

Anesthetic Management for Oophorectomy in a Patient with Lymphangiomyomatosis

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Lymphangiomyomatosis is a rare disease occurring in females of reproductive age. This disease results from the benign proliferation of smooth muscle in the muscle lining of the abdominal and thoracic lymphatics, veins, and bronchioles, and is characterized by a rapid deterioration in respiratory function resulting in death almost always within 10 yr.¹⁻³

A hormonal etiology for this disease process has been postulated, and estrogen and progesterone receptor populations and antibodies in affected lung tissue have been measured.⁴ Medical and surgical manipulations of hormonal levels have become the basis of treatment.^{5,6}

We describe here the anesthetic management of a patient with pulmonary lymphangiomyomatosis for a cervical cone biopsy and a vaginal hysterectomy and bilateral salpingo-oophorectomy.

CASE REPORT

A 43-yr-old woman with a known diagnosis of pulmonary lymphangiomyomatosis following an open lung biopsy 2 yr ago presented for surgery as a result of a cervical smear that showed cervical cell dysplasia. Consent was obtained for a cone biopsy, vaginal hysterectomy, and bilateral salpingo-oophorectomy.

She had had a general anesthetic for a neck lymph node biopsy in 1968 and a spinal anesthetic for a cervical cone biopsy in 1979, both without complication. The open lung biopsy in 1986 for diagnostic purposes of chest radiological appearances and poor pulmonary function resulted in ventilatory support for 3 days in the intensive care unit. Her exercise tolerance and measured lung function had returned to its preoperative state.

She complained of exertional angina, her last attack being 2 months before this admission. Her exercise tolerance was now severely limited and determined by symptoms of dyspnea and cough. She was receiving added oxygen at home. She also gave a 15-yr history of a nonspecific bleeding dyscrasia that had been unresponsive to a recent course of desmopressin acetate injections. She had stopped smoking 2 yr prior to this admission. Before that, she had smoked half a pack a day for 15 yr. There were no known allergies and no family history of other diseases or anesthetic complications. Her medications included aminophylline, bumetanide, metaproterenol inhalers, oxygen *via* nasal cannulae, and, recently, depoprovera.

On examination, she was a well-nourished, elderly looking woman,

134 cm tall and 70 kg in weight. She did not have cyanosis, but was mildly short of breath and was receiving oxygen *via* nasal cannulae. Chest auscultation revealed decreased bilateral basal breath sounds with expiratory crackles throughout the lung fields, and a systolic ejection murmur radiating to the left axilla. There were no signs of right heart failure or other significant clinical findings.

Hematological investigations showed normal electrolytes, a hemoglobin of 14.1 g/dl, a hematocrit of 42.2%, and platelets of 105,000/mm³. The prothrombin time was slightly prolonged at 11 s (normal range 8-10 s) and the partial thromboplastin time at 30 s was in the upper end of the normal range of 22-32 s. The bleeding time was 11.5 min and, when repeated, greater than 18 min (normal range 5-10 min). Arterial blood gases breathing spontaneously with 2.0 l/min of nasal oxygen were pH 7.39, PaCO₂ 55 mmHg, and PaO₂ 73 mmHg. The aminophylline level was within the therapeutic range. Lung function tests had been performed virtually every month since the lung biopsy, when a diagnosis of lymphangiomyomatosis had been made. The results prior to admission were forced expired volume in 1 s (FEV₁) 1.01, 34% of predicted; forced vital capacity (FVC) 1.25 l, 32% of predicted; FEV₁/FVC 81% and the best diffusional lung capacity for carbon monoxide (DLCO) 46% of predicted. Measured DLCO values ranged from 2-14.5 ml · min⁻¹ · mmHg⁻¹ using a helium and carbon monoxide single-breath technique. These lung function test results were little changed over the previous 3 months.

The left ventricular function was measured by cardiac blood pool imaging using labelled red blood cells, and gave an ejection fraction of 65%, a cardiac output of 7.6 l/min, and a cardiac index of 4.3 l · min⁻¹ · m².

The electrocardiogram showed a sinus tachycardia and mild left ventricular hypertrophy. Chest radiography showed diffuse nonspecific infiltration and fibrotic changes at both bases. There was no evidence of cystic or bullous formation and no effusions or pneumothoraces. There had been no significant change over the previous 6 months.

In view of the coagulopathy, epidural anesthesia was deemed to be contraindicated. After discussion with all concerned parties, the patient consented to general anesthesia with endotracheal intubation and was advised to expect postoperative ventilation on the intensive care unit. The patient was taken to the operating room receiving oxygen *via* the nasal cannulae. A 20-gauge right radial arterial line and a 16-gauge intravenous cannula were inserted. Monitors attached to the patient included a pulse oximeter, an end-tidal capnograph, an electrocardiograph, and a noninvasive blood pressure machine.

Following breathing 100% oxygen for 5 min, a rapid sequence induction of anesthesia using iv thiopental and succinylcholine was performed. Anesthesia was maintained with nitrous oxide and oxygen, with an FI_{O₂} of 0.5, and the oxygen saturation remained greater than 95% at all times after induction of anesthesia. Isoflurane and fentanyl, 250 µg iv, were added when indicated by increases in systolic blood pressure and heart rate. Intermittant positive pressure ventilation, facilitated by neuromuscular blockade with atracurium, to a PaCO₂ of approximately 55 mmHg, as measured by intraoperative arterial blood gas analysis and end-tidal capnography, was maintained over the 3-h operating time.

Surgery proceeded uneventfully despite a 750-ml blood loss. Anesthesia was uncomplicated, apart from a sinus tachycardia of 100 bpm

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that responded to 2 units of packed red blood cells and 1500 ml of crystalloid. There was no other difficulty in maintaining oxygenation or "normocapnia" and cardiovascular stability.

At the end of surgery, the patient was allowed to awaken, and any remaining neuromuscular blockade was reversed with iv neostigmine and glycopyrrolate. Despite full return of neuromuscular function, she was making inadequate respiratory efforts. Ventilation was controlled in the recovery room with a FI_{O_2} of 1.0. Hemodynamic stability was maintained without additional intravenous fluids or inotropic support, and ventilation was continued to ensure adequate oxygenation. Endotracheal suction for the removal of secretions produced copious fresh blood. This was probably caused by iatrogenic damage to the lining of the trachea. The association of smooth muscle hypertrophy with a degree of infection, the inflammatory process, and a bleeding dyscrasia makes the tracheal lining friable. Confirmation would have risked further trauma and respiratory compromise. Attempts to extubate the trachea were abandoned. She was discharged from the recovery area to the intensive care unit with the trachea intubated and ventilation controlled.

Six hours postoperatively, hemodynamic instability became apparent and hematological investigations confirmed hemorrhage and a coagulopathy. The patient returned to the operating room. Her trachea was already intubated; ventilation controlled; and arterial, intravenous, and, now, central venous pressure lines in position. Cardiovascular resuscitation was achieved with 6 units of whole blood and 4 units of fresh frozen plasma. Anesthesia was maintained with atracurium, fentanyl, and isoflurane. During an exploratory laparotomy, 3 l of blood and ascites were drained from the abdomen, and hemostasis was obtained by control of numerous and diffuse bleeding sites in the pelvic cavity.

Postoperatively, the patient was returned to the intensive care unit with ventilation controlled. She was aggressively treated with antibiotics, bronchodilators, and physical therapy, so that she was weaned from the ventilator over the next 5 days without serious complication, and discharged to the floor.

Ten days postoperatively, the patient was discharged home and was reported to have been making a good recovery from her surgery.

One month later, she presented herself at the Emergency Room in acute respiratory failure. Despite ventilatory support, she died.

DISCUSSION

Although lymphangiomyomatosis is a rare disease, patients will present for surgery more frequently as the etiology and treatment by hormonal manipulations are better understood. They are almost certain to have a severely compromised respiratory system and may have other system disorders that will affect the choice of anesthetic technique.

The pathological process whereby there is a proliferation of immature smooth muscle throughout the peribronchial, perivascular, and perilymphatic regions of the lungs results in a progressive restrictive and obstructive pulmonary disease with a reduction in the diffusional capacity at the alveola: capillary interface causing death, on average, within 4 yr.³

The etiology of lymphangiomyomatosis remains unknown. Traumatic rupture of the thoracic duct, neoplastic proliferation of lymphangiocytes, hamartomatous malformation, hormonally stimulated hamartoma,⁷ or a

forme fruste of tuberous sclerosis^{8,9} have all been suggested. There are marked clinical, histological, and hormonal similarities between lymphangiomyomatosis and benign metastasising leiomyomas.⁶

The exclusive female sex distribution suggests steroid hormone metabolism involvement in the pathological process.^{1,4} It has been postulated that the increase in the incidence of the disease may be related to the widespread use of estrogens, particularly in the 1960s.¹⁰ Hormone receptor assay in lung tissue and then hormonal manipulation, whether medically or surgically, have become the mainstay of management.

Oophorectomy improves clinically and radiologically advanced disease.⁵ Controversy still exists as to the significance of estrogen and progesterone receptors and their antibodies.¹¹ The clinical presentations of lymphangiomyomatosis are increasing dyspnea, hemoptysis, recurrent pneumothoraces, chylous pleural effusions, and debilitating ascites. Chest radiography demonstrates a fine reticulonodular interstitial pulmonary infiltrate and, in later stages of the disease, cystic or honeycombing effects and bullous formation. Pulmonary function tests show a combined restrictive and obstructive picture and a diminished diffusional capacity as measured with carbon monoxide breathing tests.^{3,4} It is those patients with advanced disease and severe respiratory impairment that present for oophorectomy, under general or regional anaesthetic techniques. Our patient presented with manifestations of severe pulmonary disease, tachypnea, hypoxia and hypercapnia, and radiological evidence of interstitial fibrosis and basilar collapse. There is a good correlation between the severity of the clinical and radiological features and the pathological process.¹

In the described case, serial lung volume measurements over a number of years demonstrated a worsening disease process, becoming critical in the last 12 months. The diffusing capacity measurements using a single breath technique with carbon monoxide appears to be of limited predictive value, especially if the FVC is less than 2 l.¹³

The choice of anesthetic technique in our patient is complicated further by her coagulopathy and thrombocytopenia. We were also concerned about inadequate respiratory effort in a compromised patient positioned for a prolonged period of time in lithotomy and trendelenburg positions with an epidural or spinal anesthetic. We feel the advantages of an epidural technique intraoperatively and its use postoperatively for pain relief are outweighed by the risk of an epidural hematoma in patients with coagulopathies. It is for these reasons that general endotracheal intubation anesthesia was selected in preference to a regional technique. This was acceptable to the patient even in the knowledge that she had been ventilated 3 days postoperatively following the open lung bi-

opsy and that she would probably be ventilated postoperatively again.

General anesthesia was induced and maintained by drugs with a limited duration of action in the hope that, at the completion of surgery, neuromuscular function would return and respiratory depression due to anesthetic agents would be minimized. We felt that every opportunity to re-establish spontaneous ventilation should be taken.

We believe that early extubation after the first operation was prevented by traumatic endobronchial suction and not by the anesthetics. Subsequent events resulting in exploration of the abdomen dictated the need for prolonged mechanical ventilation.

Central venous or pulmonary artery pressures were not monitored during the first anesthetic. The patient had good myocardial function as measured by the ejection fraction and the cardiac index, and the predicted blood losses and fluid shifts were not large. The possible major complication of such invasive monitoring would include a hemothorax, a serious event in patients with limited respiratory reserve. When hemodynamic instability became apparent postoperatively, a central venous pressure line was indicated, and inserted.

In summary, we have described the anesthetic management of a patient with severe pulmonary disease, due to lymphangiomyomatosis, and a coagulopathy presenting for a vaginal hysterectomy and bilateral salpingo-oophorectomy.

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Airway Management for Unilateral Lung Lavage in Children

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Bronchopulmonary lavage (BPL) has been employed since 1965¹ for treatment of alveolar filling disorders such as alveolar proteinosis or extensive bronchial obstruction such as severe asthma. Because of the lack of double-lumen

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endotracheal tubes of appropriate size, it has been difficult to perform such procedures in children. We report a simple and effective technique for unilateral lung lavage in children.

CASE REPORT

A 7-yr-old, 25-kg psychotic boy was admitted for evaluation of bilateral pneumonia and respiratory distress. Flexible bronchoscopy revealed large quantities of oil in his bronchi, which upon analysis proved to be olive oil. Because of respiratory distress and hypoxemia (oxygen saturation by pulse oximetry 84% on room air), we elected to perform large-volume BPL. This child was uncooperative, and general anesthesia was considered essential for the lavage. His size precluded use of a double-lumen endotracheal tube, and an alternate plan for airway management was needed.

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