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Case Report

Thymectomy in a Patient with Myasthenia Gravis and Crohn's Disease - Anaesthetic Challenges

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Abstract

Crohn's Disease (CD) is a chronic, nonspecific, transmural inflammation of the gastrointestinal tract. Myasthenia Gravis (MG) is a chronic autoimmune neuromuscular disease. The concurrence of both the disease is a rare entity. The combinations of Myasthenia gravis and Crohn's disease have many challenges in the perioperative period for anesthesiologist. We report successful perioperative management of a patient with Myasthenia gravis and Crohn's scheduled for thymectomy.

A sixty-two year male with a diagnosis of MG and CD was posted for trans-sternal thymectomy. He was diagnosed with CD five years back, had intermittent exacerbations and being managed with steroid and immunosuppressant. Two months back, he was diagnosed to have MG and optimized with oral pyridostigmine and azathioprine. He was managed with epidural block and general anesthesia along with endotracheal intubation. He required postoperative ventilatory support for around 2 hours and had an uneventful recovery.

The patients with myasthenia gravis associated with Crohn's disease is a rare entity. Thymectomy has been reported to improve symptoms of Crohn's disease and should be done on priority basis for such patients. These patients require vigilant management in the perioperative period to avoid any untoward event. Anaesthesia for this combination can be complicated, drug interactions have to be sought for, side effects of the drugs should be known and adjustments for the same should be made when considering the anaesthetic plan.

Keywords: Anesthesia; Challenge; Crohn's disease; Myasthenia gravis; Thymectomy

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Introduction

Crohn's Disease (CD) is a chronic, nonspecific, transmural inflammation of the intestine which may be seen throughout the gastrointestinal tract, from the oropharynx to the anus. Crohn's disease also manifests itself in many extraintestinal symptoms including eyes, skin, and joints [1]. On the other hand, Myasthenia Gravis (MG) is a chronic autoimmune neuromuscular disease which manifests as weakness of the skeletal muscles which further increases with activity. The muscular weakness is prominent with eye and eyelid movement, facial expression, chewing, talking and swallowing. However other muscles group like that involved in breathing and limb movement may also be involved. The etiology for concurrence of both these disease simultaneously in not well elucidated. However, it appears to be autoimmune in nature. The combinations of Myasthenia gravis and Crohn's disease have many challenges in the perioperative period for anaesthesiologist. These challenges are related to anaesthetic technique and anaesthetic drugs interactions during perioperative outcome. The concerns are also related to manifestation of these diseases including body system involvement. We report here successful management of aesthesia challenges posed by a combination of Myasthenia gravis and Crohn's disease in a patient for thymectomy.

Case Report

A sixty-two year male weighing 50 kg (BMI 25.5 kg/m^2) with a diagnosis of MG and CD was posted for trans-sternal thymectomy. On reviewing the history, he was diagnosed with CD five years back and was presently on oral prednisolone (10 mg) once an alternate day and oral mesalazine (500 mg) thrice a day. He had intermittent exacerbations and was managed with steroid and immunosuppressant. Presently the disease was under remission with ongoing steroid and mesalazine. Two months back, he was diagnosed to have MG and corresponded to Ossermans classification grade II, i.e., mild generalised weakness associated with ocular symptoms (diplopia). The myasthenic symptoms were optimised with oral pyridostigmine (60 mg) thrice a day and azathioprine (50 mg) once a day.

Presently on examination, patient had mild dyspnoea in supine rather than left or right lateral position. His pulse rate was 60/minute and regular and the blood pressure 134/84 mmHg. On auscultation, chest was bilaterally clear with air entry more towards the right than left. Mouth opening was three fingers and modified Mallampati grade of II, short neck, and submental fat was present. Breath holding time was more than 35 seconds. Ptosis of right eye was present. Preoperative investigations were haemoglobin 13 gm/dL, WBC 9500/mm^3, Platelets 2.13 lacs/mm^3, random blood sugar 81 mg/dL, blood urea 34 mg/dL, serum creatinine 1.15 mg/dL. Serum sodium 141 meq/L, S. potassium 3.7 meq/L. Thyroid function tests were within normal limits. Pulmonary function tests revealed mild restriction with a FVC of 70%, FEV1 of 62% and FEV1/FVC of 94% of predicted values. Chest X-ray revealed mediastinal widening and marked tracheal deviation to right. Elelctrocardiogram (ECG) indicated sinus bradycardia (heart rate of 56/min). Ejection fraction

was 60% on echocardiography. MRI of chest revealed a well defined lobulated heterogeneous soft tissue density mass 7.3 x 7.5 cm in anterior mediastinum extending posteriorly and superiorly with multiple specs of calcification noted within it. Mass was displacing trachea to right and extending to neck on the left side. The large solid mass in anterior mediastinum with infiltration was displacing branches of aorta up to sternum and upper dorsal vertebrae; trachea is pushed to right though normal in calibre. Left brachiocephalic vein was compressed. Multiple lymph nodes were seen in periphery of mass of which few are adherent. AchR Ab were positive 13.46 nmol/l (>0.4 nmol/L - positive). On bronchoscopy, trachea was deviated to right and bilateral vocal cords were normal.

Informed written consent for surgery and anaesthesia including epidural and central line were taken. Patient was kept nil per oral for 6 hours. Premedication included oral ranitidine (150 mg) and oral alprazolam (0.25 mg) in the night before surgery. Morning dose of pyridostigmine was given with sip of water. In the operating room, routine monitors (5 lead electrocardiogram, non-invasive blood pressure, pulse oximeter) were attached and 18 G intravenous access was gained in left forearm. Intravenous hydrocortisone 100 mg was administered. Epidural catheter was placed in T10-11 interspinous space in left lateral position. Patient was turned to right lateral position. Anaesthesia was induced with intravenous fentanyl (100 μg) and propofol (100 mg) in incremental doses. Mask ventilation was ensured with sevoflurane in 100% oxygen. Laryngoscopy with glidescope revealed a Cormack Lehane Grade IIa and 7.5 mm ID cuffed flexometallic tube with stylet was passed and secured at 21 cm mark. Injection atracurium 30 mg was given intravenously. Patient's lungs were ventilated on pressure controlled mode with a pressure support of 15-20 cm of water and Positive End-Expiratory Pressure (PEEP) of 5 cm of water. The frequency of ventilation was titrated to keep the end-tidal carbon dioxide between 35-40 mmHg. Anaesthesia was maintained with oxygen air mixture, sevoflurane (0.5-0.8 MAC) and propofol infusion. Epidural space was loaded with 10 mL of 0.125 % bupivacaine with 25 μg fentanyl. After sternotomy, thymus was removed and surgical duration was around 4 hours. The oxygen saturation remained more than 98% throughout the surgery. The heart rate and blood pressure remained within 20% of baseline throughout surgery. The pyridostigmine was given through Ryles tube. Patient was shifted to ICU for further management and ventilated initially on pressure controlled mode with pressure support of 10-15 cm of water and Positive End-Expiratory Pressure (PEEP) of 5 cm of water and gradually weaned to pressure support on return of respiratory efforts. The pressure support was gradually decreased from 15 cm of water to 5 cm of water gradually. He was extubated after 2 hours when respiratory efforts were adequate. The postoperative analgesia was provided with epidural morphine (2 mg) diluted in 8 mL normal saline and administered twice a day and continued for 4 days. Intravenous paracetamol 1 gm was administered thrice a day for 5 days. Postoperative course was uneventful and patient was discharged on 10th postoperative day. Postoperative steroid, pyridostigmine and mesalizine were continued and patient was advised to follow up in oncology and gastroenterology clinics.

Discussion

MG is associated with other autoimmune disorders like rheumatoid arthritis, sjögren's syndrome, systemic lupus erythematosis, diabetes mellitus, hypothyroidism and thyrotoxicosis commonly [2-4]. CD is a chronic, nonspecific, transmural inflammation of the gastrointestinal tract. However, the occurrence of both these disease is not commonly seen. There are only 3 case reports till date describing the coincidence of the two diseases.

MG has been reported to be associated with both Ulcerative Colitis (UC) and CD [5-8]. Autoimmune disorders, including MG, occur more frequently in UC than in CD. Autoimmune dysregulation is the central defect in both MG and this Inflammatory Bowel Disease (IBD). Both IBD and MG may be associated with an elevated Carcinoembryonic Antigen (CEA) and decreased peripheral lymphocyte counts that subsequently normalise following thymectomy [9]. This indicates beneficial effect of thymemctomy in patient with CD which also have concomitant MG. The immunological link between MG and IBD is highlighted by two reports of patients undergoing surgical treatment. One report of a patient with both MG and CD documented improvement in perineal and perianal disease following thymectomy for severe uncontrolled MG [7]. Another patient with both MG and UC demonstrated regression of the myasthenia following proctolectomy [10].

Anaesthesia was managed as standard for thymic mass with myasthenia gravis by avoiding neuromuscular blocking agents prior to intubation, intubating in lateral position, using minimal doses of neuromuscular blockers, epidural analgesia to decrease the dose of inhalational agents, avoiding drugs that precipitate myasthenia, continuation of pyridostigmine and planned late extubation.

Drugs used for Crohn's disease like mesalazine can have interactions with azathioprine (used as immunosuppressant for MG) and can increase the effective dose of azathioprine and chances of toxicity of which low blood count and liver toxicity are most worrisome [11]. These concerns play important role in perioperative management as low blood counts increase risk of infection bleeding. Low platelet count also raises concern for central neuraxial block like epidural catheter placement which remains paramount for adequate pain management in such patients. Monitoring renal functions in patients on mesalazine helps monitoring its side effect on kidneys, and Non-Steroidal Anti-Inflammatory Drugs (NSAIDs) should be avoided [12]. Mesalazine associated lung disease is a known entity including bronchiolitis obliterans, Bronchiolitis Obliterans with Organising Pneumonia (BOOP), and interstitial pneumonitis with the most common was being eosinophilic pneumonitis [13]. Mesalazine associated folic acid deficiency can cause anaemia. Given the gastrointestinal side effects and the possibility of aggravating mucosal inflammation, Non-Steroidal Anti-Inflammatory Drugs (NSAID) should be avoided in IBD [14]. The patients undergoing sternotomy may lead to respiratory compromise and increase morbidity because of surgical handling. So these patients require ventilatory support for varying duration in the postoperative period. Our patient required ventilatory support for around 2 hours but that was primarily because of muscle weakness due to MG.

Conclusion

Myasthenia gravis associated with Crohn's disease is a rare entity. Thymectomy has been reported to improve symptoms of Crohn's disease and should be done on priority basis for such patients. These patients require vigilant management in the perioperative period to avoid any untoward event. Anaesthesia for this combination can be complicated, drug interactions have to be sought for, side effects of the drugs should be known and adjustments for the same should be made when considering the anaesthetic plan.

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