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Case Report Open Access

Video-Assisted Thoracoscopic Surgery in an Adult Patient with Myotonic Dystrophy: A Case Report and Review of the Literature

Daniel Braunold[1], **Ahmad Mahajna**[1,2] **and Michal Barak**[1,2*]

[1]*Department of Anesthesiology, Rambam Health Care Campus, POB 9602, Haifa 31096, Israel*

[2]*The Bruce Rappaport Faculty of Medicine, Technion-Israel Institute of Technology, Haifa, Israel*

[*]**Corresponding author:** Michal Barak, Department of Anesthesiology, Rambam Health Care Campus, Israel, Tel: +972-4-777-2487; Fax: 972-4-777-2961; E-mail: m_barak@rambam.health.gov.il

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Abstract

A 52 year old intellectually disabled woman with myotonic dystrophy type 1 and a history of malignant melanoma underwent an elective thoracoscopic neoplasm resection. Pre-operatively she had very poor pulmonary function tests with a vital capacity of 1.99 L. The operation was conducted under total i.v. anesthesia with a single dose of 20 mg rocuronium and one lung ventilation was achieved with left double lumen tube. At the end of the surgery the patient regain conciseness and was extubated. Following extubation the patient quickly deteriorated into type 2 respiratory failure with significant hypoxemia. She was alternately ventilated with face mask and breathing spontaneously for about 2 h until fully recovered.

Keywords: Myotonic dystrophy; Mechanical ventilation; General anesthesia

Introduction

Myotonic dystrophy (MD) is a multisystem, autosomal dominant disorder, best known for its skeletal muscle manifestations. Its prevalence in the general population is 1-10:100,000 [1,2]. It can present as muscle wasting and fatigue, and the disease also involves the cardiovascular, respiratory, endocrine and nervous systems [1,3,4]. Cognitive dysfunctions, and intellectual and personality disturbances may appear as well. MD has been divided into two subtypes, with type 2 being the milder of the two forms.

From an anesthetic view, the management of the MD patient presents a challenge to the caretaker. Both cardiovascular and pulmonary impairments significantly increase the anesthetic risk. While an acute cardiac event may be life-threatening during surgery, post-operative quality of life and prognosis often depend upon the degree of lung function impairment. Characteristically, MD patients tend to have hypoxemia and hypercapnia, and their pulmonary function tests often show a restrictive pattern [3,5]. If a MD patient requires mechanical ventilation, they are at risk for a lengthy weaning process or becoming ventilated permanently.

Here we present an adult MD, mentally challenged patient with poor pre-operative pulmonary function, who underwent thoracoscopic surgery and subsequently had problems recovering effective breathing post-operatively.

Case Report

The patient was a 52 year old mentally challenged woman with MD type 1 and a history of malignant melanoma. She had complained of a cough, and a subsequent CT showed a mass in her right lung. FNA biopsy was deemed difficult due to mass positioning and patient compliance, so she was scheduled to have a right thorascopic wedge resection with possible lobectomy, depending on the intra-operative pathology results.

Pre-operative evaluation showed a very pleasant, cooperative and optimistic woman. Due to her moderate intellectual disability, her elderly mother accompanied her for the assessment. The patient was living in sheltered accommodation along with other handicapped people and ambulated with the aid of a walker. Upon physical examination, she was 1.52 m in height and weighed 65 kg. Her respiration was effortless, with a rate of 12-14 breaths per minute with no added breath sounds noted on auscultation. Her saturation was 76% on room air.

Pre-operative pulmonary function test showed severe restrictive lung disease with FEV1 0.75 L at 33% prediction, FEV1/FVC 115% predicted, PEF 43%, MEF 25-75 45% prediction, VC 27% prediction (1.99 L) and TLC of 1.72 L at 38% prediction. She was unable to perform CO diffusion analysis. Her ECG showed normal sinus rhythm and her echocardiogram was unremarkable with normal left ventricular ejection fraction. The anaesthetic and surgical risks were explained in detail to the patient and her mother.

The anesthetic plan was to induce and maintain GA without inhalation agents, using total i.v. anesthesia (TIVA), and to try to avoid using muscle relaxants. However, if events would necessitate the use of muscle relaxation, rocuronium would be used and would be reversed with sugammadex at the end of the procedure. The use of opioids was to be limited to a minimum, with preference for remifentanil because of the rapid termination of its effect.

General anesthesia was induced with 150 mg of propofol and 100 mcg of fentanyl. A 35 f left double lumen tube was placed in the trachea, with its bronchial port above the carina, and not in its correct position in the left main bronchus. At that time it appeared that muscle relaxation are necessary in order to introduce the tube to its place without causing trauma to the airways. Rocuronium 20 mg was administered and the tube was placed in its correct position, as verified with a fiberoptic bronchoscope. General anesthesia was maintained

with propofol and remifentanil *via* continuous drip. Local anesthetics were injected into the chest wall before trocar insertion. Ventilation parameters were: tidal volume of 350 ml, respiratory rate of 15 per min, and FiO_2 of 1.0. Blood gas analysis post-induction showed pH 7.43, paO_2 84 mmHg, $paCO_2$ 48 mmHg, bicarb 31.9 and BE +6.9. The wedge resection was performed and pathology showed malignant melanoma thus the procedure was ended. The operation took one hour during which the patient received no additional muscle relaxants.

At the end of the surgery, a chest drain was placed, the right lung was re-inflated, and TIVA turned off. Sugammadex 200 mg was given. After five minutes the patient woke up from anesthesia and began to cough on the tube. She was responsive to our comments. She was extubated and the face mask from the anesthesia machine was placed to enable monitoring of her post-extubation breathing. She was positioned in a sitting position, for her comfort and for easier breathing. Despite the ability of the patient to communicate *via* nodding and shaking her head, she was unable to ventilate effectively and could only generate minimal tidal volume while struggling for air. Shortly afterwards the patient desaturated to 55% and was ventilated *via* face mask for 5 min. Blood gasses were then taken and showed: pH 7.17, pa 02 73, $paCO_2$ 91 mmHg, bicarb 32 and BE 1.6. A second dose of 200 mg sugammadex was given.

Over the next 45 min the patient was breathing at her best capacity. When she desaturated below 75%, she was ventilated with the face mask back to normal saturations and then allowed to attempt to self-ventilate. A quick discussion was held about what the next stage should be. Options that were discussed include: (a) re-intubation and continue mechanical ventilation for a few hours, (b) non-invasive ventilation, and (c) wait and continue face mask ventilation if the patient began to deteriorate. It was decided to wait for another hour. Every 20 min a blood gas was sent for analysis. A table below shows the patient's blood gas analysis at that time (Table 1).

Time	11:55	13:21	13:36	13:50	14:05	15:00	18:15	21:00	23:30
pH	7.44	7.17	7.09	7.09	7.11	7.15	7.16	7.2	7.25
paO_2 mmHg	84	73	60	63	65	78	125	61	71
$paCO_2$ mmHg	48	91	99	109	99	95	86	80	75
HCO_3	31	33	30	33	32	29	30	31	32
BE	6.9	1.6	-2	0.6	1.2	-1.5	0.4	0.3	3.2
The time line was: extubation at 13:15, transferred to PACU at 14:45, to the ward at 20:00. BE=base excess.									

Table 1: Patient's blood gas results.

Despite the CO_2 levels climbing higher and worsening acidosis, the patient's clinical status was actually improving; she became wakeful and started to moan with extended expiratory noises. Gradually her blood gasses also showed a small improvement dropping the $paCO_2$ from 109 to 99 mmHg. She then began talking and her cough became stronger. After this, she was taken to the Post Anesthesia Care Unit, where she was monitored and treated for pain. Several hours later the patient was transferred to the Thoracic Surgery ward where a physiotherapist attended her. Later that evening, she pulled out the chest drain by herself. The next morning another session of respiratory physiotherapy was held. The rest of the hospitalization was uneventful and the patient was discharged from hospital back to sheltered accommodation after five days.

Discussion

This case presents the dilemma of anesthetizing a patient with compromised pre-operative respiratory functions for a lung resection. The British Thoracic Surgical guidelines debate whether the "cut-off" for lobectomy surgery should be predicted postoperative (PPO) FEV1 >40% and DLCO >40%, since it was suggested that these calculations can overestimate the impact of decreased lung function and, therefore, quality of life post operatively [6]. In their prospective study, Brunelli A et al. showed that PPO could be as low as 30% for FEV1 and DLCO with a post-operative mortality rate of 4% [7]. Similar results were demonstrated in several larger scale studies [8,9], leading us to believe that the predictive value of pre-operative pulmonary function tests is limited [10]. Considering the imprecision of prediction of postoperative outcomes and the necessity for surgical treatment for the patient, we tend to push the boundaries and take patients with worse pre-operative pulmonary conditions into lung surgery and general anesthesia.

In MD, the physio-pathological cause for respiratory impairment is multifactorial and includes respiratory muscle weakness, altered central ventilatory control, reduced CO_2 sensitivity, impaired ventilatory mechanics that lead to global alveolar hypoventilation, microatelectasis, and reductions in lung compliance [4,5]. For these reasons it is better to avoid tracheal intubation and mechanical ventilation in MD patients. However, this is not always possible, as was the case with our patient, where her procedure required both endotracheal intubation and ventilation. The challenge was to bring the patient back to her basic respiratory function as it was before the surgery.

One of the most important issues is the timing of extubation and the type of post-extubation respiratory support, in case it is needed. The attending anesthesiologist chose to extubate immediately after surgery. A short period of mechanical ventilation could have been undertaken in the ICU, with full monitoring of pulmonary functions, resistance and compliance. ICU ventilator is capable of providing sophisticated ventilation modes. However, the patient was extubated. The decision to extubate our patient at the end of the surgery was taken relying on several considerations: (a) the clinical impression of her muscle tone while coughing with the tube, (b) the operation was limited: thoracoscopic wedge resection [11], (c) she received a minimal dose of rocuronium, and then sugammadex, (d) it was a double lumen tube,

and (e) it was not a difficult intubation, thus she could be re-intubated if needed. Post extubation the patient deteriorated rapidly, developing significant respiratory failure. A decision had to be made about whether to re-intubate and ventilate the patient or to attempt some other form of ventilation. Given the patients history of MD along with very poor respiratory function and underlying pathology, intubation and continued mechanical ventilation could easily have turned into a prolonged, or even permanent, respiratory support. This option was kept as a last resort. Another option was to place the patient on non-invasive ventilation to help improve the hypoxemia and hypercapnia. This patient had previously had an assessment for non-invasive ventilation at home but was un-compliant with the treatment at the time. It should be noted that most guidelines suggest the use of non-invasive ventilation to improve respiratory failure with hypercapnia but show a higher failure rate with an acidosis of <7.25 [12,13]. In this case the patient's acidosis was severe and, given a previous history of failed compliance with non-invasive ventilation when at their baseline, thus it was thought that non-invasive ventilation would be unsuccessful. The third option was to face mask ventilate the patient for a few minutes whenever she was deeply desaturated and to improve immediate hypoxemia, then allow the patient to attempt to breathe alone, unaided. This cycle was repeated several times, while carefully monitoring her status both clinically and with blood gas tests. The patient required about an hour of alternate face mask ventilation and spontaneous breathing until she seemed to stabilize and recover.

Another consideration in managing the MD patient is drug selection, especially the use of neuromuscular blocking agents. Ideally one would prefer to avoid the use of muscle relaxants but intubation conditions were not ideal without the patient being paralyzed. The correct position of the double lumen tube is essential for the conduction of thoracoscopic surgery. Still, the use of sugammadex in MD patients has been shown to be effective and induces rapid reversal of blocking agents [14,15]. Maintenance of anesthesia was with TIVA instead of inhaled anesthetics since volatile anesthetics relax skeletal muscle, exacerbating and potentiating neuromuscular blocking agents which would make breathing more difficult post-operatively in this case [16].

This patient has a chronic degenerative muscular condition and very poor respiratory function. In this case there were multiple considerations to take into account, including whether which agents to use, how long to ventilate the patient, and how best to wean her off the ventilator. We believe that decisions should be based primarily upon the patient's clinical picture and not on lab results, and that patience is sometimes the best practice.

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