Clinical Anaesthesia File

Anaesthetic management of a patient with myasthenia gravis and tracheal stenosis

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Purpose: The combination of myasthenia gravis and tracheal obstruction presents a number of difficulties for anaesthetic management. This case illustrates the advantages of careful planning.

Clinical features: A 66-yr-old man with myasthenia gravis required resection of a stenosis at the site of an old tracheostomy. The primary goal was to accomplish safe management of the airway, a task made more difficult because the airway was shared with the surgeon. Awake fiberoptic examination of the tracheal stenosis performed in the operating room provided useful information in planning the subsequent anaesthetic. From this examination, it was found that the trachea could be intubated by a normal endotracheal tube passed through the stenosis over the fiberoptic bronchoscope. Intraoperatively, the orotracheal tube was withdrawn temporarily and replaced with an endotracheal tube placed by the surgeon into the distal trachea. Extubation was carried out judiciously and a plan for reintubation prepared in advance. The anaesthetic plan was modified because of the myasthenia gravis. Following careful investigation of the extent of the patient's disease and its treatment, an assessment was made of the patient's need for postoperative ventilation. The anaesthetic plan included maintenance of anticholinergic medications until the time of surgery and their early resumption postoperatively, avoidance of neuromuscular blocking agents, and careful monitoring of neuromuscular function during the anaesthetic.

Conclusion: Careful examination of the area of tracheal stenosis and a carefully considered plan for reintubation are prerequisites for this type of surgery. Clinically well controlled myasthenia gravis was managed successfully using familiar principles.

Objectif: L'association de la myasthénie grave avec une trachéosténose peut influencer la conduite de l'anesthésie. Cette observation illustre les avantages d'une préparation méticuleuse.

Caractéristiques cliniques: Un myasthénique âgé de 66 ans a subi une résection de trachéosténose sur le site de sa trachéostomie. L'objectif fondamental était de maintenir la perméabilité des voies aériennes pendant l'intervention. La fibroscopie réalisée en salle d'opération, avant la chirurgie a fourni des renseignements très utiles pour l'anesthésie. Cet examen a révélé qu'on pouvait introduire par le fibroscope une canule endotrachéale ordinaire au travers de la sténose. Pendant l'intervention, la canule endotrachéale a été retirée temporairement et remplacée par une canule insérée par le chirurgien dans la partie distale de la trachée. Après avoir planifié une réintubation éventuelle, on a précautionneusement extubé le patient. La gestion anesthésique a toujours tenu compte de l'existence de la myasthénie. Après une analyse rigoureuse de la gravité de la maladie et de son traitement, une ventilation postopératoire éventuelle a été organisée. Le planification de l'anesthésie avait prévu le maintien de la médication anticholinergique jusqu'au moment de la chirurgie et sa reprise dès que possible après celle-ci, l'abstention de myorésolutifs et le monitoring précis de la fonction neuromusculaire après l'anesthésie.

Conclusion: L'examen attentif du site de la trachéostomie et un plan détaillé pour l'anesthésie sont essentiels pour gérer ce type de chirurgie. C'est grâce à des principes reconnus, qu'on a pu gérer un cas de myasthénie grave sous contrôle.

A 66-yr-old man was scheduled for resection of a subglottic tracheal stenosis. The stenosis was at the site of an old tracheostomy that had closed. The tracheostomy had been created one year earlier for persistent vocal cord paralysis later found to be caused by myasthenia gravis (MG). The MG was treated with thymectomy, prednisone (10 mg every 2 days) and pyridostigmine (450 mg daily). The patient noted significant improvement on this regimen. His only related symptoms were mild dysphagia for solids and slight right ptosis. However over the previous 5 months, he had episodes of stridor at rest and increasing limitation of his activity level. He was an ex-smoker but had no symptoms suggesting COPD.

This patient has several problems. First, his stridor implies intermittent airway narrowing to 4.0 mm or less, hence this is a difficult airway. Further information concerning the location and severity of this obstruction is necessary to formulate a plan of airway management. The upper airway must be carefully examined. An assessment should be made of any potential difficulties for normal methods of airway control, such as fiberoptic bronchoscopy.

Second, the planned surgery poses several concerns.

Intermittent disruption of airway continuity may be required. Also, as with any major surgery in the neck, there is the risk of injury to nearby structures including the recurrent laryngeal nerve, phrenic nerve, vocal cords, esophagus, apices of the lungs, and major vessels. Rapid blood loss is possible. The location of the stenosis is important as lesions of the lower trachea or carina cannot be approached through the neck but require sternotomy or thoracotomy.

Third, the management is complicated by MG. A full history of the symptoms, onset, duration, progression, and current treatment of the patient's MG should be obtained to assess the severity of the disease. Severe disease is suggested by generalized weakness, especially if rapid in progression, and bulbar or respiratory muscle dysfunction. While certain factors including duration of MG greater than six years, history of chronic respiratory disease, pyridostigmine dose greater than 750 mg per day, and vital capacity of less than 2.9l have been suggested to be predictors of respiratory failure following trans sternal thymectomy, there is debate about their applicability for other types of surgery.^{1,2}

If bulbar muscle weakness is present, the patient is at increased risk of aspiration and preoperative pneumonia should be ruled out. Also, nutritional status may be sub-optimal if dysphagia makes oral intake difficult. The patient's treatment should also be reviewed. Pyridostigmine may alter the response to ester local anaesthetics and neuromuscular blockers, succinylcholine and mivacurium. If the patient has been treated with steroids recently, as in this case, stress-dose intravenous steroid coverage should be provided during the perioperative period. Both thymectomy and plasmapheresis may produce significant improvement in myasthenic patients.

Finally, MG may be associated with other autoimmune disorders such as thyroid and connective tissue diseases including rheumatoid arthritis.³

Physical examination showed an 81-kg man with a scar at the base of the anterior neck. He was edentulous with full cervical spine movement. His airway was assessed as Mallampati grade I. His voice was hoarse but no stridor was present at rest or when supine. The lung fields were clear to auscultation. Neurological and musculoskeletal examination was normal except for a mild right ptosis.

More information is required to complete the assessment of his airway obstruction. Bronchoscopy, CT scan of the neck, tomograms, and fluoroscopy of the stenoic segment, and flow-volume loops may provide useful

information on the location, length, and diameter of the stenosis as well as dynamic changes of the tracheal lumen with respiration. Although there were no definite symptoms or signs of COPD, he had been a long-term smoker which may increase the risk of postoperative ventilation in this situation.³ Reduced vital capacity secondary to the MG is also a risk factor. For these reasons, the patient requires pulmonary function testing and measurement of arterial blood gases. A chest x-ray will rule out underlying pulmonary pathology including aspiration pneumonia, and may show the stenosis.

His CBC, electrolytes, and glucose were in normal range. On room air, arterial blood gases were: pH 7.36, PCO₂ 40 mmHg, PO₂ 75 mmHg, and HCO₃ 22 mmol·L⁻¹. A chest x-ray was also normal. Pulmonary function testing revealed a normal FEV₁ (3.13 L), FVC (4.20 L), and diffusing capacity but inspiratory flow rates were low (PIF = 2.22 L·sec⁻¹) suggesting a variable extrathoracic obstruction. A 1.0 cm subglottic narrowing was seen on CT of his neck. The stenosis did exhibit narrowing with inspiration on fluoroscopy. A bronchoscopy performed 2 months earlier described a 2.9 mm stenosis 4–5 cm distal to the cords. A swallowing study was also done because of his dysphagia and indicated mild bulbar weakness.

It is important to review this case with the surgeon before formulating a plan for anaesthesia.

The surgeon planned to resect several complete tracheal rings at the site of stenosis. At completion of the case, the patient's chin was to be sutured to his chest to reduce tension on the anastomosis. To reduce irritation of the anastomosis by the endotracheal tube, the surgeon requested the patient be extubated as soon as possible postoperatively (preferably in the operating room).

At this point, the options for anaesthetic management should be reviewed. Although regional anaesthesia is the anaesthetic of choice in patients with MG whenever possible,³ this case could not be done with a regional technique alone. However, bilateral superficial cervical plexus blocks in combination with general anaesthesia may reduce total anaesthetic requirements thereby diminishing the likelihood of postoperative ventilatory

failure and providing some postoperative analgesia. In considering airway management, the important problems are airway obstruction, MG and surgical technique. Three concerns must be specifically addressed: airway stenosis may make passage of an endotracheal tube difficult; good exposure of the posterior tracheal wall is necessary to perform the reanastomosis; and early, safe extubation is desired.

Preoperative investigation suggest the lumen diameter at the stenosis to be approximately 2.9 mm, which would not allow passage of an endotracheal tube of adequate size. However, this degree of obstruction is not compatible with the current absence of stridor at rest. In patients with tracheal stenosis, ventilation has been achieved through an endotracheal tube placed below the cords but just above the stenosis.⁴ There is no preoperative guarantee of success with this technique especially if the stenosis varies with changes in airway pressure. For these reasons, the safest approach may be to reassess the airway using the pediatric fiberoptic bronchoscope in the operating room with the patient awake. Further management will depend on the findings at fiberoptic bronchoscopy. The fiberoptic bronchoscope may show the lumen to be larger than predicted, in which case a normal endotracheal tube can be passed with fiberoptic guidance. The surgeon may also be able to dilate the stenosis with a rigid bronchoscope or dilators to allow passage of a larger tube. Another option would be jet ventilation through a catheter passed through the stenosis. Jet ventilation would allow the surgeon an obstructed field, decrease the movement of the airway and chest, and the catheter could be passed beyond the stenosis. However, problems with its use include the need for an intravenous anaesthetic with adequate neuromuscular blockade, aerosolization of blood and debris in the surgical field, and the risk of barotrauma. Heliox may also be helpful temporarily and should be readily available. Tracheostomy may be considered if it could be performed below the obstruction. An option of last resort is femoral vein to femoral artery cardiopulmonary bypass.

During construction of the posterior wall anastomosis, the surgeon's exposure will be blocked by the endotracheal tube. The tube must therefore be pulled back above the incision and replaced either by a jet ventilation catheter⁵ or by a sterile endotracheal tube placed through the incision into the distal trachea.^{6,7} The orotracheal tube can be readvanced once the posterior anastomosis is completed.

Airway management is also a concern at time of extubation. As the patient's chin will be sutured to his chest, reintubation even with a fiberoptic bronchoscope would

be difficult. It is therefore imperative that the patient be fully awake and cooperative but with no residual muscle weakness, a challenge in this patient because of MG. Theoretically, the airway obstruction should have been relieved by the surgery but the possibility of airway compromise persists, either from oedema or from other complications such as recurrent laryngeal nerve injury. Before attempting extubation, there must be a plan for emergency reintubation if airway obstruction does occur. It is logical to extubate over a guide for reintubation. A hollow tube changer can be left in place and provides the additional advantage of providing a means for jet ventilation should the need arise. Another option would be to extubate over the fiberoptic bronchoscope. It may be necessary to remove the chin suture to facilitate reintubation as a last resort.

The other problem to be considered in the anaesthetic plan is MG. This patient has a past history of respiratory failure requiring tracheostomy due to his MG, but subsequently he has improved since his thymectomy. Prolonged postoperative ventilation is unlikely on the basis of MB because of its relatively short duration, the low dose pyridostigmine, and normal lung function.³ In preparing a myasthenic for surgery, some benefit may be obtained from preoperative plasmapheresis and continuation until the time of surgery. A neurologist should also be involved in the patient's perioperative care. If bulbar involvement is suspected, premedication with H₂-blockers and metoclopramide should be considered because of the risk of aspiration. Stress-dose intravenous steroids should be used in this case. If awake fiberoptic bronchoscopy is planned, an antisialagogue such as glycopyrrolate should be given. The intensive care unit should also be notified to expect the patient for post-operative monitoring.

Once the airway is secured, general anaesthesia can be induced. Careful monitoring, especially to ensure adequate ventilation, is important because of ongoing airway manipulation. Additional monitors to be considered are an arterial line and an oesophageal or precordial stethoscope. Large bore intravenous access is necessary because of the risk of rapid blood loss. Goals for the general anaesthetic are to have a deeply anaesthetized patient during airway manipulation to prevent coughing, to avoid depressants of neuromuscular function, and to have a cooperative fully-awake patient for extubation. Patients with MG will have unpredictable responses to both depolarizing and non-depolarizing neuromuscular blockers so consideration should be given to their avoidance. If they are to be used, careful monitoring with a nerve stimulator is mandatory and shorter-acting agents may be of benefit.

Actual course

After discussion with the patient and surgeon, it was decided to proceed with an awake fiberoptic bronchoscopy to assess the stenosis. Following insertion of an arterial line, the patient was premedicated with glycopyrrolate 0.2 mg *iv* and midazolam 0.2 mg *iv*. Aerosolized lidocaine was used to anaesthetise the upper airway. Heliox, a rigid bronchoscope, and the jet ventilation catheter and ventilator were available in the operating room. The 4.7 mm diameter bronchoscope easily passed through the subglottic stenosis and the patient was able to breath around it. A 7.0 armoured orotracheal tube was advanced over the bronchoscope and through the stenosis so the cuff lay just above the carina.

General anaesthesia was then induced with propofol and fentanyl, and maintained with fentanyl, isoflurane, oxygen, and air. No neuromuscular blockers were given. Bilateral superficial cervical nerve blocks were done with bupivacaine 0.25%. During anastomosis of the posterior tracheal wall, the orotracheal tube was temporarily pulled back with ventilation via through a sterile endotracheal tube passed by the surgeon through the incision into the distal trachea. When the anastomosis was complete, the orotracheal tube was readvanced. Near the end of the surgery, the volatile anaesthetic was discontinued and a propofol infusion started. The patient's chin was sutured to his chest to maintain neck flexion. A 0.035 inch diameter Teflon-coated guide wire was passed through the endotracheal tube. The patient was allowed to awake gradually as the propofol infusion rate was reduced and alfentanil was titrated to maintain respiratory rate. The patient was extubated once fully awake and possessing good motor power and a tidal volume of 600–900 ml. The wire was left in place to act as a guide to be placed through the suction port of a fiberoptic bronchoscope to facilitate reintubation. He remained stable in the intensive care unit and was restarted on pyridostigmine given via a nasogastric tube. He made an uneventful recovery with significant improvement in his stridor.

Discussion

Tracheal stenosis

Patients with tracheal stenosis present with stridor, shortness of breath on exertion, and inability to clear secretion. Tomograms of the neck, bronchoscopy, and

flow-volume loops are good tests to diagnose and evaluate a stenotic lesion. Plain x-rays of the neck or chest or CT scanning of the neck may also be helpful. On pulmonary function testing, several patterns can be seen on the location of the obstruction and whether the stenosis is fixed or varies with changes in pressure across the wall. A variable extrathoracic obstruction as in this patient will cause obstruction to inspiration as the airway is no longer held open by positive pressure as in the expiratory phase. The converse is true for a variable intrathoracic lesion where obstruction is maximal in exhalation. Negative pressure in the chest cavity holds the intrathoracic airways open in inspiration only. A fixed obstruction at any level affects both the inspiratory and expiratory phases.⁷

SURGICAL TECHNIQUE

Treatment of the stenosis can involve dilatation, laser excision, or resection.⁵ During resection, a common approach is to pull the orotracheal tube above the incision while a sterile tube placed through the incision until the posterior anastomosis is done.^{6,7} The patient's chin is usually sewn to his chest for two weeks postoperatively.^{6,7} There is controversy regarding the timing of extubation but early extubation has been successful in 66.7–84.2% of patients.^{4,6} Complications include leak at the anastomosis, vocal cord paralysis, restenosis, dysphagia, and atelectasis.

ANAESTHETIC CONSIDERATIONS

Bronchoscopy in the operating room is suggested to assess the stenosis and dilate it if necessary to facilitate ventilation. Subsequent options for airway management are to proceed with fiberoptic intubation if an endotracheal tube can pass through the lesion, ventilate the patient with an endotracheal tube placed just above the stenosis,⁴ jet ventilate, perform a tracheostomy, or use cardiopulmonary bypass.⁴ The patient may need close monitoring of airway patency post-operatively in the intensive care unit.

Myasthenia gravis

Myasthenia gravis is an autoimmune disorder due to production of antibodies against the acetylcholine receptor resulting in a reduction of the number of functional receptors at the neuromuscular synapse.⁸ Repetitive stimulation of a motor nerve results in progressively diminishing action potentials in the muscle (fatigue).³ Clinically, MG presents with fluctuating weakness that may only involve the extra-ocular muscles, producing ptosis or diplopia (14%), or may become more generalized (86%).³ The disease is typically more severe and rapidly progressive in males.³ Bulbar muscle weakness

causes dysphagia, dysarthria, and risk of aspiration. Myasthenia gravis is often associated with abnormalities of the thymus gland and other autoimmune disorders. Treatment involves immune suppression with steroids or azathioprine, boosting neuromuscular transmission with cholinesterase inhibitors, removing antibodies by plasmapheresis, and thymectomy.

The main concern for anaesthetists are perioperative respiratory and bulbar muscle strength, and the effects of treatment. An assessment of the risk of post-operative ventilation should be made. Criteria that have been proposed to predict risk following trans sternal thymectomy are listed above. Careful attention to preoperative pulmonary function testing may also be helpful in predicting postoperative respiratory problems;² however, concurrent tracheal disease in this case limits its utility.

Plasmapheresis may be helpful in preparing patients with MG for anaesthesia and surgery. There is controversy concerning whether anticholinesterases should be continued perioperatively as vagal responses can be potentiated, metabolism of ester local anaesthetics and succinylcholine inhibited, and requirements for non-depolarizing neuromuscular blockers possibly increased. In most cases, the patient's medical condition prevents cessation of the anticholinesterase medication. Sedative premedication should be avoided since these patients have little respiratory reserve and often have increased risk of aspiration. Regional anaesthesia may be preferred in some instances. Response to muscle relaxants is unpredictable. Patients may be resistant to succinylcholine due to diminished numbers of available receptors but sensitive to non-depolarizing agents. Muscle relaxants should therefore be avoided, or if used monitored closely and shorter-acting drugs chosen. The trachea should be extubated only once normal muscle strength has returned.

Conclusion

The combination of tracheal obstruction and MG presents a number of difficulties in anaesthetic management. The goal of the anaesthetist is to accomplish safe management of the airway, a task made more difficult because the airway will be shared with the surgeon. In this case, the tracheal stenosis was less severe than initially thought; however, careful preoperative assessment and a well planned anaesthetic approach were essential. In the operating room, awake fiberoptic examination of the tracheal stenosis provided useful information in planning the subsequent anaesthetic. Based on this examination, it was found that the trachea could be intubated with a normal endotracheal tube passed through the stenosis. Intraoperatively, the orotracheal tube was temporarily withdrawn and replaced by an endotracheal

tube placed by the surgeon into the distal trachea. Extubation was carried out judiciously and a plan for reintubation prepared in advance. Myasthenia gravis did not present significant problems in this case. Following careful investigation of the extent of the patient's disease and its treatment, an assessment of the patient's need for postoperative ventilation was made. The anaesthetic plan included maintenance of anticholinergic medications until the time of surgery and their early resumption postoperatively, avoidance of neuromuscular blocking agents, and careful monitoring of neuromuscular function during the anaesthetic.

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