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Successful Anesthetic Management in Pregnant Lady with Eosinophilic Granulomatosis with Polyangiitis

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Abstract Vasculitis syndromes rarely co-exist with pregnancy; for example, Eosinophilic Granulomatosis with Polyangiitis (EGPA) (formerly known as Churg-Struss Syndrome) is even rarer. Vasculitis syndromes often have a poor prognosis, making diagnosis very problematic, and generally, we do not have much literature about the anesthetic management of these rare types of vasculitis. Our study reports a case scenario of a 38- year-old woman who is pregnant and has been diagnosed with EGPA. She completed a successful pregnancy and was subject to emergency category II cesarean section delivery under regional anesthesia at University College Hospital Galway-Ireland. The patient coped well during the pregnancy apart from the reoccurrence of an asthma attack and allergic rhinitis at mid-pregnancy, which responded to an upsurge in steroid dosage. Clinical effects and choice the type of anesthesia and it's relation to vasculitis are discussed.

Keywords: *Eosinophilic Granulomatosis with polyangiitis, vasculitis syndrome, anesthesia, pregnancy, diagnosis*

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1. Introduction

A necrotizing small-vessel vasculitis with extravascular granulomas and eosinophilic infiltrates is known as Eosinophilic Granulomatosis with Polyangiitis (EGPA) [1,2]. EGPA generally appears between the ages of 7 and 74, with a mean onset age of 38 to 54. EGPA has been reported in children as young as four years old, according to a recent study of the illness in the paediatric population [3,4]. The annual incidence is predicted to be between 0.11 and 2.66 new cases per million persons, with a prevalence of 10.7 to 14 cases per million adults. EGPA has not clearly established ethnic predisposition or gender preponderance [4]. EGPA is a rare form of necrotizing vasculitis affecting both medium and small systemic blood vessels. Polyarteritis Nodosa (PAN), Granulomatosis with Polyangiitis (GPA) and Microscopic Polyangiitis (MPA) are some of the other subtypes observed in the wide category [5]. The co-occurrence of asthma, peripheral eosinophilia and rhinosinusitis distinguishes EGPA from the other disorders in the group respiratory tract involvement, such as asthma, allergic rhinitis and pulmonary infiltrates, are the most prevalent clinical symptoms [6].

EGPA pathogenesis remains idiopathic since factors that trigger EGPA development are yet to be identified. On a cellular level, EGPA is characterized by a robust Th2-type immune response. Th2-linked cytokines, IL-13, IL-5, and IL-4, may trigger severe eosinophilia in EGPA. Conversely, migration of Eos to inflammatory sites is likely controlled by eotaxin-3 [7,8].

Based on EGPA, there is no specific test to confirm the disorder since signs and symptoms are identical to those of other diseases; hence can be problematic to diagnose [9]. The early indicators of the disease include asthma and sinusitis, which are often common; therefore, diagnosis may not be made until the inflammation has resulted in serious harm to organs and nerves [9]. If treatment to be postponed due to a late diagnosis, it may result in heart failure, leading to death. Thus, a quick diagnosis and aggressive medication are required to treat EGPA patients effectively.

People with EGPA present hypersensitivity to the airway, cholinesterase deficiency, and multiple organ dysfunctions. Hence, the anesthetic management of EGPA patients can be problematic and challenging. There are only a few studies on conducting anesthesia for EGPA patients. Hence, this study reports on the anesthetic management of a EGPA patient as well as reviews the available medical literature on the condition.

2. Case Description

A 38-year-old female, gravida 1 para 0, with a long history of Eosinophilic Asthma, chronic rhinosinusitis,

and atopy on the background of EGPA, was admitted at 38 weeks of pregnancy with spontaneous rupture of membrane [SROM] so urgent category II cesarean section was performed, under spinal anesthesia.

The patient was on maintenance prednisolone 10mg, Fluticasone/Vilanterol inhaler, Tiotropium bromide inhaler, Pulmicort, and Flixonase nasal drops. Her pregnancy course was uneventful apart from a few episodes of shortness of breath, green nasal discharge, and postnasal drip at 26 weeks of pregnancy. She responded well to increasing the maintenance dose of prednisolone to 20 mg for 5 days.

All the routine preoperative investigations, including routine blood test, blood biochemistry, 12 leads ECG, were within the standard limits. During the preoperative physical examination, sputum, mild cough, and rhinorrhea were detected. The report from chest X-ray performed 2 days prior to hospital admission revealed minimal patchy opacity in the left lower zone of the lung. In addition, the report from pulmonary function tests was within the required limits with a FVC of 80%, a FEV_1/FVC of 94%, and FEV_1 of 96% of the predicted values.

The patient was prepared, and admitted to the operating theatre. Her informed consent for the recommended spinal neuraxial anesthesia was obtained. The 8-hours fasting state was completed.

Routine basic monitoring included heart rate 80 beats/min, electrocardiogram, noninvasive blood pressure [BP] 130/65 mmHg, and pulse oximetry with SpO2 at 95% in room air were conducted and these baseline parameters were recorded in anesthesia chart. An intravenous [IV] access was secured with a 16 G cannula, and our patient was preloaded with 500 ml of CSL.

Under aseptic technique, a spinal block was performed at L2/L3 space in sitting position with 25-gauge Quincke needle, with an effective first attempt to reach the subarachnoid space and amount of 12 mg of hyperbaric bupivacaine, 20 mcg fentanyl, and 100 mcg morphine was injected into the subarachnoid space after confirmation of a clear and free backflow of cerebrospinal fluid [CSF]. Five minutes after lying the patient flat, the sensory level of the block was found to be at T4. There were no difficulties or complications over the course of the procedure.

The intraoperative period was straightforward with hemodynamic stability with minimal vasoactive cardiovascular support that stopped by the end of surgery. A total amount of 1.3 L of intravenous CSL was given. The 15-degree left tilt was performed to improve venous preload and avoid pressure over IVC and Aorta. A stress dose of 100 mg hydrocortisone and anti-emetic prophylaxis by ondansetron 4mg were given before skin incision. Cesarean section was performed uneventfully, and four minutes after skin incision, a full-term baby was born with an Apgar score of 8/10 and 9/10 at 1 and 5 minutes, respectively. The weight of the baby was 2800 g. There were 700 ml of intraoperative blood loss with no complications. At the end of C-section, analgesia was supplemented with paracetamol 1g rectally.

The postoperative course was smooth and straightforward, uneventful recovery room stayed for 45 minutes, and spinal block wears off 6 hours postoperatively. Postoperative pain relive, including regular paracetamol 1 g, regular Targin 5 mg/2.5 mg and PRN oxynorm 10 mg were supplemented. Postoperative day 1 patient was mobilized and on postoperative day 3 patient was discharged home.

The patient signed her consent to write the case report, which was approved by the local ethics committee in UCH Galway.

3. Discussion

General anesthesia (GA) and Spinal anesthesia (SA) can be employed interchangeably in specific surgeries. Each has its pros and cons and may lead to unique impacts on perioperative outcomes. In general anesthesia, a patient is in an unconscious state and are not aware or do not feel other sensations. Conversely, in spinal anesthesia, an anesthesiologist injects anesthetic agents near roots of nerves to anaesthetize the region of the body where surgery is needed. In our case study, the SA, a regional form of anesthesia, was chosen due to its numerous advantages. SA involves using small quantities of a local anesthetic agent into the subarachnoid space to yield a temporary sensation loss and motor functions [10]. The Anesthesiologist positions the needle underneath the L2 in an adult client to prevent spinal cord trauma [10]. SA offers perfect operating conditions for surgical processes beneath the umbilicus, hernia repair, gynecologic/obstetric processes of the uterus, genitourinary procedures, and orthopedic procedures downwards the hip. According to El-Radaideh et al. [10], SA is the preferred anesthetic method when giving anesthesia for individuals undergoing elective cesarean section. The threat of fetal and maternal problems linked with SA is minimal compared to general anesthesia.

There are several advantages associated with the neuraxial blockade. According to Pierce et al. [11], SA is associated with the reduced surgical period, duration in the post anesthesia care unit (PACU), and postoperative pain. In addition, spinal anesthesia leads to reduced postoperative nausea, cases of urinary retention, and more satisfactory cost-efficiency with SA. Other advantages include it being easy to execute, offers perfect surgery conditions for the surgeon, keeping patient's airway patent, and lowered pulmonary problems than general anesthesia [11]. Moreover, SA results in decreased cases of the formation of pulmonary emboli and deep vein thrombosis than general anesthesia.

SA was selected for the pregnant lady with EGPA because anesthetic management of such patients could be complicated by the deficiency of cholinesterase, hypersensitive airway, and multi-organ failure [12]. Due to the necrosis of the vessels of multiple organs in EGPA (formerly known as CSS) , its prominent symptoms include eosinophilia, asthma, and extravascular granuloma. Symptoms may be a risk to pregnant women, and in some women, they may be unable to conceive. The symptoms such as asthma, allergy, and pulmonary infiltrate can be a challenge to general anesthesia, which accounts for the reason why spinal anesthesia was used for the woman in the case study [12]. Besides, there were no contraindications for regional anesthesia such as mono- or polyneuropathies. Hence, spinal anesthesia is used to

decrease the stimulation of airways and ensure smooth pain-free intraoperative and postoperative course.

In conclusion, people with EGPA present with hypersensitivity of the airway, cholinesterase deficiency, and multiple organ dysfunctions. Therefore, anesthetic management of EGPA patients can be problematic and challenging. General anesthesia and Spinal anesthesia can be employed interchangeably in specific types of surgeries. Each has its pros and cons and may lead to unique impacts on perioperative outcomes [13]. Spinal anesthesia is the favored anesthetic method when giving anesthesia for patients experiencing cesarean section. The threat of fetal and maternal complications linked to SA is minimal compared to general anesthesia. Spinal anesthesia was selected for the pregnant woman with EGPA because anesthetic management of such patients could be complicated by deficiency of cholinesterase, hypersensitive airway and multi-organ failure.

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Conflict of Interest

Authors declare that there is no conflict of interest.

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