Anaesthesia for resection of phaeochromocytoma in a patient with myasthenia gravis: A case report

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Abstract

Introduction
Myasthenia gravis is a disease affecting the nicotinic acetylcholine receptor of the post-synaptic membrane of the neuromuscular junction, causing muscle fatigue and weakness. Pheochromocytoma is a neuroendocrine tumour of the medulla of the adrenal glands or extra-adrenal chromaffin tissue that secretes excessive amounts of catecholamines. We report a rare case of myasthenia gravis and pheochromocytoma, two anaesthetically challenging conditions co-existing in the same patient, and discuss the anaesthetic considerations and management during the surgical resection of pheochromocytoma.

Case report
A 74-year-old lady with ocular myasthenia gravis and ventricular septal defect was diagnosed with pheochromocytoma during a surveillance computed tomography scan post-thymectomy and radiotherapy. She was optimised preoperatively by various multidisciplinary teams prior to the resection of pheochromocytoma. Our anaesthetic technique included thoracic epidural analgesia, general anaesthesia with the use of Bonfils endoscope for intubation and total intravenous anaesthesia for maintenance, cautious dosing of muscle relaxants and the use of pharmacological agents during tumour manipulation.

Case report
A 74-year-old lady, 172 cm, 48 kg, presented with a background history of hypertension, diabetes mellitus, hyperlipidemia, small perimembranous ventricular septal defect and ocular MG. Her MG has been stable on pyridostigmine 120 mg tds. She underwent total thymectomy for stage II thymoma 2 years ago, followed by postoperative radiotherapy. Surveillance CT thorax done 6 months post-thymectomy showed an incidental finding of a soft tissue mass 2.8 × 2.4 cm in the gastrohepatic region, which was found to be enlarging in the repeat CT abdomen pelvis performed 3 months later. She was referred to surgery and endocrinology. Twenty-four-hour urine collection revealed elevated levels of epinephrine, metanephrine and normetanephrine, and the diagnosis of PCC was made. In preparation for open resection of PCC, she was started on alpha blockade with phenoxybenzamine 10 mg bd, and the dosage was increased progressively. Subsequently, beta blockade was added. Neurology review confirmed ocular MG with no generalised MG symptoms or signs. Cardiology review deemed the patient at low cardiac risk for operation.

We report a case of MG and PCC, two anaesthetically challenging conditions co-existing in the same patient, and discuss the anaesthetic considerations and management during the surgical resection of PCC. Searching Medline data from 1966 to July 2012 revealed no previous case reports of anaesthetic management of these two diseases in the same patient.

Conclusion
Perioperative management of phaeochromocytoma remains an anaesthetic challenge. We have described a safe and effective strategy for anaesthesia in this unusual patient with concomitant myasthenia gravis.
nitroprusside 0.04 µg/kg/min were used briefly for about 7 min. Upon resection of PCC, BP was 85/38 mmHg. A total of IV ephedrine 12 mg, IV phenylephrine 700 mcg and IV noradrenaline at 0.03 µg/kg/min were administered to maintain the blood pressure (target MAP 65 mmHg). Operative finding was that of a 4 × 3cm paraganglioma medial to the left adrenal gland. During the 4-h operation, the patient received a total of 3 l of Ringer’s lactate, 500 ml of hydroxyethyl starch (Voluven) and 1 unit (293 ml) of blood. The estimated blood loss was about 300 ml, the lowest haematomirt being 24%. TOF tested with a peripheral nerve stimulator was 4 with no fade 3 h after the muscle relaxant was given at induction with no top-up dose subsequently. Although she was breathing spontaneously, she appeared to be weak clinically (her hand grip strength was diminished); hence we decided to keep the patient intubated till she was served her usual dose of pyridostigmine.

She returned to the ICU postoperatively where pyridostigmine was resumed immediately. She was then extubated the following day. Chest physiotherapy and incentive spirometry were instituted. Her postoperative BP was 110–130/60–70 mmHg. All antihypertensives were hence continued. Epidural ropivacaine 0.2% with 2 mcg/ml fentanyl at 8 ml/h infusion achieved sensory block between the levels T4–12, and the epidural catheter was removed on postoperative day (POD) 2. The postoperative course was complicated by complete consolidation-collapse of her right lung lower lobe and partial collapse of right lung middle lobe, requiring re-intubation on POD 3, IV antibiotics, mucolytics and intensive chest physiotherapy. She was extubated 3 days later and discharged from hospital stable on POD 14. BP control at home was 120–130 mmHg systolic. Twenty-four-hour urine collection repeated in the outpatient setting was normalised.

**Discussion**

Thorough preoperative evaluation, continuing the daily pyridostigmine, careful monitoring during surgery, sufficient respiration prior to extubation and sufficient respiration and analgesia post-surgery minimise the risk of postoperative mechanical ventilation for MG patients. In addition, this MG patient with PCC should ideally be managed by an experienced team of endocrinologists, endocrine surgeons and anaesthetists. Our patient was optimised preoperatively by various multidisciplinary teams prior to the resection of PCC.

Preoperative pharmacological control of the adverse effects of circulating catecholamines is essential to control arterial pressure, heart rate and arrhythmias and to allow blood volume to be restored to normal. Phenoxybenzamine has been the mainstay of preoperative control of blood pressure in patients with PCC. It produces non-competitive blockade as a result of covalent binding to the receptor, preventing the effects of surges of catecholamine release during the preoperative period. In this patient, phenoxybenzamine was started in small doses and increased gradually until she complained of the side effects of postural hypotension and a stuffy nose. Subsequent introduction of beta-adrenoceptor antagonist (atenolol) is aimed at limiting tachycardia with or without cardiac arrhythmias caused by the secreting tumour and at blocking excessive cardiac sympathetic drive secondary to suppression of the presynaptic alpha-2-regulating mechanism by phenyoxybenzamine.

Prolonged exposure of the circulation to high-circulating noradrenaline concentrations results in constriction of both arteriolar and venous segments with a marked decrease in circulating blood volume. Induction of anaesthesia may cause widespread venodilation, leading to profound arterial hypotension. She was therefore admitted a few days prior to the operation for intravenous

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**FOR CITATION PURPOSES:** Loh SKN, Chong SY. Anaesthesia for resection of phaeochromocytoma in a patient with myasthenia gravis: A case report. OA Anaesthetics 2013 Aug 01;1(2):14

All authors abide by the Association for Medical Ethics (AME) ethical rules of disclosure.
hydration. She was admitted to ICU a day prior to the resection. Intra-articular and central venous lines were inserted, so as to guide blood pressure and fluid management. She was generously given 5.6 l of fluids over 24 h to increase her CVP to >8 mmHg.

For this patient, we have elected to use the combination of total intravenous anaesthesia (propofol and remifentanil) and segmental mid-thoracic epidural analgesia for surgery. This prevents the patient from coughing on the tracheal tube intraoperatively and provides satisfactory conditions for surgical incision and exposure of the tumour. Manipulation of the PCC during open surgery, however gently performed, causes a brisk haemodynamic pressor response. In our case, a combined regional and general anaesthetic technique with the use of selective adrenergic antagonists helps suppress haemodynamic surges in response to tumour manipulation.

MG patients present an increased sensitivity to neuromuscular depolarising blocking agents (NMBA) because of reduced number of receptors. Once the epidural block is effective, it can minimise the need for further doses of intravenous opioid or NMBA during open abdominal surgery. It is essential to use neuromuscular monitoring during surgery and ensure full recovery (TOF >90%) prior to terminating the anaesthesia. In our case, a combined regional and general anaesthetic technique with the use of selective adrenergic antagonists helped suppress haemodynamic surges in response to tumour manipulation.

Case report

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Acknowledgement

Support was provided solely from institutional sources.

Abbreviations list

CT, computerised tomography; CVP, central venous pressure; MG, myasthenia gravis; NMBA, neuromuscular depolarising blocking agent; PCC, phaeochromocytoma; POD, postoperative day; TOF, train of four.

References