding the risks of general anesthesia and positive pressure ventilation. However, NA may cause hypotension by reducing preload and SVR. Increased dilatation of epidural vessels during pregnancy increases the likelihood of inadvertent vein insertion can result in spinal hematoma [5]. Secondary spinal cord infections must be taken into consideration too. On the other hand, pulmonary aspiration of gastric contents and failed endotracheal intubation is one of the major causes of maternal mortality and morbidity during general anesthesia. Considering all factors, the choice of anesthesia should be made according to the accompanying comorbidities of the patient.

While our patient was receiving infertility treatment, she was diagnosed with PLG deficiency and got pregnant as a result of the treatment. As it is known, pregnant women are in the risk group for difficult intubation, so airway obstruction may occur after a possible traumatic intubation in a pregnant woman with PLG deficiency. These patients may be complicated by laryngeal, tracheal, and bronchial obstructions by viscous fibrin-rich secretions and ligneous masses [6]. Considering these complications and the patient's safety, we decided to apply atraumatic SA, but we did preparation for difficult airway management, including video laryngoscope. Decision on primary modality of

anesthesia was the most challenging because of the emergency of C/S, pathophysiology of the PLG deficiency, and lack of any information about NA in similar cases.

PLG deficiency is a rare condition that can progress with infertility. In the literature, we could not find any article about C/S anesthesia in PLG deficiency and therefore, we are presenting our successful NA application in pregnant women with PLG deficiency.

Conclusion

In conclusion, thorough medical history, physical examination, knowing physiological changes in pregnancy and pathophysiology of the rare disease is very important in the choice of anesthesia method. Keeping everything above in mind, the most effective and safe anesthesia method should be preferred, together with the multidisciplinary team.

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