Case Report

Anaesthetic challenges in pulmonary alveolar microlithiasis – a rare disease treated with bilateral lung transplantation, first case from India

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ABSTRACT

Pulmonary Alveolar Microlithiasis (PAM) is a rare autosomal recessive end stage lung condition characterized by widespread alveolar deposition of calcium microliths. It is worth emphasis india has reported 80 cases out of 1022 cases worldwide but there is no report on lung transplant and and anaesthetic management in this category of patient. This report present the anaesthetic challenge in management of first indian patient with the diagnosis of PAM, who underwent bilateral lung transplantation. Bilateral lung transplantation is one of the most challenging surgeries that require the care of a cardiothoracic anesthesiologist. Utilization of extracorporeal circulation has allowed a safer performance of this procedure in patients with severe cardiopulmonary compromise. Intraoperative management is a pivotal part of the patient's care, as it contributes to the patient's overall outcome.

Keywords: Anaesthetic management, bilateral lung transplant, extracorporeal membrane oxygenation, pulmonary alveolar microlithiasis, transesophageal echocardiography

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INTRODUCTION

Pulmonary Alveolar Microlithiasis (PAM) is a rare autosomal recessive end stage lung condition characterized by widespread alveolar deposition of calcium microliths.[1] it is worth emphasis India has reported 80 cases out of 1022 cases worldwide but there is no report on lung transplant and anesthetic management in this category of patient.[1] This report present the anesthetic challenge in management of first Indian patient with the diagnosis of PAM, who underwent bilateral lung transplantation with extracorporeal membrane oxygenation (ECMO) support. Transesophageal echocardiography (TEE) plays an important role in evaluation and decision making during the surgery.

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CASE REPORT

A 54-year-old female patient presented with progressive dyspnoea eventually becoming oxygen and NIV dependent. Her past history is significant for hypothyroidism and systemic hypertension on regular medications. On physical examination there was mild respiratory distress with peripheral cyanosis, marked clubbing and bilateral fixed inspiratory crackles. On presentation her Arterial Blood Gas (ABG) was suggestive of severe hypoxemia with room air pO_2 of 47.2 mmHg, and SpO_2 of 82.8%.

Chest X ray showed 'white out lungs' with opacification of bilateral lung parenchyma also obscuring all mediastinal

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details [Figure 1a]. HRCT chest revealed diffuse dense calcification of lung parenchyma including the subpleural, peribronchovascular and centrilobular distribution [Figures 1b, 2a and b]. Thus, a diagnosis of pulmonary alveolar microlithiasis was established. Pulmonary Function Test (PFT) suggested restrictive lung defect. ECHO demonstrated Good biventricular systolic function and severe Pulmonary Hypertension (PASP 80 mm Hg). Ventilation perfusion scan R > L. Patient was put on a program for pulmonary rehabilitation and optimization of general condition and underwent bilateral lung transplantation with a size and ABO matched donor.

Following appropriate donor selection and communication with the procurement team at the donor site, patient prepared, and baseline investigations sent, 20-gauge radial artery and a large bore 16-gauge cannula placed. Anesthesia was induced gradually to avoid any sudden decrease in systemic pressure or increase in PVR with intravenous (iv) injection etomidate 0.3 mg/kg and iv injection fentanyl 5 mcg/kg. Muscle relaxation was achieved with iv injection rocuronium 0.6 mg/kg. The patient was intubated with left sided double lumen endotracheal tube (DLT) that is positioned using fiberoptic bronchoscopy. Anesthesia was maintained with oxygen, air, isoflurane, and fentanyl. A left femoral arterial line is placed. Venous access is established in the right neck and left groin. If the patient is high risk or the donor lungs of marginal quality, it is prudent for the team to place the right venous neck line in the left neck in the event that post-operative extracorporeal membrane oxygenation (ECMO) may be required (the right neck would be used for a cannula during veno-venous ECMO). Placement of a pulmonary artery (PA) catheter is performed. A transesophageal echo (TEE) probe is placed in the esophagus and routine evaluation performed.

The patient is positioned supine with arms abducted, supported, and padded above the head to expose both the chest and the axillary regions. The incision used for bilateral lung transplantation was the clamshell incision.

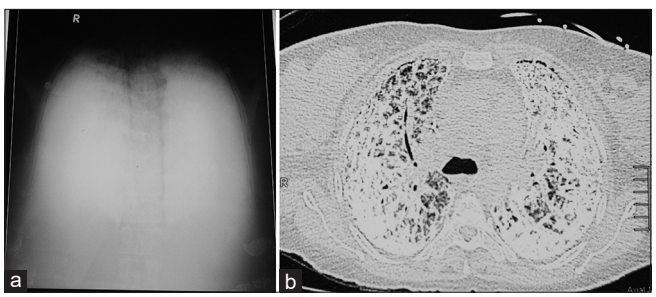

Figure 1: (a) Chest X Ray showing "white out lung". (b) CT showing dense calcific fibrosis and subpleural cyst

Before stapling the PA, it was snared down using a tourniquet for 5–10 minutes to assess hemodynamic stability. In the event of escalating PA pressure, the decision to use ECMO/cardiopulmonary support should be made. This patient had severe PAH and after PA clamp, PA pressure rose to suprasystemic inspite of nitric oxide, so we decided to use VA ECMO. Central VA ECMO was established after adequate heparinization, and right and left pneumonectomy was performed. After anastomosis care was taken to maintain the mean PA pressure not to exceed 20 mmHg for 10 min to facilitate controlled pressure reperfusion conditions. The lung was ventilated on 40% oxygen before the weaning off from VA ECMO. Following the procedure, she was shifted to CT Isolation ICU with nitric oxide, inotropes, and mechanical ventilator support. Post-operative sedation and analgesia maintained with inj fentanyl, inj dexmedetomidine, and inj paracetamol.

Bronchoscopy and Echo was done at bedside in periodic intervals. She was extubated, Inotropes weaned off and RT feeds started on 2nd POD. Triple regimen immunosuppressants were started on 1st POD and her renal parameters were kept under close observation. She was mobilized on 3rd POD. Her airway was supported with intermittent BiPAP following next 3 days. She was discharged from ICU POD 7.

DISCUSSION

Pulmonary alveolar microlithiasis is a rare autosomal recessive disease characterized by a intra-alveolar accumulation of calcium calculi or microliths. The first case of PAM was reported by Harbitz in 1918 hence, the name Harbitz' syndrome. PAM is characterized by Clinic-Radiologic dissociation. Typically, asymptomatic patients present with diffuse fine sand like infiltrates on chest radiograph called "sandstorm lung". Bronchoscopic Trans Bronchial Lung Biopsy (TBLB) and Broncho alveolar Lavage (BAL) help establish definitive diagnosis of PAM. No definite treatment other than lung transplantation has been proven to change the course of disease.[1,2]

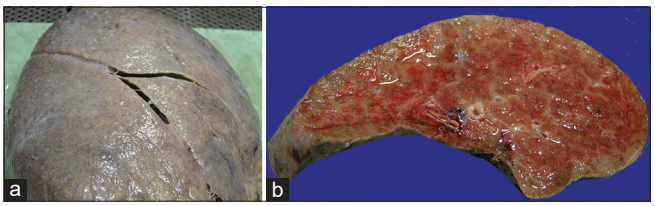

Figure 2: (a) Lung explant with granular external surface. (b) Cut surface with granular appearance

Our patients was NIV dependent with end-stage lung disease (PAM) with severe PAH. Induction is a crucial part of the surgery. Etomidate seems to be preferred in cases of existing hemodynamic compromise as it is described to offer more hemodynamic stability.[3] Opioids blunt the sympathetic response to laryngoscopy and may prevent increases in PVR during intubation. Inhalation agents (isoflurane, sevoflurane) provide bronchodilation but may also cause vasodilation and myocardial depression and should be introduced slowly. TEE monitoring allows the anesthesiologist to detect changes in right heart function and size, worsening of tricuspid regurgitation and filling status of the left ventricle. This information becomes important in deciding if extracorporeal life support (ECLS) is necessary, like this case we had to go on VA ECMO because of supra systemic PA pressure.[4] Obstruction of the pulmonary vein flow (due to clot, kinking or narrow anastomosis) may present with hypoxia.[5] TEE becomes a significant tool in differentiating this condition from other causes such as acute graft rejection or reperfusion injury. Turbulence of flow, pulmonary vein diameter of <0.5 cm, peak systolic flow velocity (PSFV) >1 m/s and pulmonary vein-left atrial pressure gradient (PVLAG) ≥10–12 mmHg support the diagnosis of pulmonary vein stenosis.[5] In general, large volumes of fluids should be avoided as the donor lung is susceptible to edema.[6] Vasopressors and inotropes are used to counteract the hemodynamic effects of anesthetics and positive pressure ventilation. Patients with restrictive lung disease may require higher respiratory rate and higher airway pressures to generate adequate tidal volumes, as they have low compliance. PEEP helps recruit collapsed alveoli and increases the FRC. I: E ratio closer to 1:1 may minimize peak inspiratory pressures. Minimization of V/Q mismatch throughout the lungs is key.[7] Utilization of nitric oxide and inhaled iloprost may improve the V/Q by selectively dilating the pulmonary vessels only in the ventilated areas.[8] If PaO2 continues to decline, $PaCO_2$ to increase and the patient becomes severely acidotic despite all maneuvers, CPB or ECMO should be initiated.[4]

Lung reperfusion is a critical step during lung transplantation. Blood flow is slowly (over 10-15 min) introduced to the new lung.[9] The transplanted lung is gently ventilated with low FiO_2 (close to room air), initially barely inflated with a slow respiratory rate (RR). Gradually the tidal volume, RR, and PEEP are increased to the appropriate levels based on lung compliance. According to protective ventilation strategies double lung ventilation with 6–7 ml/kg, predicted body weight of donor, plus PEEP protects against lung injury. FiO_2 40% is desirable to avoid risk of free radical-induced oxygen toxicity.[7,9]

Primary graft dysfunction (PGD) after lung transplantation occurs in the early period following reperfusion of the allograft with incidence reported between 10% and 57%.[10]

The denervated lung has a decreased ventilatory response to hypercapnia and decreased cough reflex resulting in decreased pulmonary clearance requires frequent bronchoscopy. Close postoperative care, adjustments in ventilation setting, triple therapy of immunosuppressants, hemodynamic support, fluid status, and pain control is crucial.

CONCLUSION

PAM remains a challenging disease worldwide. Bilateral lung transplantation is one of the most challenging surgeries that require the care of a cardiothoracic anesthesiologist. The complexity of the procedure demands that all teams are in communication with each other at all times. Utilization of extracorporeal circulation has allowed a safer performance of this procedure in patients with severe cardiopulmonary compromise. Intraoperative management is a pivotal part of the patient's care, as it contributes to the patient's overall outcome.

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Conflicts of interest
There are no conflicts of interest.

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