A case of progressive pulmonary alveolar proteinosis with lymphadenopathy revealed on ^{18}F-FDG PET/CT

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Abstract

Pulmonary alveolar proteinosis (PAP) is a rare lung disease, which may cause repeating infections. A 36-year-old man had repetitive admissions to our hospital, beginning two years ago, due to episodes of severe dyspnea. Serial computed tomography(CT) scans revealed extensive ground-glass opacities with interlobular/intralobular septal thickening, diffuse consolidations in both lungs and enlarged lower paratracheal lymph nodes. The first biopsy of the right lung and of a mediastinal lymph node showed no evidence of malignancy.Fluorine-18-fluorodeoxyglucose positron emission tomography/CT(^{18}F-FDG PET/CT) was performed in June 2020 following a case of clinical and radiological deterioration to exclude the possibility of malignancy. Positron emission tomography/CT showed increased ^{18}F-FDG uptake in the both lungs and in enlarged mediastinal lymph nodes, with maximum standardized uptake value(SUVmax) of 13.5 and 9.2, respectively. Computed tomography-guided biopsy of the right lower lobe supported the diagnosis of pulmonary alveolar proteinosis.

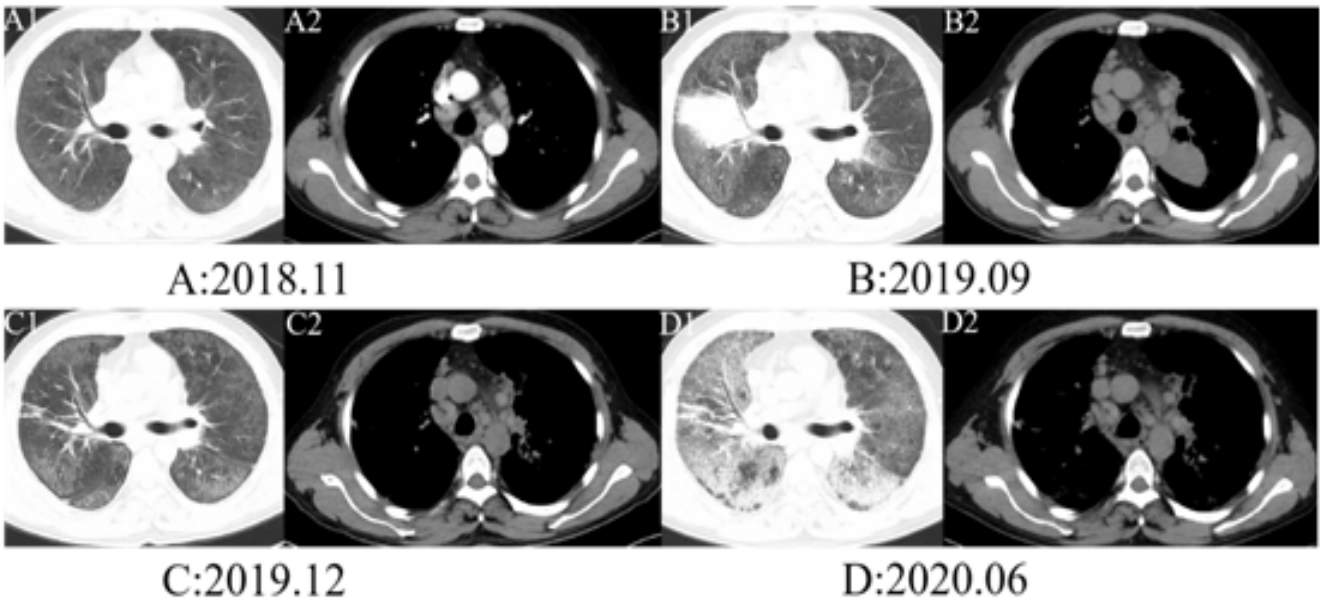

Figure 1. A 36-year-old man had repetitive admissions to our hospital, beginning two years ago (about 11/2018), due to severe dyspnea. His medical history revealed no remarkable events. The chest computed tomography (CT) (A1: pulmonary window, A2: mediastinal window) at that time showed diffuse ground-glass opacities in both lungs and multiple lymph nodes in the mediastinum with the lower paratracheal's largest diameter of 13mm, which were diagnosed as interstitial pneumonia and lymph node reactivity changes. After anti-inflammatory treatment, the above symptoms improved. After ten months (09/2019), the patient presented with dyspnea, chest pain, cough, sputum, hemoptysis, and fever for two weeks. The blood examination showed elevated white blood cells of 17.39x10^9/L. The chestCT (B1: pulmonary window, B2: mediastinal window) indicated diffuse ground-glass opacity with focal consolidation in the right upper lobe and enlarged mediastinal lymph nodes. Infectious disease was suspected. Biopsy of the right lung and of a mediastinal lymph node showed non-necrotizing granulomatous inflammation and lymph node tissue with a few carbon deposits. Three months after anti-inflammatory treatment, the above symptoms improved. Chest CT (C1: pulmonary window, C2: mediastinal window) on 12/2019 showed that the consolidation in the right upper lob had fully disappeared with persistent fibrous tissue and diffuse ground glass opacities. However, after six months (06/2020), this patient developed dyspnea again. The chest CT (D1: pulmonary window, D2: mediastinal window) showed significant deterioration with extensive ground-glass opacities, interlobular/intralobular septal thickening, diffuse consolidations in both lungs and enlarged lower paratracheal lymph nodes.

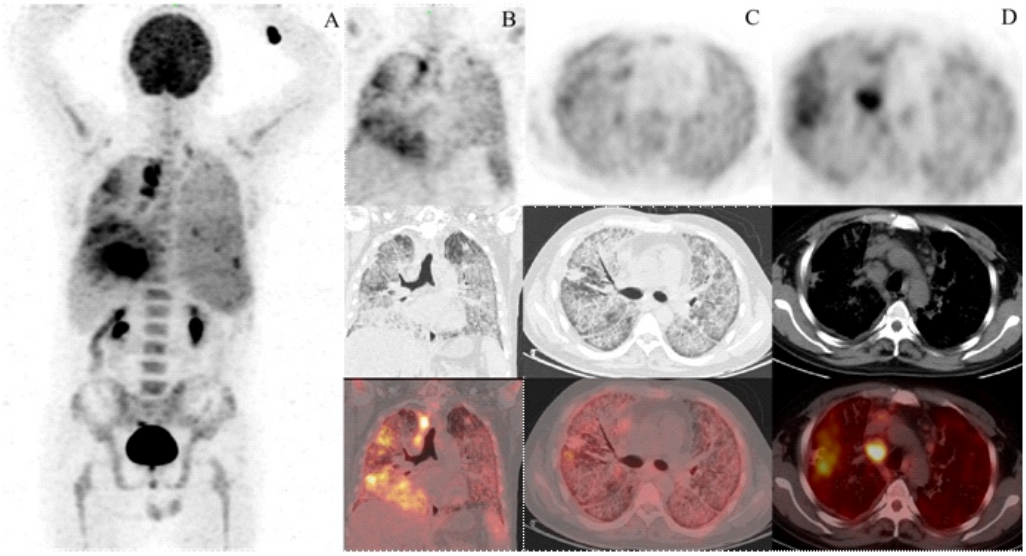

Figure 2. Tumor marker examination showed elevated CEA and NSE (35.1ng/mL, normal range: less than 5ng/mL; NSE: 60.4ng/mL, normal range: less than 20.4ng/mL). Fluorine-18-FDG PET/CT was performed in 06/2020 to exclude the possibility of malignancy. On the MIP image (A), increased ^{18}F-FDG uptake was observed in both lungs and mediastinal area with SUVmax of 13.5 and 9.2, respectively. The coronal and axial images (PET, CT, PET/CT fusion in B and C) showed extensive ground-glass opacities with interlobular/intralobular septal thickening, diffuse consolidations in both lungs, called as 'crazy paving', and enlarged lower paratracheal lymph nodes(d:14x11mm)(PET, CT, PET/CT fusion in D). Computed tomography-guided biopsy of right lower lung showed a large amount of protein exudate in the alveolar cavity and fibrous scar tissue hyperplasia with part of the consolidation in the lung tissue. Immunohistochemical results found positive PAS and D-PAS and negative AB and WS, which supported the diagnosis of pulmonary alveolar proteinosis. EBUS-guided biopsy of mediastinal lymph showed lymph nodes deposited by carbon dust, supporting inflammatory lymph node.

Pulmonary alveolar proteinosis (PAP) is a syndrome characterized by the accumulation of alveolar surfactant and dysfunction of alveolar macrophages[1]. The three main types of PAP are autoimmune, primary, and secondary [1]. The typical chest computed tomography (CT scan) of PAP revealsextensive ground-glass opacities with interlobular/intralobular septal thickening, called as "crazy paving pattern" [2]. Approximately 5% of PAP patients, mostly untreated patients, may present with opportunistic infections [3], which may cause high ^{18}F-FDG uptake on PET/CT, corresponding to this patient.The unique feature in our case compared to previously published is the presence of enlarged mediastinal lymph nodes, due to associated infection[4-6]. This is an interesting case showing the evolving radiologic pattern of PAP, which PET/CT readers should bear in mind, and guide the suitable site for histology which will finally infer the diagnosis.

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