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ABOUT COVER

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AIMS AND SCOPE

The primary aim of *World Journal of Clinical Cases* (*WJCC*, *World J Clin Cases*) is to provide scholars and readers from various fields of clinical medicine with a platform to publish high-quality clinical research articles and communicate their research findings online.

WJCC mainly publishes articles reporting research results and findings obtained in the field of clinical medicine and covering a wide range of topics, including case control studies, retrospective cohort studies, retrospective studies, clinical trials studies, observational studies, prospective studies, randomized controlled trials, randomized clinical trials, systematic reviews, meta-analysis, and case reports.

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CASE REPORT

Unexpected diffuse lung lesions in a patient with pulmonary alveolar proteinosis: A case report

Li Jian, Qi-Quan Zhao

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Abstract

BACKGROUND

Pulmonary alveolar proteinosis (PAP) often presents nonspecifically and can be easily confused with: (1) Idiopathic interstitial lung fibrosis; (2) alveolar carcinoma; (3) pulmonary tuberculosis; and (4) other lung diseases such as viral pneumonia, mycoplasma pneumonia, and chlamydial pneumonia.

CASE SUMMARY

Diagnosis: In this case, a patient was diagnosed with PAP through transbronchial cryobiopsy (TBCB) and quantitative metagenomic next-generation sequencing, which confirmed the impairment of surfactant turnover as the underlying cause of PAP. Interventions: High-volume total lung lavage was performed for this patient. Outcomes: The patient's clinical condition had improved significantly by the 6-month follow-up, with a 92% finger oxygen saturation. A repeat chest computed tomography scan revealed scattered patchy ground-glass shadows in both lungs, which was consistent with alveolar protein deposition but with a lower density than in the radiograph from October 23, 2022.

CONCLUSION

TBCB has unique advantages in diagnosing atypical alveolar protein deposition, particularly for enabling the early detection of PAP. This information can help patients take preventive measures to prevent or halt PAP development by avoiding dusty environments and seeking treatment with total lung lavage and inhaled granulocyte macrophage colony-stimulating factor.

Key Words: Diffuse lung lesions; Pulmonary alveolar proteinosis; Quantitative metagenomic next-generation sequencing; Transbronchial cryobiopsy; High-volume double lung

lavage; Case report

Core Tip: Pulmonary alveolar proteinosis (PAP) is frequently misdiagnosed due to the absence of typical map-like and pavement-like manifestations in the lungs or the absence of typical pathological findings. Transbronchial cryobiopsy has unique advantages in diagnosing atypical alveolar protein deposition, particularly for enabling the early detection of PAP. This information can help patients take preventive measures to prevent or halt PAP development by avoiding dusty environments and seeking treatment with total lung lavage and inhaled granulocyte macrophage colony-stimulating factor.

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INTRODUCTION

Pulmonary alveolar proteinosis (PAP) often presents nonspecifically and can be easily confused with: (1) Idiopathic interstitial lung fibrosis; (2) alveolar carcinoma; (3) pulmonary tuberculosis; and (4) other lung conditions such as viral pneumonia, mycoplasma pneumonia, and chlamydial pneumonia. PAP is frequently misdiagnosed due to the absence of typical map-like and pavement-like manifestations in the lungs or the absence of typical pathological findings. Written informed consent was obtained from the patient for the publication of this case report and accompanying images.

CASE PRESENTATION

Chief complaints

Cough, sputum production, and shortness of breath for 2 mo.

History of present illness

After catching a chill 2 mo ago, there was a cough with white frothy sputum. Shortness of breath occurred after physical activity but improved with rest. Other accompanying symptoms include chills, fever (exact temperature unknown), headache, and dizziness.

History of past illness

The patient had no medical history.

Personal and family history

A 40-year-old female who works in a furniture factory with a history of significant dust exposure.

Physical examination

Temperature: 37.2 °C; Pulse: 90 beats per minute; Respiration rate: 20 breaths per minute; Blood pressure: 108/72 mmHg; Oxygen saturation: 93%. The patient is conscious and cooperative during the physical examination. No significant abnormalities were found during the examination of the lungs, heart, and abdomen.

Laboratory examinations

Further investigation through electron bronchoscopy and quantitative metagenomic next-generation sequencing did not reveal any significant abnormalities. Transbronchial cryobiopsy (TBCB) was performed to obtain 3 pieces of lung tissue. Pathological examination of tis tissue showed localized fibrous tissue hyperplasia, a thickening of some alveolar septa, and an eosinophilic structureless material in the focal alveolar lumen. The histological findings were combined with the clinical findings to obtain the diagnosis. Special staining results were as follows: Periodic Acid Schiff (+), Periodic Acid Schiff Diastase (±), and Congo red (-).

Imaging examinations

The patient had a finger oxygen saturation of 75%, and a chest computed tomography (CT) scan revealed diffuse infectious lesions in both lungs, suggesting possible opportunistic pneumonia (Figure 1).

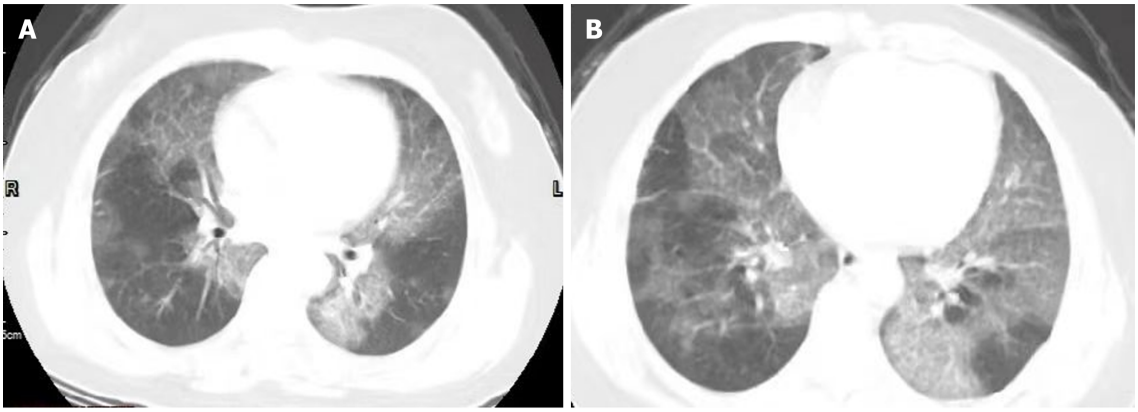

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Figure 1 Chest computed tomography scan shows diffuse infectious lesions in both lungs with the possibility of opportunistic pneumonia, and it was recommended to combine these findings with the findings of the clinical and treatment follow-up. A: The right middle lobe, left lingula, and both lower lobes of the lungs exhibit diffuse ground-glass opacities; B: Both basal segments of the lower lobes of the lungs show diffuse ground-glass opacities.

The patient underwent high-volume double lung lavage. At the 6-mo follow-up, a repeat chest CT scan revealed scattered patchy ground-glass shadows in both lungs, which was consistent with alveolar protein deposition but with lower density than the October 23, 2022 radiograph (Figure 2).

FINAL DIAGNOSIS

The final diagnosis was PAP.

TREATMENT

High-volume double lung lavage was performed on the patient.

OUTCOME AND FOLLOW-UP

The patient's clinical condition had improved significantly by the 6-mo follow-up, with a 92% finger oxygen saturation.

DISCUSSION

PAP is a rare lung disease characterized by the accumulation of phospholipid proteins within the alveoli[1,2]. It often presents insidiously, with typical symptoms including exertional dyspnea progressing to dyspnea at rest, accompanied by cough, white sputum, fatigue, and weight loss. PAP can be classified into the following three types[2-4]: Primary PAP is the most common type and is typically associated with abnormalities in the production of granulocyte-macrophage colony-stimulating factor (GM-CSF) antibodies. These antibodies inhibit the function of macrophages, leading to the obstruction of proteinaceous material clearance within the alveoli. Secondary PAP is caused by other diseases or factors such as certain infections, malignant tumors, immunodeficiency, and so on. Congenital PAP is a rare genetic disease caused by mutations in the genes encoding pulmonary surfactant proteins. The treatment approach depends on the type of PAP[5]: For primary PAP, whole-lung lavage is a common therapeutic method used to remove proteinaceous deposits from the alveoli. In some cases, GM-CSF therapy may also be employed. The treatment of secondary PAP involves addressing the underlying condition. The management of congenital PAP is more complex and typically includes surfactant replacement therapy, antibiotic treatment for infections, and supportive care. In some refractory cases, lung transplantation may be the only viable treatment option. PAP is more common in middle-aged and young adults and is approximately three times more common in men than in women. Dust exposure, particularly exposure to silica dust, can cause PAP. It is believed that PAP may be a nonspecific response to certain stimuli, leading to the breakdown of alveolar macrophages and the production of PAS-positive proteins[3,6-8]. PAP is a challenging disease to diagnose clinically, and a definitive diagnosis is usually made through histopathological analyses[9]. The primary methods of obtaining tissue specimens include transbronchial forceps biopsy (TBFB), percutaneous biopsy, and surgical lung biopsy (SLB). TBFB has limited diagnostic efficacy due to the small size and poor quality of the specimens obtained, making it difficult to meet

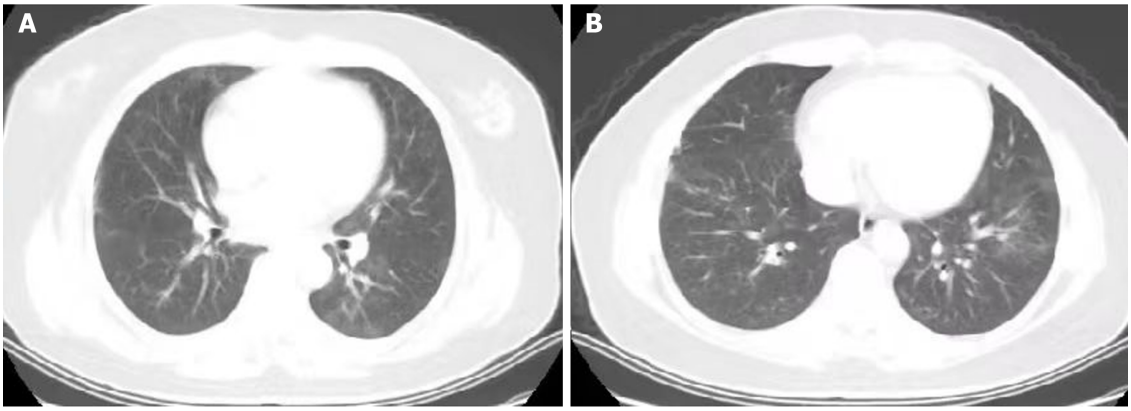

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Figure 2 Chest computed tomography scan shows scattered patchy ground-glass shadows in both lungs, which was consistent with alveolar protein deposition, with lower density than the October 23, 2022 film. A: The diffuse ground-glass opacities in the right middle lobe, left lingula, and both lower lobes of the lungs have significantly resolved compared to previous findings; B: The diffuse ground-glass opacities in both basal segments of the lower lobes of the lungs have significantly resolved compared to previous findings.

pathological analytical requirements. Percutaneous lung puncture biopsy is also associated with the problem of small specimens often not meeting pathological analytical requirements and carries the risk of pneumothorax and hemopneumothorax. SLB is less commonly used due to its high invasiveness, high cost, and limitations in patients with reduced cardiopulmonary function.

CONCLUSION

PAP is often nonspecific and can be easily confused with other lung diseases and conditions, such as idiopathic pulmonary fibrosis, lung cancer, tuberculosis, viral pneumonia, mycoplasma pneumonia, and chlamydial pneumonia. The aim of management is to improve symptoms and quality of life. TBCB is a safe and effective technique for lung tissue biopsy and is used primarily for the etiological diagnosis of diffuse lung disease but also for biopsying localized lesions in the lung periphery. TBCB is associated with less trauma, larger and higher-quality specimens, fewer complications, and significantly lower costs than SLB. TBCB has unique advantages in the diagnosis of atypical alveolar protein deposition, particularly for enabling the early detection of PAP. This information can help patients take preventive measures to prevent or halt PAP development by avoiding dusty environments and seeking treatment with total lung lavage and inhaled GM-CSF.

FOOTNOTES

Author contributions: Jian L contributed to writing the manuscript and participated in helpful discussions; Zhao QQ is the guarantor of this work.

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