Anaesthetic management for oophorectomy in pulmonary lymphangiomyomatosis

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Pulmonary lymphangiomyomatosis is an idiopathic disease, resulting in severe respiratory impairment. Bilateral oophorectomy has led to objective and subjective amelioration of the pulmonary pathology. In the anaesthetic management of such a patient, careful attention must be paid to pulmonary and systemic haemodynamics, and gas exchange. We describe the successful anaesthetic management of a 34-year-old female, using epidural anaesthesia, and pulmonary artery catheterization. Although the intraoperative and immediate postoperative courses were heralded by marked cardiorespiratory stability, refractory respiratory failure developed, and she died five months after surgery.

Pulmonary lymphangiomyomatosis is a debilitating disease occurring exclusively in women of childbearing age. Fewer than 100 cases have been reported in the English language literature.

Pathologically, there is a proliferation of immature smooth muscle throughout the peribronchial, perivascular and perilymphatic regions of the lung and retroperitoneum.[1]

Patients present with increasing dyspnoea, haemoptysis, recurrent pneumothoraces and chylous effusion. Chest radiographs demonstrate a fine reticulonodular pattern which progresses to cystic changes and "honeycombing." Pulmonary function studies reveal a mixed obstructive and restrictive defect and a grossly diminished diffusing capacity of carbon monoxide (Dco). Survival greater than ten years after diagnosis is rare.

Although idiopathic, the uniqueness of the patient population suggests a hormonal aetiology.[2] To this end, measurements of estrogen and progesterone receptors in affected lung tissue have been made and medical and surgical hormonal manipulations attempted. These include therapy with progesterone, androgens, tamoxifen (an estrogen receptor antagonist) and bilateral oophorectomy.[2-4]

We describe the anaesthetic management for oophorectomy in a patient with pulmonary lymphangiomyomatosis.

Key words

ANAESTHETIC TECHNIQUES: epidural, epidural narcotics; PULMONARY FUNCTION; LUNG: pulmonary lymphangiomatosis.

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Case report

The patient, a 34-year-old white female, had presented three years previously with increasing shortness of breath of one year's duration. Physical examination and chest *x*-ray were unremarkable at that time but dyspnoea was incapacitating with minor activity. Pulmonary function tests (Table I) showed a mixed obstructive and restrictive defect and a severely depressed Dco. Arterial blood gases while breathing room air were within normal limits (Table II).

An open lung biopsy revealed pulmonary lymphangiomyomatosis. No tissue was submitted for receptor assay.

Despite treatment with theophylline, salbutamol inhalations, prednisone, progesterone and tamoxifen, her pulmonary status continued to deteriorate. PaO_2 on room air had fallen to 49 mmHg at rest and home oxygen therapy had become a requisite of day-to-day life. Bilateral oophorectomy was offered to the patient as a last resort.

The patient was admitted to the Surgical Intensive Care Unit for preoperative evaluation and preparation. Physical examination revealed a cachectic female, mildly short of breath at rest while receiving oxygen by "nasal prongs" at 4 $L \cdot min^{-1}$. Preoperative weight was 38.6 kg and height 157 cm. Breath sounds were diminished bilaterally with fine crackles heard at both bases. Heart sounds were normal and there was no evidence of right heart failure. Haemoglobin was 128 $g \cdot L^{-1}$; electrolytes, renal and liver function tests and coagulation studies were within normal limits. Chest *x*-ray showed bilateral apical bullae and basal nodularity. An electrocardiogram was normal.

TABLE I Pulmonary function tests results

Time	Predicted	3 yrs preop	2 yrs preop	1 yr preop	SICU admission	3 days postop	2 months postop
FEV_1 (L)	2.72	1.42	0.82	0.68	0.47	0.45	0.67
FVC (L)	3.36	2.72	1.61	1.56	1.57	1.35	1.53
TLC (L)	4.8	4.85	5.0	4.34	3.82	3.08	NA
RV(L)	1.55	2.13	3.2	2.7	2.34	1.61	NA
FRC (L)	2.4	2.78	4.1	3.14	2.66	2.2	NA
Dco	23	3.34	1.45	NA	3.03	1.69	NA

A pulmonary artery catheter was inserted via the right median basilic vein and a #20 cannula into the right radial artery. Baseline haemodynamics and blood gases are presented in Tables II and III; cardiac output was measured in triplicate by the thermodilution technique. A slight degree of pulmonary hypertension was demonstrated and was not altered by increasing the FiO_2. Marked shortness of breath precluded studies being done with the patient breathing room air.

She was taken to the Operating Room after receiving 100 mg of intravenous hydrocortisone. Five hundred ml of lactated Ringer's solution was given, and an epidural catheter was inserted at the L_2-L_3 interspace. A test dose of 3 ml of bupivacaine 0.5 per cent with 1:200,000 epinephrine was injected without incident. Over the ensuing 45 minutes, a total of 8 ml of three per cent 2-chloroprocaine and 10 ml of 0.5 per cent bupivacaine were carefully titrated to achieve a T_6 level of sensory blockade. It was hoped that the combination of drugs would allow a reasonably rapid achievement, as well as an adequate maintenance of the desired level of anaesthesia.

Surgery was performed uneventfully. Five mg of diazepam, 5 mg of prophylactic ephedrine and a further 500 ml of lactated Ringer's solution were given intravenously. The patient had no subjective worsening of her respiratory status intraoperatively, nor any significant changes in blood gases and haemodynamics. (Tables II and III).

Postoperatively, in the SICU, haemodynamics and blood gases remained unchanged (Tables II and III). Epidural morphine 5 mg every 10 to 12 hours was used to control abdominal pain and no systemic analgesics were needed. She was discharged to the ward three days later and home two weeks postoperatively. Despite aggressive nutritional support and intramuscular progesterone, her respiratory status continued to deteriorate and she died five months after surgery. Post mortem examination revealed pathological changes in the lung consistent with pulmonary lymphangiomyomatosis, a chylous right pleural effusion and no evidence of cardiac enlargement.

Discussion

A patient with advanced pulmonary lymphangiomyomatosis presents a major challenge to the anaesthetist and intensivist. Respiratory impairment is generally far advanced by the time oophorectomy is contemplated. This is manifested by marked hypoxaemia, bullous lung disease with a recurrent pneumothoraces, chylous pleural effusions and ascites. Work of breathing is excessive and patients are frequently malnourished. As seen with other forms of chronic lung disease, these patients are prone to frequent upper and lower respiratory tract infections.

The cardiovascular system is generally intact. Pulmonary artery pressure and pulmonary vascular resistance were mildly elevated in our patient and did not change while breathing more enriched concentrations of oxygen. This was undoubtedly due to perivascular smooth muscle proliferation,[1] and the effects of chronic hypoxaemia.

TABLE II Arterial blood gas results

Time	3 yrs preop	2 yrs preop	6 mos preop	SICU admission	SICU admission	Post-epidural	2 hrs postop	2 hrs postop	3 days postop
FiO_2	0.21	0.21	0.21	$4LO_2$-NP	$8LO_2$ NRBM	$8LO_2$ NRBM	$4LO_2$-NP	$8LO_2$ NRBM	$4LO_2$-NP
PO_2 (mmHg)	88	66	49	77	234	212	73	188	62
PCO_2 (mmHg)	33	31	30	39	42	41	46	49	44
pH	NA	NA	NA	7.41	7.42	7.40	7.35	7.33	7.39
HCO_3 ($mEq \cdot L^{-1}$)	NA	NA	NA	25	27	25	25	26	27
BE	NA	NA	NA	+1	+3	+1	−1	−1	+2

NP = nasal prongs
NRBM = non rebreathing mask

TABLE III Haemodynamic results

Time	*SICU admission*	*Postepidural*	*2 hrs postop*	*12 hrs postop*
Arterial pressure (mmHg)	120/54	110/55	104/52	110/50
Pulmonary arterial pressure (mmHg)	37/5	38/5	40/12	42/13
RA pressure (mmHg)	5	5	14	14
Wedge pressure (mmHg)	7	6	16	16
Cardiac output ($L \cdot min^{-1}$)	4.2	4.33	4.31	4.3
SVR (dyne sec cm^{-5})	1352	1219	1021	1021
PVR (dyne sec cm^{-5})	286	277	190	190
Hg ($g \cdot L^{-1}$)	128	116	118	116

We chose epidural anaesthesia for several reasons: (1) The patient had required a period of postoperative mechanical ventilation for several days after an open lung biopsy had been performed several years earlier. She had found this especially traumatic and specifically requested some form of regional anaesthesia. (2) Because of her apical bullae, we hoped to avoid positive pressure ventilation and the increased risk of pneumothorax. (3) Use of an epidural as opposed to a spinal anaesthetic allowed careful titration and avoidance of a high motor block which would have been catastrophic to someone with such limited respiratory reserve. (4) We were able to use epidural morphine for postoperative analgesia. Shulman *et al.*[5] found a more rapid return to baseline pulmonary function in post-thoracotomy patients given epidural as opposed to parenteral morphine. Pulmonary function testing on the third day postoperative showed a slight worsening of our patient's restrictive defect, with the functional residual capacity decreasing from 2.6 litres to 2.2 litres and the PaO_2 while receiving oxygen by "nasal prongs" at $4\ L \cdot min^{-1}$, falling from 77 mmHg to 62 mmHg (Tables I and II). This would be in keeping with the postoperative changes in pulmonary function rviewed by Craig.[6]

We felt pulmonary artery catheterization was a valuable adjunct. It demonstrated a slight degree of fixed pulmonary hypertension and enabled us to monitor this and other haemodynamic variables in the perioperative period.

In summary, we described the successful management of a patient with pulmonary lymphangiomyomatosis presenting for oophorectomy. Cardiorespiratory stability was maintained using invasive haemodynamic monitoring and epidural anaesthesia.

References

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Résumé

La lymphangiomyomatose pulmonaire est une maladie idiopathique, entraînant un trouble respiratoire grave. La pathologie pulmonaire s'est améliorée de façon objective et subjective suite à une ovariectomie bilatérale. On doit porter une attention particulière à l'hémodynamique pulmonaire et systémique et à l'échange gazeux dans la conduite anesthésique chez ce genre de patiente. Nous décrivons une conduite anesthésique réussie che une patiente de 34 ans, où l'on a utilisé une anesthésie péridurale et un cathétérisme de l'artère pulmonaire. Bien qu'une stabilité cardiorespiratoire marquée se soit manifestée en période intraopératoire et en période postopératoire immédiate, une défaillance respiratoire réfractaire s'est développée et la patiente est décédée cinq mois après la chirurgie.