

中文題目：多專科團隊會議在間質性肺病上的應用

英文題目：Multidisciplinary discussion in a patient with interstitial lung disease

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Introduction

Identifying and determining the cause of interstitial lung disease can be difficult. Multidisciplinary discussion (MDD) is now viewed as the diagnostic decision-making in interstitial lung disease (ILD). We reported a case of interstitial lung disease with diagnostic challenges.

Case Report

A 63-year-old man with a history of hypertension and gouty arthritis presented to our emergency department with a 2-month history of worsening exertional dyspnea and hypoxemia. He had smoked 3 packs per day for 30 years, and just quit for 2 months. On Physical examination, the oxygen saturation was 70% in ambient air; the breathing sounds revealed diffuse inspiratory Velcro crackles. Chest radiographs showed bilateral reticular opacities. CT of the chest revealed multiple cysts and emphysema in bilateral upper lungs, and honeycombing with traction bronchiectasis predominantly in bilateral lower lungs. Langerhans cell histiocytosis was considered by image. Serologic testing for connective tissue disease was unremarkable. We used high flow nasal cannula for oxygen support, and FiO₂ remained high. Empirical antibiotics and systemic glucocorticoid (equivalent of 2mg/kg/day prednisolone) were administered but the response was limited. For further diagnosis and treatment, he received video-assisted thoracoscopic biopsy of right upper, middle and lower lobes. His lung condition and hypoxemia deteriorated thereafter and he died 11 days after the biopsy. Pathological diagnosis confirmed end-stage interstitial lung disease with negative immunostains for Langerhans cells. After a multidisciplinary discussion (pulmonologist, radiologist, pathologist and chest surgeon), the differential diagnoses included combined pulmonary fibrosis and emphysema (CPFE), smoking-related interstitial pneumonia, or end-stage Langerhans cell histiocytosis.

Discussion

About 10% of ILD cases remain unclassifiable. The 2013 ATS/ERS statement highlighted the importance of a multidisciplinary diagnostic approach. In this case, the clinical, radiologic, and pathological findings demonstrate discordance. CT findings suggested pulmonary Langerhans cell histiocytosis (PLCH), combined pulmonary fibrosis and emphysema (CPFE), or usual interstitial pneumonia (UIP), but none of which was typical. Pathologic findings are unable to make a definite diagnosis due to coexisting patterns, end-stage fibrosis, and inability to take adequate sampling. Several issues were discussed during the multidisciplinary review, including (1) the most likely differential diagnoses (2) the optimal timing for biopsy, weighting diagnostic value against risks of the procedure (3) limitations on lung biopsy. Despite the unfavorable outcome for this patient, the diagnostic strategy can be helpful in difficult ILD cases.

Conclusion

In this case, though the diagnosis remains inconclusive, the multidisciplinary discussion (MDD) improved the knowledge of interstitial lung disease.