Case Report

Bilateral Whole Lung Lavage by Lung Isolation in a Child with Pulmonary Alveolar Proteinosis: A New Technique

Brajesh Kaushal, Sandeep Chauhan, Suruchi Hasija

Department of Cardiac Anaesthesia, Cardiothoracic Centre, AIIMS, New Delhi, India

ABSTRACT

We present two rare cases of children who presented with progressive exertional breathlessness and dry cough. They also had history of bluish discoloration of mucous membranes, hands and feet on exertion. Both were diagnosed to have pulmonary alveolar proteinosis after a high-resolution computed tomography and bronchoalveolar lavage. They were subjected to bilateral whole lung lavage (WLL) as a salvage therapy. Bilateral WLL was performed in a single sitting with the help of a modified endotracheal tube. The anesthetic technique included a modified cuffed endotracheal tube for accomplishing WLL. After the procedure, both children improved clinically and functionally.

Keywords: Bilateral whole lung lavage, modified endotracheal tube, pulmonary alveolar proteinosis

Address for correspondence: Dr. Suruchi Hasija, Department of Cardiac Anaesthesia, 7th Floor, Cardiothoracic Centre, All India Institute of Medical Sciences, New Delhi - 110 029, India.
E-mail: suruchi_hasija@hotmail.com
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INTRODUCTION

Pulmonary alveolar proteinosis (PAP) is an uncommon disease characterized by intra-alveolar accumulation of surfactant proteins and lipids, impeding gas exchange leading to hypoxia and cyanosis.[1] Patients with extensive disease have an increased alveolar/arterial pressure difference which increases further after exertion.[2] Whole lung lavage (WLL) remains the gold standard for treating PAP. Recently, granulocyte-macrophage colony-stimulating factor (GM-CSF) replacement therapy has been suggested as the first-line therapy for PAP.[3] WLL is a cumbersome procedure that may require venovenous extracorporeal membrane oxygenation (V-V ECMO) support. Improved technology and experience in cardiopulmonary anaesthesia have made this procedure relatively simple and safe. Various techniques have been described in the literature regarding WLL in PAP.

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WLL procedure has been performed in the left lung while ventilating the right lung using a double-lumen tube, and on right lung 9 days later.[4,5] Paquet and Karsli[6] performed WLL in a 2-year-old child with PAP using an airway assembly consisting of two cuffed tracheal tubes (3.0 and 3.5 mm ID) and the angled and Y-connectors from a standard double-lumen tube. The 3.5-mm ID tube was inserted in the left bronchus (bronchial tube), and the 3.0-mm ID cuffed tube was placed in the trachea [endotracheal (ET) tube]. They first performed left lung lavage followed by right lung lavage 2 days after the first procedure.

CASE REPORT

Case 1

A 3.5-year-old male child (height 87.6 cm, weight 8.6 kg) presented in the paediatric outpatient department with the complaints of dry cough and progressive shortness

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of breath on mild exertion for a duration of 8 months. His mother also gave history of bluish discoloration of hands, feet and lips on exertion and crying. He also had history of failure to thrive and recurrent respiratory tract infections, but his clinical and radiological findings remained unchanged even after antibiotic treatment. His general physical examination revealed poor built and malnourishment. On auscultation, bilateral inspiratory crackles were present in the basal lung fields. Chest radiograph showed bilateral diffuse small pulmonary opacities reminiscent of a miliary pattern [Figure 1a]. His routine serological and biochemical investigations were within normal limits. His baseline oxygen saturation was 90% on room air, but he desaturated further on minimal exertion. High-resolution computerized tomography scan reveal thickening of inter- and intralobular septal lines with ground glass opacities [Figure 1c]. Bronchoscopy showed clear airways, and the effluent bronchial lavage fluid examination confirmed the diagnosis of PAP.

The child was posted for bilateral WLL under general anaesthesia, the most successful treatment described for PAP.[7] It necessitates modified airway assembly because doublelumen tubes are not available for children. The airway assembly was constructed by increasing the length of the ET tube by railroading one size bigger uncuffed tube over the proximal (connector) end of the cuffed ET tube. For the left lung, a 3.0-mm cuffed ET tube was lengthened by a 4.0-mm uncuffed ET tube railroaded over its proximal end. For the right side, a 3.5-mm cuffed tube was railroaded by a 4.5-mm uncuffed ET tube in the same manner [Figure 2a].

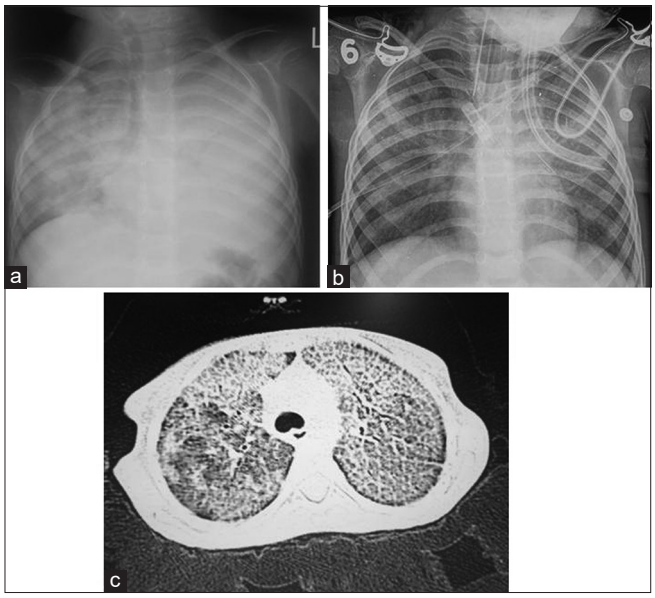

Figure 1: Chest X-ray of the child before whole lung lavage (WLL) (a), after WLL (b), and highresolution computerized tomography chest image before WLL (c)

This newly constructed airway assembly increased the working length of the ET tubes and enabled bilateral WLL in a single sitting with one ET tube used for lung lavage, while the other ET tube used for ventilating the opposite lung. Previous case reports used two cuffed ET tubes, but placed one ET tube in the bronchus and the other ET tube in the trachea, and the WLL procedure was performed separately on both the sides two days apart.

In this index case, bilateral WLL was performed in a single sitting, which was helpful to hasten recovery and curtail the cost of the procedure. The child was induced with intravenous ketamine (2 mg/kg), fentanyl (2 μg/kg), and cisatracurium (0.15 mg/kg), and the 3.5-mm modified cuffed ET tube was placed in the right bronchus; and after inflating the cuff, airway entry was checked. Thereafter, the 3.0-mm modified cuffed ET tube was passed through the glottis behind the previous ET tube and placed in the left bronchus and air entry was checked in the same manner [Figure 2b].

Both the tubes passed easily and without resistance, and the cuffs were inflated just enough to ensure proper lung isolation and adequate ventilation. Tube positioning was verified with a 2.2-mm OD flexible fiberoptic bronchoscope (Olympus, Center Valley, PA, USA). It was ensured that the patient could tolerate single lung ventilation with 100% oxygen. V-V ECMO personnel and hardware were on standby. Anaesthesia was maintained with intermittent fentanyl (1 μg/kg), cisatracurium (0.03 mg/kg) and isoflurane (1 MAC). Left lung lavage was performed with aliquots of 100–120 mL lukewarm saline. Chest clapping was performed during the procedure, and lavage returns were collected by gravity after each aliquot. The first sample was thick and milky [Figure 2c] but gradually cleared during the procedure. A total of 1500 mL of saline was instilled.

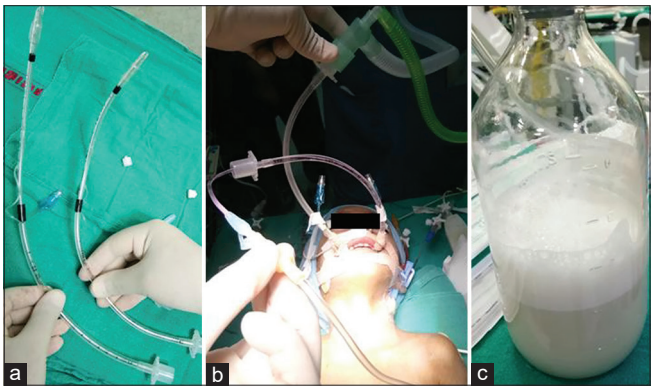

Figure 2: Modified endotracheal (ET) tube for lung isolation during whole lung lavage (WLL). The length of the ET tube was increased by connecting the proximal end of bigger size uncuffed ET tube over the distal end of smaller size cuffed ET tube (a). Patient with the modified ET tubes in situ during WLL procedure (b). Milky return lavage fluid (c)

The same procedure was repeated on the right lung while ventilating the left lung. After overnight ventilation in the intensive care unit, the patient was extubated uneventfully. After the procedure, the patient were better both clinically and radiologically [Figure 1b]. Two days later, the patient was able to maintain adequate oxygenation on room air and was discharged. On the first follow-up visit, the patient had shown significant clinical improvement.

Case 2

A 3-year-old male child weighing 8 kg was also admitted with similar complaints, and radiological and other investigational findings. He was also considered for bilateral WLL under general anaesthesia. The right bronchus was intubated with 3.0-mm modified cuffed ET tube and the left bronchus with 2.5-mm modified cuffed ET tube in the same manner. WLL was performed on the left side with aliquots of 60 mL lukewarm saline while ventilating the right lung until the return fluid was cleared. The right lung was then subjected to WLL in a similar fashion while ventilating the left lung.

DISCUSSION

PAP is a rare disease of unknown etiology with an estimated incidence of 0.36 per million.[8] The disease is classified as congenital, idiopathic (primary) and secondary. The majority (>90%) of cases fall into idiopathic category, which is considered to be an autoimmune disorder with development of anti-GM-CSF antibodies.[9] Patients with PAP are prone to infection.[10]

We constructed a simple and reliable airway assembly that can be used to provide lung isolation for lung lavage or other such procedures in small children in a single sitting. Bilateral WLL in a single sitting is beneficial for the patient in terms of early recovery and reduced cost. Essentially, two ET tubes were passed through the glottis, one endobronchially in the left side and the second in the right side. The more diseased left lung was lavaged first to improve gas exchange and limit desaturation during subsequent right lung lavage. The advantage of this technique was effective lung isolation, the ability to collect lavage returns by gravity or suction, the option of differential lung ventilation after lavage, and the ability to perform bilateral lavage as single sitting procedure. Fiberoptic bronchoscopy can be used to ensure continued lung isolation throughout the procedure. A possible disadvantage of this technique is the potential for vocal cord edema, stridor, or mucosal damage. To minimize this risk, the smallest sized tubes that allowed adequate ventilation and lavage were used. The tubes were inserted along the long axis of the glottic opening to minimize lateral stretch of the vocal cords. Our patients did not develop any hoarseness after the procedure. In addition, hydrocortisone was administered before the procedure. A major advantage of this approach for lung lavage in small children seems to be higher lavage returns when compared with other techniques. In conclusion, we report the successful use of two cuffed ET tubes (one left bronchial and one right bronchial) in small children (<12 years of age for whom commercial double-lumen tracheal tubes are not available) to perform bilateral WLL as a single procedure without the noteworthy complications. This way, V-V ECMO and its attendant complications could be averted.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient (s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initial s will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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