Respir Case Rep 2022;11(1):36-40 DOI: 10.5505/respircase.2022.98470

A Case of Pulmonary Alveolar Proteinosis in which a New Approach to the Traditional Whole Lung Lavage Technique is Preformed

Geleneksel Total Akciğer Lavaj Tekniğine Yeni Bir Yaklaşımın Uygulandığı Bir Pulmoner Alveolar Proteinozis Olgusu

Goksel Altinisik[1], Nilufer Yigit[1], Beste Metin[1], Merve Türkarslan[1], İlknur Hatice Akbudak[2]

Abstract

Pulmonary alveolar proteinosis is a rare interstitial lung disease that is characterized by an accumulation of surfactant composed of proteins and lipids in the alveoli. Whole Lung Lavage (WLL) protocols, which have been used for more than half a century and are still considered the optimum approach, vary according to centers, and there is no standard lavage technique. In this case report we describe a method that does not require technology in which the patient's chest is vibrated with strong and rhythmic movements, using it instead of the manual vibration method used in our WLL applications for the mechanical washing of the alveolar space. In this case report, we also draw attention to the fact that intensive care hospitalization can be prevented by CPAP treatment in the recovery room before hypoxemia deepens. In conclusion, our case report shows that more lung areas can be included in the lavage through the addition of the easily accessible and applicable vibration method that we describe to the WLL technique. The study also reports on our observations that CPAP support applied during postoperative recovery can reduce the hospitalization of patients in intensive care after WLL.

Key words: Bronchoalveolar lavage, continuous positive airway pressure, postural drainage, Pulmonary Alveolar Proteinosis.

Özet

Pulmoner alveolar proteinozis, proteinler ve lipitlerden oluşan yüzey aktif maddenin alveollerde birikmesiyle karakterli nadir görülen bir interstisyel akciğer hastalığıdır. Yarım asrı aşkın süredir kullanılan ve günümüzde de altın standart tedavi kabul edilen Tam Akciğer Lavajı (TAL) protokolleri merkezlere göre değişmekte standart bir lavaj tekniği bulunmamaktadır. Alveoler boşluğun mekanik yıkamasının esas olduğu TAL uygulamalarımızda kullandığımız manuel vibrasyon yöntemi yerine kullanarak gözlemsel olarak daha iyi sonuç aldığımız, hastanın göğüs kafesini güçlü ve ritmik hareketlerle titreştiren, teknoloji gerektirmeyen bir yöntemi bu olgu sunumunda tanımlamayı amaçladık. Bu olgu sunumumuzda ayrıca, hipoksemi derinleşmeden derlenme odasında uygulanacak CPAP tedavisiyle yoğun bakım yatışının önleyebileceğine dikkat çekmeyi amaçladık. Sonuçta olgu sunumumuz, TAL tekniğine eklenmek üzere tariflediğimiz kolay erişilebilir ve uygulanabilir olan vibrasyon yöntemiyle daha fazla akciğer alanının lavaja katıldığını, postoperatif derlenme sırasında uygulanacak CPAP desteği ile TAL sonrası hastaların yoğun bakıma yatışlarının azalabileceği yönündeki gözlemlerimizi içermektedir.

Anahtar Sözcükler: *Bronkoalveoler lavaj, sürekli pozitif havayolu basıncı, postural drenaj, Pulmoner Alveoler Proteinoz.*

[1]Department of Chest Diseases, Pamukkale University, Denizli, Turkey
[2]Department of Anesthesiology and Reanimation, Pamukkale University, Denizli, Turkey

[1]Pamukkale Üniversitesi, Göğüs Hastalıkları Anabilim Dalı, Denizli
[2]Pamukkale Üniversitesi, Anesteziyoloji ve Reanimasyon Anabilim Dalı, Denizli

Submitted (Başvuru tarihi): 28.07.2021 ***Accepted (Kabul tarihi):*** 25.10.2021

Correspondence (İletişim): Nilufer Yigit, Department of Chest Diseases, Pamukkale University, Denizli, Turkey

e-mail: nilufer_savurmus@hotmail.com

Pulmonary alveolar proteinosis is a rare interstitial lung disease that is characterized by the alveolar accumulation of a surfactant consisting of proteins and lipids, of which three types have been defined: congenital, autoimmune (idiopathic) and secondary (1). Ground-glass infiltrations and crazy paving patterns are seen in lung computed tomography (CT). Although open lung biopsy is considered the optimum approach, a pulmonary alveolar proteinosis diagnosis can be made without biopsy based on clinical and radiological findings, milky bronchoalveolar lavage (BAL) fluid and periodic acid-Schiff positive globules (2). Although good results have been reported with treatments such as GM-CSF administration; plasmapheresis, aimed at reducing the level of anti-GM-CSF antibodies; and rituximab (3-4), the optimum treatment is whole lung lavage (WLL). The lavage technique used in this treatment protocol, which has been used for more than half a century, may not be standard due to technological limitations. For this reason, it has been reported that centers that decide to apply WLL make use of processes that have been self-developed over time with the gradual improvement of the lavage technique (5).

A total of nine PAP patients have been followed-up by our clinic for over 10 years (6). Although there are differences in the number of WLLs endured by each patient, the application has remained unchanged from what was learned from the first patient, and similar results have been obtained with nuances (6,7). In previous approaches, the process was terminated after the color of the washing liquid gradually became lighter and consistently clears, and so, there was no need for further attempts at the standard procedure. During the WLL of our last patient, a real-time urgent innovation was required to resolve the problem of premature clear returns of lavage fluid that were inappropriate with the clinical and radiological condition of the patient. With the wisdom of collective thinking, a novel method aided in the reaching of a striking result. To encourage an exchange of views on entering the approach into routine practice, we present it to our colleagues in the field.

CASE

The patient (female; aged 42) had been diagnosed with pulmonary alveolar proteinosis based on the result of a lung biopsy obtained by an external center. She had no accompanying disease and was an active smoker with a 10-year history. As a cleaning worker, she had been regularly exposed to chemicals for 12 years. A physical examination revealed bilateral crackles that were predominantly in the lower zones, as well as lung auscultation and finger clubbing. A lung CT revealed a diffuse crazy paving pattern (Figure 1).

WLL was performed respectively to the right and left lungs with a 1 week interval between in May 2020. Due to clinical and radiological worsening 1 month after discharge, the same approach to WLL was planned. The preferred side was the right lung and the same procedures were performed, although the patient developed gradually increasing dyspnea and desaturation in the recovery room, which was attributed to the atelectasis of lung areas despite the recruitment maneuver. We concluded that the patient could have benefited from the noninvasive mechanical ventilator application, and so Continuous Positive Airway Pressure (CPAP) treatment was applied to the patient, and rapid improvement was observed.

The details of recent WLL

The patient again underwent WLL of the left lung 10 days later, but with a modified technique. In general, the density and the color of each portion of washing fluid during the WLL process changes from milky to watery. In our patient, very dense milky liquid with excessive sediment was first observed, with gradually lighter liquids returned in consecutive collections. However, unexpected fluctuations were observed in the color and sediment of the liquid as the process continued. While significant clarification was observed at the sixth liter, the following liquid was thicker than the first sample. While the fluid became clearer again, the lack of a stable clearance was a cause of concern for the operation team. It was believed that postural drainage was not as effective for the posterior parts of lung in the supine position, leading the anesthesiology technician to suggest adapting a vibration technique to remedy the situation. Thus, a circular vibration motion similar to shaking was applied to the patient using a strip of surgical cloth placed on the lower part of the patient's chest to stimulate the clearance of lower lobe segments (Figure 2). After a slight fluctuation was noted in the color darkness, the process was continued until a distinct dark color and consistency was noted at the 14th liter. After observing permanent clarification, the process was completed with 18 L (Figure 3).

The patient recovered more easily and faster than with previous applications and was discharged the next day under long-term oxygen support. The patient was advised to apply to the hospital in the event of any clinical worsening.

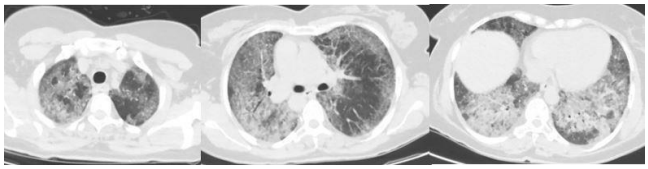

Figure 1: *Bilateral crazy paving pattern in thorax CT*

Figure 2: *A circular vibrating motion similar to shaking was applied to stimulate cleansing of the lower lobe segments with the help of a strip-shaped surgical cloth placed on the lower part of the patient's chest*

The permission of the patient and the legal person was obtained for the reporting of the case as a case report.

DISCUSSION

WLL is performed by pulmonologists, thoracic surgeons and anesthesiologists, and is an uncommon treatment method for centers other than in the usual surgical interventions due to the need to hold the operation room for a long time only for the general anesthetic requirement. Being a rare, but well-described condition, innovations are infrequent. Describing this first-time application of a novel addition to the traditional WLL approach opens it to discussion among clinicians who perform WLL for PAP.

It would be beneficial to provide details of the standard approach to WLL according to literature, where we learned how to perform WLL for our first case. This was before the advent of video media via the Internet, but our patient was insistent on being lavaged by our team. Generally, WLL is performed under general anesthesia with the use of a double-lumen tube. Approximately 500–1000 mL of fluid is applied to the lung to which WLL is to be applied and the washing continues, while single lung ventilation is applied to the other side. Manual chest percussions are performed during fluid delivery, and the fluid is drained from the lungs to another container under the effect of gravity. The isotonic saline solution is heated to 37°C, and given in 500–1000 cc portions to a single lung at a time, with a total of 15–20 L. The recovered lavage fluids are collected in separate glass containers and examined macroscopically. Initially, the liquid is opaque and milky, and becomes lighter towards the end of the procedure (4). According to our previous experiences, for the addition to the procedure of percussion to the lavaged hemithorax, we position the operating table for the entire WLL procedure to facilitate the principles of postural drainage, allowing the fluid to enter and exit from different areas of the lung. When the collected fluid becomes clear without any re-darkening, we terminate WLL with the recruitment maneuver and extubation. Over more than 10 years, nine PAP patients have been followed by our clinic. Although there are differences in the number of WLL procedures undergone by each patient, the application has not changed since the first patient, and similar results were obtained, with some nuances (6). The process is terminated following the gradual lightening of the dark washing liquid, as learned from medical sources and experienced colleagues. For our previous PAP patients, the WLL technique worked very well, and no adaptations to the standard procedure were needed.

In the case we present here, unlike in other case examples presented in literature and our experiences, there were remarkable fluctuations in the color of the fluid. To increase the effects of the percussion motion and positioning of the operating table, a circular vibration/shaking movement was applied using a surgical cloth located at the lower part of the rib cage as an addition to the lavage procedure. We consider this to be a valid alternative to the traditional vibration movement, as in the case presented here the change in color of the exiting liquid collected in the glass bottles suggested the process can be successful.

Oxygenation and ventilation vary at different stages of WLL, and so hypoxia is the leading postoperative complication (8). During filling up of the non-ventilated lung with lavage fluid, there is ipsilateral pulmonary capillary bed compression resulting in a diversion of blood flow toward the ventilated lung. This effect can temporarily decrease the shunt created by the single-lung ventilation and help with oxygenation. Conversely, blood flow to the non-ventilated lung increases during the drainage of the lavage fluid from the non-ventilated lung, leading to hypoxemia by increasing the shunt (8). Various recruitment maneuvers can be used to reduce the hypoxemia associated with this mechanism. In recent patients, we applied manual intermittent double lung ventilation as a norm at the end of the procedure. Unlike in previous applications, the deep hypoxemia continued in the recovery room. Because of observing the patient not fully conscious related to recent general anesthesia, noninvasive mechanical ventilation was been avoided especially when the patient was severely dyspneic. However, in the our recent experience, CPAP therapy was applied to achieve positive pressure ventilation to support the ventilation-perfusion balance and to reduce pulmonary edema to gain time before transferring the patient to the intensive care unit, where no beds were available. Saturation values started to increase within seconds of the initiation of CPAP therapy. CPAP treatment has been used for many years in the postoperative intensive care follow-up of patients who have undergone WLL, but we consider it necessary to mention in this case report when it should be applied rather than the application of CPAP treatment.

Figure 3: a: *lavage view of the same patient after the procedure performed 1 month ago* ***b,c:*** *Difference in sediment levels of whole lung lavage and stabilization of opening in sediment after 18 liters*

CPAP treatment started in the postoperative recovery room provides a fast and permanent response to hypoxemia, and prevents unnecessary intensive care unit admissions.

CONCLUSION

The highly instructive case presented here provided us with two very important experiences that we considered worth sharing with our colleagues. We share these gains in the belief that it will accelerate the development of the management of diseases, especially orphan diseases, in which there are no definitive results in terms of diagnosis, follow-up and treatment. First, we have contributed to procedures aimed at the cleansing of lower lobe segments with a circular motion, which had a stimulating effect in bringing about a fluctuation in the color of the returned lavage fluid during the procedure. Second, it seems valuable for a fast and permanent response to apply CPAP treatment when a deep hypoxemia occurred postoperatively.

ACKNOWLEDGEMENT

We would like to thank Semih Eren, an anesthesia technician who worked devotedly during the procedure.

CONFLICTS OF INTEREST

None declared.

AUTHOR CONTRIBUTIONS

Concept - G.A., N.Y., B.M., M.T., İ.H.A.; Planning and Design - G.A., N.Y., B.M., M.T., İ.H.A.; Supervision - G.A., N.Y., B.M., M.T., İ.H.A.; Funding - G.A., N.Y., B.M., M.T.; Materials - N.Y., B.M., M.T.; Data Collection and/or Processing - N.Y., B.M., M.T.; Analysis and/or Interpretation - G.A., N.Y.; Literature Review - N.Y.; Writing - G.A., N.Y., B.M., M.T.; Critical Review - G.A., İ.A.H.

YAZAR KATKILARI

Fikir - G.A., N.Y., B.M., M.T., İ.H.A.; Tasarım ve Dizayn - G.A., N.Y., B.M., M.T., İ.H.A.; Denetleme - G.A., N.Y., B.M., M.T., İ.H.A.; Kaynaklar - G.A., N.Y., B.M., M.T.; Malzemeler - N.Y., B.M., M.T.; Veri Toplama ve/veya İşleme - N.Y., B.M., M.T.; Analiz ve/veya Yorum - G.A., N.Y.; Literatür Taraması - N.Y.; Yazıyı Yazan - G.A., N.Y., B.M., M.T.; Eleştirel İnceleme - G.A., İ.A.H.

REFERENCES

1. Riley L, Wang T, Ataya A. Pulmonary alveolar proteinosis (PAP). Am J Respir Crit Care Med 2019; 200:16-7. [CrossRef]
2. Jouneau S, Ménard C, Lederlin M. Pulmonary alveolar proteinosis. Respirology 2020; 25:816-26. [CrossRef]
3. Trapnell BC, Inoue Y, Bonella F, Morgan C, Jouneau S, Bendstrup E, et al. Inhaled molgramostim therapy in autoimmune pulmonary alveolar proteinosis. N Engl J Med 2020; 383:1635-44. [CrossRef]
4. Malur A, Kavuru MS, Marshall I, Barna BP, Huizar I, Karnekar R, et al. Rituximab therapy in pulmonary alveolar proteinosis improves alveolar macrophage lipid homeostasis, Respir Res 2012; 13:46. [CrossRef]
5. Suzuki T, Trapnell BC. Pulmonary alveolar proteinosis syndrome. Clin Chest Med 2016; 37:431-40. [CrossRef]
6. Altınışık G, Ugurlu E, Bir F, Cengiz B, Trapnell BC. What we have learned from an unpredictable teacher: Autoimmune Pulmonary Alveolar Proteinosis: The 2018 International Rare Lung Diseases Research Conference and LAMposium, 6-9 September 2018, Cincinnati, Ohio: P-05.
7. Smith BB, Torres NE, Hyder JA, Barbara DW, Gillespie SM, Wylam ME, et al. Whole-lung lavage and pulmonary alveolar proteinosis: Review of clinical and patient-centered outcomes. J Cardiothorac Vasc Anesth 2019; 33:2453-61. [CrossRef]
8. bdelmalak BB, Khanna AK, Culver DA, Popovich MJ. Therapeutic whole-lung lavage for pulmonary alveolar proteinosis: A procedural update. J Bronchology Interv Pulmonol 2015; 22:251-8. [CrossRef]