CLINICAL VIGNETTE

A Rare Case of Granulomatosis with Polyangiitis and Its Anesthetic Implications

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Background

Granulomatosis with Polyangiitis (GPA), formerly known as Wegener's granulomatosis, is one of the most commonly presented forms of systemic vasculitis that typically involve small to medium sized blood vessels[1]. GPA is one of the conditions associated with anti-neutrophil cytoplasmic antibody, an autoantibody believed to amplify the inflammatory process. Its reported annual incidence is 10 cases per million. GPA is observed in all ethnic backgrounds but Caucasians, particularly those of northern European ancestry, have a stronger predilection for this disease. It is equally seen in males and females with a mean age of diagnosis of 50 years.[1] The three pathological hallmarks of GPA are granulomatous inflammation, vasculitis and necrosis. GPA typically presents in a subacute fashion with a prodromal phase starting with benign symptoms such as sinusitis and decreased hearing.

The first manifestations of GPA often include nasal involvement where patients present with persistent rhinorrhea or less commonly, epistaxis and nasal obstruction. Classical clinical features include nonspecific constitutional symptoms including fatigue, myalgias, weight loss and fevers. Additional symptoms involving multiple organ systems include persistent upper respiratory tract and ear infections, migratory or polyarticular arthritis, nodular or cavitary lung lesions and rapidly progressive glomerulonephritis.

Concerning symptoms of disease progression include conductive and sensorineural forms of hearing loss due to granulomatous involvement of the middle or inner ear. Inflammatory lesions of the eye can lead to possible orbital pseudotumors, peripheral keratitis, episcleritis and conjunctivitis. For anesthesiologists, airway involvement is the most concerning. These include inflammation affecting the mouth causing strawberry gums and tongue ulcers, inflammation affecting the trachea leading to subglottic stenosis and scarring below the vocal cords and inflammation affecting the lungs causing pulmonary infiltrates and nodule.

Case Report

A 51-year-old female presented to our preoperative anesthesia clinic for a wide local excision of the vulva and argon beam ablation for biopsy confirmed vulvar intraepithelial neoplasia. Her past medical history included Granulomatosis with Polyangiitis, seizure disorder, congenital tongue defect, left eye blindness, gastroesophageal reflux disease, and post-menopausal spotting. She had no known allergies and current medications were intravenous immunoglobulin once a month, abatacept once a month, methotrexate once a week, and prednisone 16 mg daily.

On physical exam, the patient had a body mass index of 32 with normal vital signs. Airway exam revealed a Mallampati classification III view, adequate mouth opening, but decreased thyromental distance. There were no abnormal lesions or ulcers seen in the oral pharynx, but the dentition was notably decayed. The rest of the physical exam was otherwise normal.

On the day of surgery, the patient was brought to the operating room. Standard monitors consisting of non-invasive blood pressure, EKG, heart rate, pulse oximetry, and capnography were placed. After providing supplemental oxygen via face mask, the patient was given intravenous medications for sedation using midazolam, fentanyl, and propofol. She was prepped, draped, and given local anesthetic with lidocaine 1% at the site of incision. The patient tolerated the sedation well without airway obstruction or other complications. At the end of the procedure, she was taken to the post anesthesia care unit for recovery, did well and was discharged home later that day.

Discussion

Granulomatosis with Polyangiitis (GPA) is a systemic vasculitis that involve small to medium sized blood vessels. GPA can be associated with physiological changes secondary to inflammation that an anesthesiologist should be mindful of when preparing to anesthetize a patient with this disease. Anesthetic management of patients with GPA depends on which of the various organ systems are involved.[2] A difficult airway should be anticipated due to the possible involvement of the upper respiratory tract. Inflammation of the mucosal lining of the upper airway can lead to increased granulation or fibrous tissue formation which can cause airway stenosis. Stenosis of the subglottis and proximal trachea are important complications of GPA that appears in 10% to 16% of patients at some point during the course of their disease.[3] In these patients, symptoms typically range from cough and shortness of breath to potentially life-threatening stridor.

Ideally, prior to their operation, indirect laryngoscopy should be done to rule out airway narrowing. Also in patients with GPA, a difficult airway cart should be readily available and

easily accessible. Items in this cart should include a fiberoptic bronchoscope, different sizes of straight and curved laryngoscopes, oral and nasopharyngeal airways as well as instruments needed for emergency tracheotomy and cricothyroidotomy. Instrumentation of the airway should be done gently and mindful of potential bleeding due to probable friable tissue and granulomas in the airway. In the post-operative recovery period there is an increased risk of laryngeal swelling after extubation.

Pulmonary and cardiovascular concerns that an anesthesiologist should be aware of include dyspnea secondary to pulmonary infiltrations, nodules, fibrosis or hypertension. In patients with cardiac valve destruction or myocardial ischemia secondary to GPA, careful attention should be taken to maintain stable hemodynamics. In these patients, it may be beneficial to obtain cardiac workup including electrocardiogram and/or echocardiography to exclude associated cardiac pathology. In addition, performing an arterial catheter placement may be contraindicated in certain patients with peripheral arteritis due to the potential risk of digital infarcts.[4]

Lastly, due to potential renal involvement in patients with GPA (including rapidly progressive glomerulonephritis), we need to be mindful of administering renally excreted anesthetic drugs. In patients with renal involvement, maintenance dose should be lowered by 20-50% in highly protein bound agents. Additionally, drugs such as midazolam and diazepam depend on renal excretion of its active metabolites. Hyperkalemia should be ruled out in these patients as it can result in arrhythmias. Furthermore, hypertension, coagulation defects and anemia should be considered in patients with renal failure.[5]

Conclusion

This 51-year-old female with Granulomatosis with Polyangiitis and multiple medical comorbidities underwent monitored anesthesia care for a wide local excision of the vulva and argon beam ablation. There were no issues throughout the procedure and she was successfully anesthetized with midazolam, fentanyl, and propofol without any complications. However, one should be aware of the potential anesthetic complications that can arise in these patients including narrowing of the upper airways, dyspnea secondary to pulmonary and cardiovascular abnormalities, and renal involvement which may need adjustment of medication dosing.

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