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CASO CLÍNICO

Anesthesia for Bronchoalveolar Lavage in Alveolar Proteinosis: A Case Report

Anestesia para Lavagem Pulmonar em Proteinose Alveolar: Caso Clínico

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Palavras-chave
Anestesia Geral; Lavagem Pulmonar; Proteinose Alveolar; Ventilação Unipulmonar

ABSTRACT

Pulmonary alveolar proteinosis is a rare disorder characterized by alveolar accumulation of lipoproteinaceous material secondary to abnormal processing of surfactant by macrophages. Its current mainstay treatment is whole lung lavage. We describe the complexity of the anesthetic management of a patient submitted to a single-lung lavage. Being an uncommon lung disease, the anesthetic approach presents a challenge for both pulmonologists and anesthesiologists. Reporting is thus necessary to share experience and promote discussion.

RESUMO

A proteinose alveolar é uma doença rara caracterizada pelo acúmulo alveolar de material lipoproteináceo secundário ao processamento anormal de surfactante pelos macrófagos. O tratamento *gold standard* consiste na lavagem pulmonar total. Descrevemos a complexidade do manuseio anestésico de um paciente com proteinose alveolar submetido a uma lavagem unipulmonar. Por ser uma patologia rara, a experiência na abordagem anestésica é um desafio, tanto para pneumologistas como para anestesiologistas, pelo que se torna primordial a partilha de experiências com promoção à sua discussão.

INTRODUCTION

Pulmonary alveolar proteinosis (PAP) is characterized by abnormal intra alveolar surfactant accumulation caused by a deficit in macrophage activity, which can be congenital, secondary, or acquired (the latter being the most common).[1-7] With an annual incidence and prevalence of 0.36 and 3.70 cases per million, respectively, it is difficult to accumulate significant clinical experience related to this disorder.[1,2,4,6,7] Typical patients include male smokers aged between 30 and 50 years, although PAP also arises in patients with coexisting autoimmune disorders.[1,2,6,7] Patients usually present progressive dyspnea associated with minimally productive cough or fatigue, weight loss and low-grade fever.[1-4,7] Physical examination is often normal[2] and biochemical abnormalities include elevated serum levels of lactate dehydrogenase (LDH) and other protein products of pulmonary epithelial cells.[1,7] The pulmonary function test (PFT) is consistent with restrictive lung disease, although it can be normal.[1-7]

The gold diagnostic standard is open-lung biopsy,[1,2,4,6] with diagnosis confirmed in approximately 75% of clinically suspected cases, by findings of a "milky" effluent from bronchoalveolar lavage.[1,2,4-6] The prognosis may vary from spontaneous resolution to rapid progression with respiratory insufficiency, with 5-year survival rates ranging from 75% to 95%.[2,4,6-7] Many therapies have been used to treat PAP, but whole lung lavage (WLL) remains the standard approach.[1-4,6,7] Although complex WLL is well tolerated, and can be associated with anesthetic intraoperative complications.[1]

Being an uncommon lung disease, the anesthetic approach presents a challenge for both pulmonologists and anesthesiologists.[8] For this reason, the presentation of case reports is relevant for practitioners to discuss and improve on best diagnostic and treatment practices.

CASE REPORT

A 46-year old man was diagnosed with acquired PAP by lung biopsy with video-assisted thoracoscopic surgery presented history of weight loss, dyspnea, cough and mucous yellow sputum in the previous four months. Chest X-ray showed bilateral interstitial and alveolar infiltrates (Fig. 1) and PFT a moderate restrictive ventilatory alteration with a moderated defect in diffusion (Table 1). Relevant history included professional exposure to paints and varnishes, past smoking history of 15 years (four units/pack per year), and treatment

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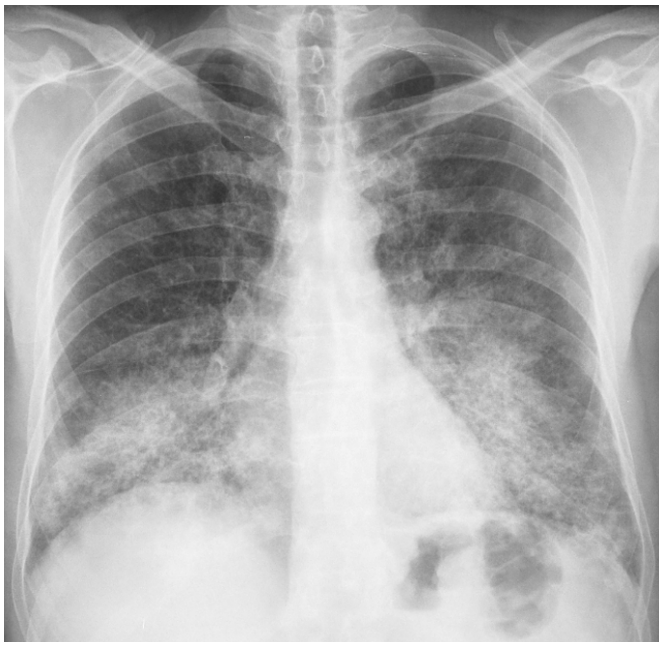

Figure 1. Pre right bronchioalveolar lavage chest X-ray showing bilateral interstitial and alveolar infiltrates

Table 1. Comparison of Pre and Post WLL Pulmonary Function Test

	Pre WLL	Post WLL
FEV1	1.97 L (60%)	2.01 L (62%)
FVC	2.57 L (64%)	2.87 L (72%)
Tiffeneau Index	90%	89%
DLCO	41%	42%
TLC	3.65 L (60%)	4.22 (69%)
FRC	2.17 L (68%)	3.01 L (95%)
RV	1.25 L (65%)	1.5 L (77%)

Legend:
WLL – whole lung lavage; **FEV1** - forced expiratory volume in first second; **FVC** - forced vital capacity; **FEV1/FVC** – ***Tiffeneau Index***; **DLCO** - diffusion capacity for carbon monoxide; **TLC** - total lung capacity; **FRC** – functional residual capacity; **RV** - residual volume; **L** – litres

with oxygen 1 L/min every night since the diagnostic of PAP. In addition to thoracoscopy, the patient had already been surgically submitted to nasal septoplasty. No usual medication and no other relevant medical history.

Given the clinical deterioration of the patient, a right WLL was proposed.

The patient was preoperatively evaluated by an anesthesiologist and classified as ASA III, with a functional capacity of 4 METS. The patient did not perform kinesiotherapy prior to WLL due to the short timespan between clinical assessment and the procedure. On arrival to the operating room, the patient was hemodynamically stable, breathing room air at rest with arterial oxygen saturation ($SatO_2$) of 97% in the supine position and arterial partial pressure of oxygen (PaO_2) of 67 mmHg. The patient was monitored as per ASA standards, including in addition bispectral index (BIS), invasive arterial pressure and urinary output monitoring. Before starting the procedure, active heating methods (patient and fluids) were used to prevent hypothermia. To prevent airway edema and bronchoreactivity induced by the procedure, hydrocortisone 100 mg and methylprednisolone 125 mg were administered prior to induction. After pre-oxygenation, a general anesthesia was induced with 100 mcg of fentanyl, 170 mg of propofol and 40 mg of rocuronium and maintained with sevoflurane (for a target BIS maintained between 40 and 60). Neuromuscular block was continuous monitored and rocuronium administered as needed (to keep train of four responses between 1 and 2). Lung separation was achieved by placing a 39F left double-lumen endobronchial tube into the left mainstem bronchus and its position was confirmed by auscultation and fiberoptic bronchoscopy. The cuff manometer was used to monitor bronchial cuff pressure throughout the procedure (pressure was kept near 30 cm H_2O) and a good sealing by fiberoptic bronchoscopy was confirmed at the beginning of the procedure. The patient was placed in the right lateral decubitus and the tube position was rechecked by auscultation. One-lung ventilation of the left lung began just before initiation of lavage of the right lung and protective ventilation was used (volumes 4 to 8 mL per Kg of ideal weight, positive end-expiratory pressure from 7 to 12 cm H_2O), being adjusted in alignment with observed compliance, $SatO_2$ and blood gases (Table 2). At the start of the lavage procedure, an infusion of 100 mL of saline solution at 1-minute intervals was instilled until reaching the functional residual capacity (FRC) of the lung to be washed. Afterwards, repeated cycles of instillation of 500 mL of 0.9% saline solution with 0.6 mmol of sodium bicarbonate at body temperature were administered, followed by passive drainage under gravity. During fluid inflow and outflow, airway pressure and tidal volume were monitored. Initially, milky fluid effluent was obtained (Fig. 2) and a total lavage of 14.8 L was necessary to get clear fluid effluent (Fig. 2). The procedure lasted approximately 3 hours and 20 minutes and $SatO_2$>95% with PaO_2>100 mmHg were maintained. At the end, in order to passively leave the volume corresponding to the FRC, drainage maneuvers were performed, mobilizing the patient to the right, left, Trendelenburg and proclive. Next, two-lung ventilation began and recruitment maneuvers were applied to restore expansion of both lungs. The double lumen endotracheal tube was exchanged for 8.0 mm single-lumen tube and the patient was transferred to the Post-Anesthesia Care Unit with ventilatory support under general intravenous anesthesia. After three hours, the patient was extubated and two hours later transferred to the pneumology ward. A chest X- ray as performed the day after the procedure showing improvement in the infiltrates of the right lung (Fig. 3). The patient started kinesiotherapy in the immediate post-operative period and continued after hospital discharge. Fifteen days later, the PFT was repeated, now compatible with a moderate obstructive ventilatory alteration positive for bronchodilation test and maintaining a moderated defect in diffusion (Table 1). The patient provided consent to the publication of this report.

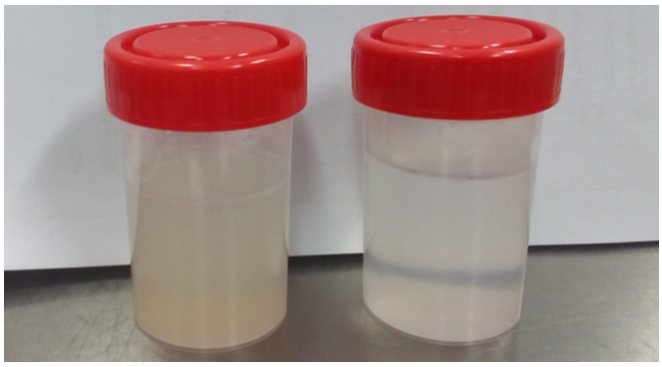

Figure 2. Alveolar lung lavage obtained from the patient, noticeably the aspirated liquid was progressively less cloudy (from left to right)

DISCUSSION

Currently, WLL is the standard treatment for PAP, being associated with an improvement in dyspnea and in objective parameters.[1-7]

The initial aspirated material has a milky appearance and foamy supernatant, revealing an important protein content (Fig. 2) that clears up during the procedure, obtaining the same aspect of the instilled material due to the removal of the lipoprotein material from the alveolar space (Fig. 2).[1,3,6]

Response to WLL and respective physiologic improvement includes increased forced vital capacity, total lung capacity, higher PaO_2 at rest and with exercise, and improved diffusing capacity of the lungs for carbon monoxide, with decreased alveolar arterial gradient and shunt fraction.[1] Anesthesia for WLL is challenging for many reasons, the primary being intraoperative refractoriness, which tends to be more common while the first lung is being lavaged. The anesthetic approach of this patient was performed safely and effectively. In this case, the patient was pre-oxygenated to avoid low oxygen saturation, maintaining $SatO_2$>95%. Furthermore, the use of one-lung ventilation and instillation of large volumes of fluid in the setting of pre-existing respiratory failure place the patient at risk of profound hypoxemia and of flooding of the ventilated lung.[5,7] The pathophysiology of hypoxemia is related to ventilation-perfusion mismatch in lung lavage: during the filling phase, perfusion of the non-ventilated lung is reduced by compression of the pulmonary vasculature and hence shunt is reduced; however during the drainage phase reperfusion of the non-ventilated lung increases shunt causing hypoxemia.[1,3,7] Although the positive end-expiratory pressure applied to the ventilated lung during the drainage phase may augment the shunt, its use is recommended because during the filling phase of the non-ventilated lung, the ventilated lung may improve oxygenation.[1,3]

Pre-oxygenation before induction is necessary to prevent desaturation.[1,3,7] In this case, after anesthetic induction was advocated to ventilated the patient for 20 minutes with a fraction of inspired oxygen (FiO_2) 100%, avoiding failure of denitrogenation of lungs that can leave nitrogen bubbles in the alveoli and limit the effectiveness of the lavage.[1,3,7]

The WLL is performed under general anesthesia, allowing

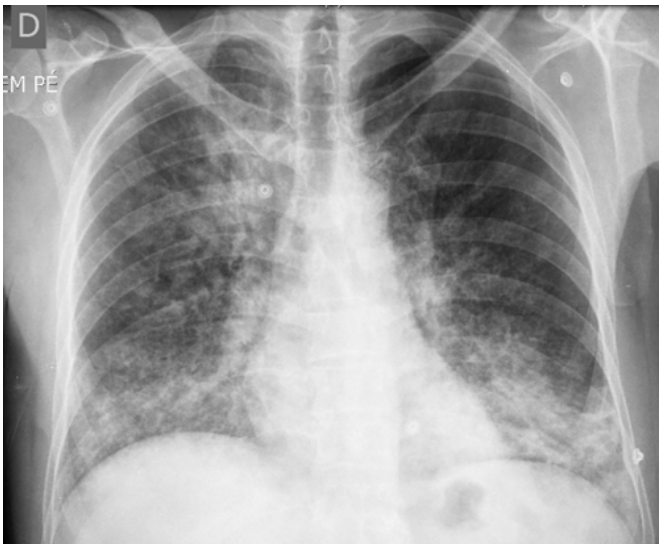

Figure 3. Pos right bronchioalveolar lavage chest X-ray showing improvement ground-glass opacification of the right lung

the two lungs to be separated by endobronchial intubation; one lung is ventilated while the other is flooded with warm saline to progressively wash out the alveolar space. We used a left-sided double-lumen endotracheal tube because the use of a right sided tube tends to block the orifice of the right upper lobe bronchus.[1,3,5,7] Despite being an anesthetic challenge, we used inhalation anesthesia instead of intravenous, taking advantage of the bronchodilator effect of sevoflurane. However, being an airway procedure, it leads to pollution and as such, in these situations, total intravenous anesthesia is one of the safest methods.[8] Traditionally, the procedure is conducted in lateral decubitus and carried out carefully to avoid endotracheal tube movement, as strict placement is essential to prevent lung flooding.[1,3,5,7] This position has the advantage to minimize blood flow to the non-ventilated lung, but maximizes the possibility of spillage.[1,3]

The most common adverse event associated with this procedure is hypoxemia, which can be minimized by increasing the concentration of O_2. However, given the patient good general condition, there was no need for significant increases in FiO_2. During drainage, a way to increase $SatO_2$ by applying high positive end expiratory pressure (PEEP) can induce an increase in pulmonary vascular resistance and hypoxia, so we must use adequate PEEP values guiding the patient's compliance, in order to prevent this complication.[8]

Other complications include pneumothorax, pleural effusion and hydropneumothorax.[1,6,7]Infusion of large volumes of saline in lungs is associated with mediastinal shift, increased intrathoracic pressure and central venous pressure, higher arterial oxygen tension and hypotension.

Increase in airway pressure or decrease in tidal volume may indicate reduced compliance of the ventilated lung and fluid leakage, so monitoring is necessary.[3] Moreover, monitoring to detect the loss of lung isolation can be done by: the appearance of bubbles in the lavage fluid draining from the lavage side, appearance of rales and rhonchus on the ventilated side and imbalance between the instilled and the drained volumes.[1]

Fiberoptic bronchoscopy inspection confirms if flooding of the ventilate lung has occurred.[3]

To restore pH to 7.2-7.4, 0.6 mmol/L of sodium bicarbonate were added to the saline solution and infused by gravity from a height of 30 cm above the midaxillary line.[7] A total of 14.8 L of fluid were instilled and 14.3 L obtained (differential of 500 mL). Because WLL is long and done with large volumes, temperature is monitored to avoid hypothermia and can be maintained using lavage fluid warmed to body temperature, warming blanket and intravenous fluid warmer.[1,3,7]

To prevent ischemia to the extremities pillows in the dependent axilla, under the head and between the thighs were used.[1,5] This procedure is suspended when there is hemodynamic instability, if peripheral oxygen saturation remains low, there is flooding of the contralateral lung or, when the primary objective is reached: the fluid is almost as clear as the instilled saline.[7] Despite the long duration of the procedure, good hemodynamic stability of the patient was achieved, without hydroelectrolyte changes that did not justify replacement, as shown by the values (Table 2).

Thus, understanding the pathophysiology of the disease and the hemodynamic changes to which the patient is subjected, together with a pre-procedure anesthetic assessment and a thoughtful planning, is essential for the success of the procedure.

CONCLUSION

Due to the rarity of PAP and the lack of randomized trials, there are no specific guidelines for the anesthetic management of WLL.[2] The success of the procedure, without complications it is due not only to the patient's good general clinical condition and indication for WLL at an early stage but also to good team coordination.

Preparation, knowledge of the task at hand, as well as an effective communication and performed in a multidisciplinary approach, with coordination between physical therapists, anesthetists and pulmonologists is essential and made it possible to perform safely.

We believe it is necessary to report similar cases that enable the sharing these type of experiences.

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Table 2. Arterial blood gas analysis during right lung lavage

	FiO_2	PEEP	PaO_2	HCO_3^-	$SatO_2$	K^+	Na^+
Pre-Procedure	0.21	-	71	23	96	4.1	136
Post Induction/Pre-Procedure	1	5	345	24.5	100	3.5	135
One Lung Ventilation – Pre Lung Lavage	0.6	7	190	25	100	3.5	136
One Lung Ventilation – During Lung Lavage	0.6	7	187	25	99	3.8	137
End of procedure	0.6	5	107	24	99	4	137
Both lungs ventilation pos-procedure	0.6	5	187	26	100	3.5	137
Post-extubation	0.4	-	91	26	98	3.7	138
15 days after procedure	0.21	-	83	25	97	3.6	139

Legend:
$PaCO_2$- partial pressure of carbon dioxide; PaO_2 - partial pressure of oxygen; HCO_3^- - bicarbonate; $SatO_2$ - oxygen saturation; K^+ - potassium; Na^+ - sodium; FiO_2 - fraction of inspired oxygen; PEEP – positive end expiratory pressure.