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Case Report

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Anesthesia for Unilateral Lung Lavage in a Patient with Pulmonary Alveolar Proteinosis: A Case Report

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ABSTRACT

Background: Pulmonary alveolar proteinosis (PAP) was first described in 1958. Basically, PAP is caused by an impairment of surfactant clearance or abnormal surfactant production, according to various pathogenetic mechanisms and different etiologies. In 90% of cases, it is an autoimmune disease compromising immunoglobulin (Ig)-G and anti-GM-CSF. The objective of this report is to show a case of a patient with severe PAP who was admitted to the emergency room with dyspnea and chest pain and was followed up by the pulmonology outpatient clinic and indicated for pulmonary lavage.

Case Report: A 38-year-old female patient, weighing 95 kg and measuring 155 cm tall, with dyspnea and chest pain with intensity 6/10 for 9 months, which had worsened for 3 months. Admitted to the emergency room with dyspnea, asthenia and chest pain. Diagnosis of PAP is performed through laboratory and imaging tests, with pulmonary lavage under intravenous general anesthesia being indicated. During right pulmonary lavage, 14 liters of warmed saline solution were infused, and 13.6 liters of fluid were drained, lasting 4 hours and resulting in a marked improvement in the symptoms, with the patient being extubated in the ICU 12 hours after the end of the procedure.

Conclusion: Pulmonary alveolar proteinosis is a rare disease that requires special attention during procedures such as whole lung lavage. Anesthetic management presents unique challenges, especially in patients with low pulmonary reserve and risk of hypoxemia, as observed in the reported case.

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Introduction

Pulmonary alveolar proteinosis (PAP) was first described in 1958, found in 27 patients with a remarkable disease of the lung that consists of the filling of the alveoli by a PAS-positive proteinaceous material, rich in lipid [1]. Basically, PAP is caused by an impairment of surfactant clearance or abnormal surfactant production, according to various pathogenetic mechanisms and different etiologies. The usual physiological consequence of PAP is impairment of gas exchange, which can lead to dyspnea, hypoxemia, or even respiratory failure and death [2]. Surfactant accumulation can compromise pulmonary gas transfer and cause several clinical manifestations such as hypoxemic respiratory failure and death [3]. A retrospective study evaluating patients with PAP between 2002 and 2016 in a hospital of the Brazilian Health System (SUS) found only 12 patients [4].

Autoimmune PAP is the most common pathophysiological mechanism, accounting for 90% of documented cases [5]. Autoimmune PAP is initiated by immunoglobulin (Ig)-G anti-

granulocyte macrophage colony-stimulating factor (anti-GM-CSF) antibodies, which decrease functional alveolar macrophages [6]. Prevalence has been reported to be from 0.2 and 3.7 to 40 cases per million depending on the country [5,7]. Autoimmune PAP accounts for approximately 90% of cases, while 4% is secondary PAP, 1% is congenital PAP, and undetermined PAP-like disease represents the remaining 5%. The average age of patients at diagnosis is reported to be between 39 and 51 years, although ages range from newborn to 72 years [7].

The objective of this report is to show a case of a patient with severe PAP who was admitted to the emergency room with dyspnea and chest pain and was followed up by the pulmonology outpatient clinic and indicated for pulmonary lavage.

Case Report

The case report was registered on the Plataforma Brasil with number CAAE: 84296924.6.0000.5078 and approved by the Ethics and Research Committee of the Hospital das Clínicas of the Federal University of Goiás with number 7,227,848. The patient signed the Free and Informed Consent Form and authorization for publication in a medical journal.

A 38-year-old female patient, weighing 95 kg and measuring 155 cm tall, with dyspnea and chest pain with intensity 6/10 for 9 months, which had worsened for 3 months, came to the pulmonology outpatient clinic of the Hospital das Clínicas de Goiânia for clinical evaluation due to clinical worsening. The patient reported dyspnea on minimal exertion, which worsened in the supine position and slightly improved in the standing position. She described asthenia and chest pain as continuous, with an intensity of 6/10, and ventilator dependent. She denied irradiation of chest pain or other ischemic characteristics. She denied other symptoms. The patient was referred to in the yellow room of the unit's emergency room for further investigation and management of the case.

The patient denied other comorbidities besides allergies, anxiety, depression and alcoholism but reported a history of smoking (10 cigarettes per day - she quit 17 years ago). The medications she used continuously were quetiapine 50 mg once at night and aerolin 3 times a day, which was prescribed by a physician in the Family Health Program without a diagnosis of asthma or other lung disease. She denied previous surgeries. During the physical examination, she was in good general condition, with the presence of digital clubbing, diffuse snoring with decreased and symmetrical chest expansion, respiratory rate of 25 breaths per minute and oxygen saturation of 90% under a nasal catheter at 3 L/min.

Laboratory tests were within normal limits, except for increased CRP 2.27 mg/Dl. Computed Tomography (CT) revealed extensive diffuse "paving stone" ground-glass opacities in both lungs. CT angiography revealed the pulmonary artery trunk and branches with preserved caliber and absence of pleural effusion. The findings suggested diffuse alveolar damage, admitting an atypical infectious etiology, pulmonary congestion or even alveolar hemorrhage and alveolar proteinosis. There was no tomographic signs of pulmonary embolism. The electrocardiogram showed sinus rhythm and transthoracic echocardiogram indicated an ejection fraction of 52% with left ventricular systolic function at the lower limit of normal and mild mitral regurgitation.

During bronchoscopy with biopsy, an inadvertent pneumothorax was triggered on the right, confirmed by chest X-ray. The patient was admitted to the surgical center for chest drainage without complications and transferred to the Intensive Care Unit (ICU) in a stable condition, but with mild respiratory distress, using a face mask at 15 L/min without good lung expansion. A spontaneous pneumothorax on the left was also evidenced, requiring chest drainage, and a bronchopleural fistula on the right. To improve the patient's clinical condition, a total lung lavage was proposed in two stages (Figure 1).

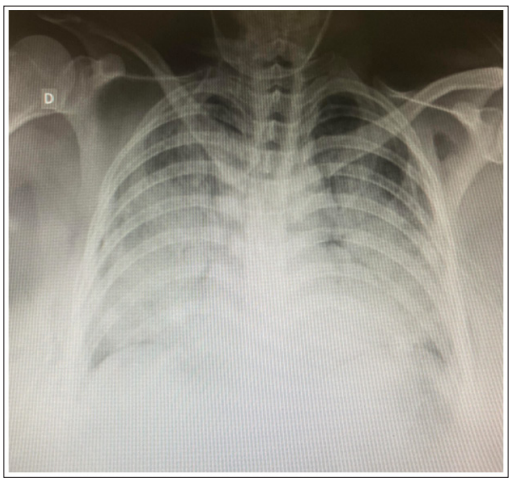

Figure 1: Pre-Lavage Chest Radiograph

During the pre-anesthetic visit, the patient was drowsy, using an oxygen mask at 8 L/min, dyspneic and tachycardia, without the use of vasoactive drugs and afebrile. Total intravenous general anesthesia was chosen. The patient was monitored with cardioscope, invasive blood pressure, pulse oximetry, central (Esophageal) temperature, diuresis, capnography and gas analyzer, in addition to serial blood gas collection. Peripheral and central vein catheterization was performed for infusion of fluids and drugs. The patient received 100% oxygen by mask for 3 minutes, and general anesthesia was initiated by the total intravenous technique and ventilatory support through the anesthesia machine Mindray A7. Propofol was administered by infusion at a target dose of 3 µg/mL and fentanyl at a dose of 3 µg/kg, rocuronium bolus 0.6 mg/kg. Anesthesia was maintained with remifentanil (0.1 µg/kg/min), propofol (3 µg/mL) continuously and ketamine 0.2 mg/kg/h. For orotracheal intubation, a double-lumen endobronchial tube (DLT number 35) was used on the left, and positioning was checked with auscultation and endobronchial optical fiberoscopy, and the tube was fixed. After 20 minutes of bilateral ventilation with 100% inspired oxygen fraction, with a tidal volume of 380 mL, PEEP of 6 and peak pressure of 40 mmHg, left-sided single-lung ventilation was initiated to lavage the contralateral lung.

With the patient in the supine position, 0.9% sodium chloride heated to 37°C was used for lavage and infusion was performed by gravity at a height of approximately 50 centimeters above the lung level (Midaxillary line). The fluid was infused with the patient in a prone position and drained in Trendelenburg. Manual chest percussions were performed to help loosen the lipoprotein material from the alveoli and the fluid was then drained by gravity into a suitable container located at floor level.

Infusions and drainages were performed repeatedly until the drained fluid was clear (or in case of hemodynamic intolerance of the patient). Due to the difficulty in maintaining adequate oximetry and arterial partial pressure of oxygen levels, FiO2 of 1.0 was used throughout the procedure. For single-lung ventilation, the Volume-Controlled mode (VCV) was used, with a tidal volume

of 6 ml/kg, respiratory rate between 20-25 breaths per minute, peak pressure of up to 40 cmH2O and PEEP between 5-7 cmH2O.

Fourteen liters of warmed saline solution were infused, and 13.6 liters of fluid were drained (Figure 2). The patient presented respiratory acidosis during lavage, decreased saturation, and significant hypoxemia, requiring the procedure to be interrupted several times for bilateral ventilation, alveolar recruitment maneuvers, and correction of ventilatory disorders. To improve the acidosis, ventilation parameters were changed, increasing tidal volume and respiratory rate (Table I).

Figure 2: Containers for Collecting the Solution Demonstrate the Clearing of the Fluid Drained in the First Procedure (Washing of the Right Lung)

Table 1: Parameters Evaluated before, During and after Lung Lavage

PARAMETERS	BEFORE LAVAGE	DURING LAVAGE	AFTER LAVAGE
pH	7.36	7.22	7.16
pO_2 (mmHg)	62.3	91.9	66.7
pCO_2 (mmHg)	48.4	68	68.6
HCO_3 (mmol/L)	26.8	27.7	24
BE (mmol/L)	0.8	-1.3	-5.9
Ht (%)	38	39	40
Hb (g/dL)	12.9	13.1	13.6
Sat O_2 (%)	89.3	95	86.7
Na^+ (mmol/L)	139.6	139.1	140.8
K^+ (mmol/L)	3.07	3.13	3.34
Ca_2^+ (mmol/L)	1.15	1.11	1.07
Cl^- (mmol/L)	103	106	107
Lactate (mmol/L)	0.93	0.92	0.76

At the end of the lavage, an intravenous diuretic (furosemide 40 mg) was administered, manual ventilation was performed to restore lung expansion, the double-lumen endotracheal tube was exchanged for a single-lumen tube and the patient was transferred under sedation to the ICU to be extubated in the next few hours to improve ventilatory patterns, aiming to improve residual absorption of the infused liquid.

The surgical procedure lasted approximately 4 hours. The patient remained hemodynamically stable without the need to start an infusion of vasoactive drugs. She was extubated without complications in the ICU 12 hours after admission for the procedure. After the hospital discharge, she was instructed to continue outpatient monitoring and to assess the need for the contralateral procedure. There was clinical, radiological and laboratory improvement after the procedure. There was no need for a new approach or lavage of the left lung. No pulmonary function tests were performed.

Discussion

Lung lavage is a well-established procedure in the treatment and control of PAP and should be used with selective intubation and a double-lumen tube, with it being important to check the position of the tube, which was successfully performed in the right lung to wash the right lung with 14 liters of warmed saline solution being infused, and 13.6 liters of fluid were drained, lasting 4 hours.

The clinical symptoms of PAP range from mild to urgent with nonspecific symptoms such as dyspnea in 39% of patients and cough in 21% of patients, most patients are smokers from 53% to 85%. Hemoptysis, fever and chest pain are rare complaints of autoimmune PAP and should consider another diagnosis. Fever may be present in 24% of patients with secondary PAP due to concomitant hematologic malignancies or opportunistic infections. However, 33% of patients may be asymptomatic at the time of presentation. The patient was admitted to the emergency room with dyspnea on minimal exertion, moderate continuous chest pain and asthenia, the most common symptoms in PAP.

In the case of bilateral PAP, lung lavage can be performed bilaterally, in the same procedure, with extracorporeal circulation or in two separate stages of a few days, which seems safer, as there may be an initial worsening in gas exchange [8]. The Whole-Lung Lavage (WLL) is the current standard treatment and consists of sequential lavage of each lung to mechanically remove the residual material from the alveoli. Although WLL is considered safe, unexpected complications can occur. Common complications associated with WLL include hypoxemia, hemodynamic instability, hypothermia, and hydrothorax or pneumothorax [9]. In the present case, the WLL was unilateral, and the patient presented respiratory acidosis during lavage, decreased saturation, and significant hypoxemia, requiring the procedure to be interrupted several times for bilateral ventilation, alveolar recruitment maneuvers, and correction of ventilatory disorders.

The most used anesthesia technique involves the use of a DLT to achieve lung isolation. The most affected lung is usually lavaged first, and if both lungs are equally affected, the left lung, given its smaller volume, is lavaged first. In the present case, WLL was performed on the right lung. Total intravenous anesthesia is the technique of choice for WLL [9,10]. It guarantees uninterrupted administration of the anesthetic, regardless of lung ventilation, while avoiding contamination of the operating room due to halogenated agents. Because of the large volume of fluid infused into the WLL, some patients may absorb large amounts of this fluid during lavage, so a restrictive approach to perioperative intravenous fluid administration should be considered to avoid volume overload.

Recently, the combination of Ultrasound (US) was used to assess the amount of fluid contained in the lung [11]. It is essential to monitor non-ventilated and ventilated lungs using US to avoid complications during the procedure. The availability of US during the WLL procedure may allow early diagnosis of complications and improve patient prognosis. In the present case, it was not possible to use US to assist in lung assessment during the WLL procedure.

Conclusion

Pulmonary alveolar proteinosis is a rare disease that requires special attention during procedures such as whole lung lavage. Anesthetic management presents unique challenges, especially in patients with low pulmonary reserve and risk of hypoxemia, as observed in the reported case. Total Intravenous Anesthesia (TIVA) is the most frequently used anesthetic technique for a WLL. TIVA is useful because the anesthesia team can be assured of continuous administration of the anesthetic to the patient, and it also prevents contamination of the operating room with inhaled anesthetic during the procedure. The PAP is a very rare disease, which limits the number of patients available, making it difficult to conduct controlled research trials. There was clinical, radiological and laboratory improvement after the procedure in the right lung, and there was no need for a new approach or lavage of the left lung. However, the publication of case reports such as this in several journals will likely lead to future advances in the treatment of these patients.

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