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CASE REPORT

Clinical Aspects and Evolution under Current Treatments on Pulmonary Alveolar Phospholipoproteinosis Patients

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A 34-year-old female smoker (4 packs a year), with the following symptoms: dry cough, moderate dyspnea, fatigue, night sweats and no exposure to respiratory poisoning or contact with tuberculosis. Clinical findings at admission: normal temperature, Hippocratic fingers and cyanosis, diminished bilateral breath sounds, fine bilateral rattling respiratory sounds, rhythmic cardiac noises, oxygen saturation 95%, pulse 87, blood pressure 125/80 mm Hg. Chest computed tomography describes infiltration images of intra-alveolar and intra-bronchial with matte glass pattern, confluent and stretched across all lung segments. Pulmonary biopsy: preserved pulmonary architecture, thick alveolar septs and terminal bronchioles, alveoli and macrophages loaded with lipoprotein material. Bronchial aspirate: negative BAAR, no tumor cells. Bronchoalveolar lavage: opalescent, abundant PAS + appearance. total cell numbers – 4.3 million, macrophages – 34.1%, lymphocytes – 56.9%, neutrophils – 10%, eosinophils – 0.4%, epithelial cells – 33%.The treatment option chosen for this case was total bronchoalveolar lavage for therapeutic purposes. The patient had a rapid favourable evolution and at discharge was recommended a periodic imaging and functional control.

Key Messages: Bronchoalveolar lavage is the optimal diagnostic method. Moderate/ severe forms, total bronchoalveolar lavage is recommended. Systemic corticotherapy or immunosuppression is not indicated. GM-CSF administered subcutaneously or by nebulization. Rituximab/ plasmapheresis are under evaluation. Pulmonary transplantation is indicated in patients who do not respond to therapeutic bronchoalveolar lavage repeated 6-12 months.

INTRODUCTION

Pulmonary alveolar proteinosis (PAP) is a rare pulmonary disease that concerns pulmonary interstitium through an abnormal and excessive accumulation of surfactant inside the alveoli, which can affect gas exchange and can lead to dyspnea and limited efforts.[1] It is a diffuse pulmonary disease of unknown etiology and characterized by the accumulation in the distal airspace of a positive amorphous PAS lipoproteinate material that does not alter the pulmonary architecture.[2] The following forms of PAP have been defined: a) congenital PAP, where mutations may occur: GM-CSF receptor level, surfactant genes or a transport defect; b) secondary PAP, exposure caused by: exposure to dust, haematogenic diseases or allogeneic bone marrow transplantation.[3] In 30% of cases the PAP patients are asymptomatic. At pathophysiological level, an imbalance between the homeostatic mechanism by surfactant production and the alveolar macrophage clearance has been identified and that results in the accumulation of lipoproteinic material in the alveoli. Frequently PAP affects males at a ratio of 4:1, at an average age of 35 years.[4]

CASE HISTORY

A 34-year-old female smoker (4 packs a year), with the following symptoms: dry cough, moderate dyspnea, fatigue, night sweats and no exposure to respiratory poisoning or contact with tuberculosis. No other past hospitalizations. Clinical findings at admission: normal temperature, Hippocratic fingers and cyanosis, diminished bilateral breath sounds,

fine bilateral rattling respiratory sounds, rhythmic cardiac noises, oxygen saturation 95%, pulse 87, blood pressure 125/80 mm Hg. Differential blood count: leukocytes 13.650/mm^3, lymphocytes 4.75/mm^3, hemoglobin 17.70 g/dl, hematocrit 48%, urea 28 mg/dl, creatinine 1.7 mg, total cholesterol serum 210 mg/dl, triglycerides 221 mg/dl. Arterial gasometry with PaO_2 – 73 mm Hg, $PaCO_2$ – 44 mm Hg, pH – 7.44. Lung function tests: FVC – 91.9%, FEV1 – 93.9%, FEV1/VC – 85.69%, PEF – 73.6%, MEF – 115.3%, and DLCO – 53.8%. The 6-minute walk test: 650 m with a value of 87.90% of the predicted value with significant desaturation (95% to 83%). The degree of dyspnea on the Borg scale was 2. Electrocardiogram and echocardiography were both normal. Chest X-ray describes diffuse micronodular opacities disseminated bilaterally with the tendency to confluence.[5] **(Fig. 1)**

Chest computed tomography describes infiltration images of intra-alveolar and intrabronchial with matte glass pattern, confluent and stretched across all lung segments.[6] **(Fig. 2)**

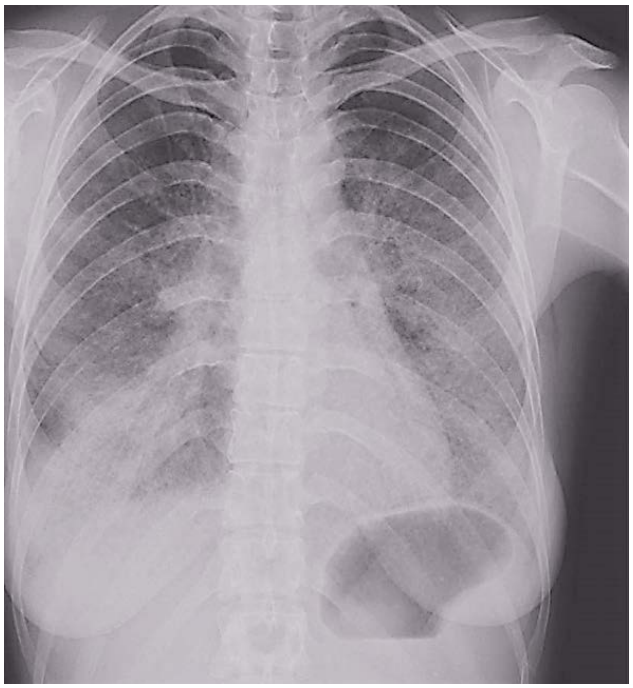

Figure 1. Postero-anterior chest radiography.

Fibrobronchoscopy: bilateral diffuse bronchial appearance with no active lesions of the mucosa. Pulmonary biopsy: preserved pulmonary architecture, thick alveolar septs with terminal bronchioles, alveoli and macrophages loaded with lipoprotein material.[7] **(Fig. 3)**

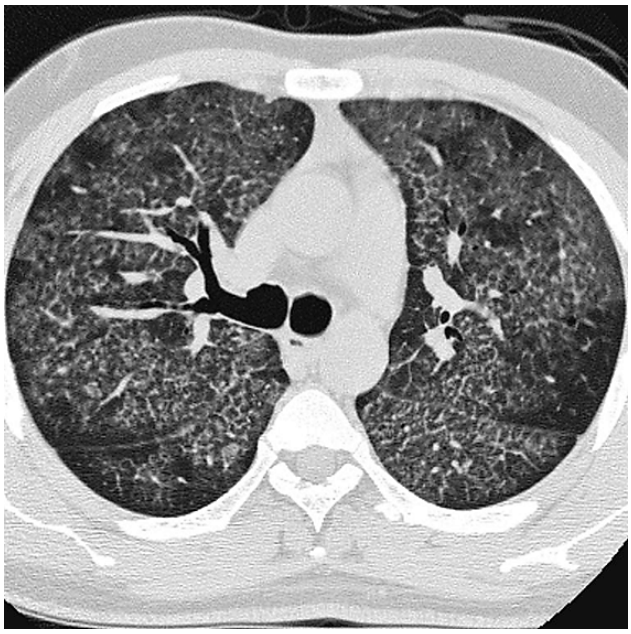

Figure 2. High resolution computed tomography..

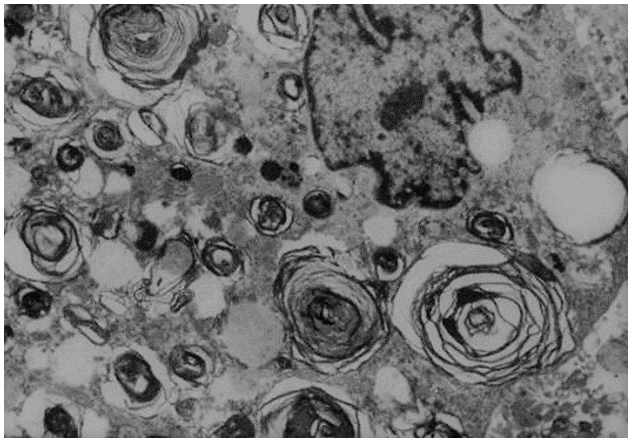

Figure 3. Histopathology sample.

Bronchial aspirate: negative BAAR, no tumor cells. Bronchoalveolar lavage: opalescent, abundant PAS + appearance, total cell numbers – 4.3 million, macrophages – 34.1%, lymphocytes – 56.9%, neutrophils – 10%, eosinophils – 0.4%, epithelial cells – 33%. **(Fig. 4)**.

After discussing smoking cessation methods, the treatment option chosen for this patient was total bronchoalveolar lavage for therapeutic purposes.[8] *The patient had a rapid favourable evolution and at discharge was recommended a periodic imaging and functional control. At six-month evaluation, the patient's overall condition was within normal limits, but was regularly monitored for possible changes in the state of health.* **(Fig. 5)**

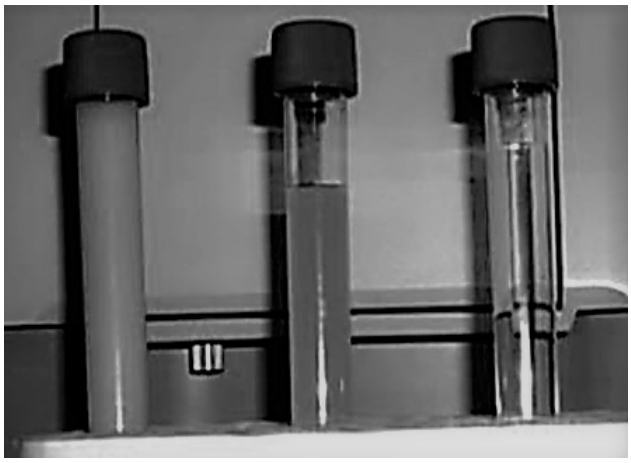

Figure 4. Test glass tubes containing the bronchoalveolar lavage fluid.

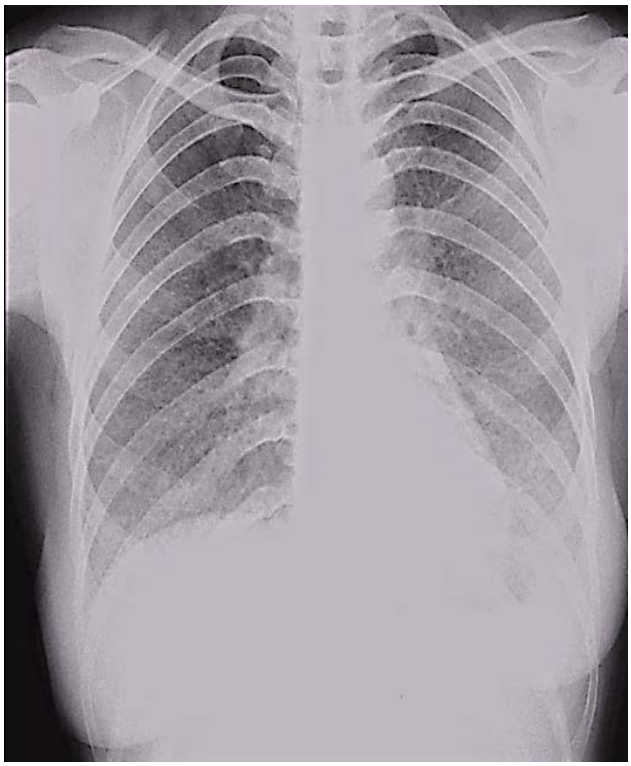

Figure 5. Chest X-ray post-total bronchoalveolar lavage.

DISCUSSION

In moderate or severe forms, total bronchoalveolar lavage is recommended. Systemic corticotherapy or immunosuppression is not indicated. Other variants of treatment: GM-CSF administered subcutaneously or by nebulization. Rituximab or plasmapheresis are under evaluation. Pulmonary transplantation is indicated in patients who do not respond to therapeutic bronchoalveolar lavage repeated at 6-12 months.[10]

CONCLUSION

Pulmonary alveolar proteinosis is a rare disease that poses difficulties in diagnosis through uncharacteristic symptoms. Differential diagnosis includes diseases with the same radiological aspect: cardiogenic pulmonary edema, pneumocystis jiroveci pneumonia, non-small cell lung cancer or small cell lung cancer, sarcoidosis. Bronchoalveolar lavage is the optimal diagnostic method. Treatment management depends on the progression of the disease.[9]

REFERENCES

1. Borie R, Danel C, Debray MP, et al. Pulmonary alveolar proteinosis. Eur Respir Rev 2011;20:98-107.
2. Shah PL, Hansell D, Lawson PR, et al. Pulmonary alveolar proteinosis: clinical aspects and current concepts on pathogenesis.Thorax 2000;55:67-77.
3. Seymour JF, Presneill JJ. Pulmonary alveolar proteinosis: progress in the first 44 years. Am J Respir Crit Care Med 2002;166(2):215-35.
4. Rosen SH, Castleman B, Liebow AA. Pulmonary alveolar proteinosis. N Engl J Med 1958;258(23):1123-42.
5. Godwin JD, Müller NL, Takasugi JE. Pulmonary alveolar proteinosis: CT findings. Radiology 1988; 169(3):609-13.
6. Murayama S, Murakami J, Yabuuchi H, et al. "Crazy paving appearance" on high resolution CT in various diseases. J Comput Assist Tomogr 1999;23(5):749-52.
7. Carey B, Trapnell BC. The molecular basis of pulmonary alveolar proteinosis. Clin Immunol 2010;135(2):223-35.
8. Suzuki T, Sakagami T, Young LR, et al. Hereditary pulmonary alveolar proteinosis: pathogenesis, presentation, diagnosis, and therapy. Am J Respir Crit Care Med 2010;182(10):1292-304.
9. Claypool WD, Rogers RM, Matuschak GM. Update on the clinical diagnosis, management, and pathogenesis of pulmonary alveolar proteinosis. Chest 1984;85(4):550-8.
10. Trapnell BC, Suzuki T. Pulmonary alveolar proteinosis. In: Grippi MA, Elias JA, Fishman JA, et al. editors. Fishman's Pulmonary Diseases and Disorders. 5th ed. New York, NY: McGraw-Hill Educational; 2015. 1028-37.

Клинические аспекты и развитие современного лечения больных с лёгочным альвеолярным фосфолипопротеинозом

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Нами представлен случай курильшицы 34 лет (4 пачки в день) со следующими симптомами: сухой кашель, умеренная диспнея, усталость, ночное потоотделение и отсутствие контакта с дыхательной интоксикацией или контакта с туберкулёзом. Клинические данные при поступлении: нормальная температура, «пальцы Гиппократа» и цианоз, ослабленные двусторонние дыхательные шумы, нормальный двусторонний храп дыхательных шумов, ритмичное сердцебиение, кислородная сатурация 95%, пульс 87, артериальное давление 125/80 mm Hg. КТ-визуализация грудной клетки показала инфильтрационную внутриальвеолярную и внутрибронхиальную визуализацию с симптомом „матового стекла", простирающуюся по всем сегментам лёгких. Биопсия лёгкого: сохранённая структура лёгких, толстые альвеолярные перегородки и терминальные бронхиолы, альвеолы и макрофаги, заполненные липопротеиновым материалом. Бронхиальный аспират: отрицательный BAAR, отсутствие опухолевых клеток. Бронхоальвеолярный лаваж: опалесцирующий, обильный PAS+ вид. Общее количество клеток - 4,3 млн., макрофагов - 34,1%, лимфоцитов - 56,9%, нейтрофилов - 10%, эозинофилов - 0,4%, эпителиальных клеток - 33%. Лечение, предпринятое в этом случае, представляло собой полный бронхоальвеолярный лаваж в терапевтических целях. Состояние больной быстро пошло на поправку, и при выписке были предписаны периодическая образная диагностика и функциональный контроль.

Ключевые элементы: Бронхоальвеолярный лаваж является оптимальным методом диагностики. При умеренных / тяжёлых формах рекомендуется полный бронхоальвеолярный лаваж. Системная кортикотерапия или иммуносупрессия не назначаются. GM-CSF (гранулоцитарно-макрофагальный колониестимулирующий фактор) применяется подкожно или путём распыления. Терапия ритуксимабом / плазмой ферреса находится на стадии оценки. Лёгочная трансплантация назначается пациентам, состояние которых не улучшается в течение 6-12 месяцев при проведении лечебного бронхоальвеолярного лаважа.