Desquamation Interstitial Pneumonia Presenting as a Solid Mass in the Lung: a Case Report.

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Case Report

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Abstract

Background: Desquamation interstitial pneumonia (DIP) is a rare type of idiopathic interstitial pneumonia. High resolution computed tomography (HRCT) shows ground glass opacities without the peripheral reticular opacities characteristic of usual interstitial pneumonia (UIP). Here we report a case of DIP presenting as a solid mass in the lung, which is rarely described in the literature.

Case presentation: A 77-year-old man presented with dry cough and dyspnea on exertion for one month was admitted to our hospital. The lung CT scan showed there was a pulmonary mass in right lower lung and the patient was scheduled for surgery. He was a sculpture artist, non-smoker and the previous medical history included only mandatory spondylitis without other connective tissue diseases. Nine days post-last CT, the FDG PET/CT demonstrated the mass in the right lower lung was significantly hypermetabolic (SUV value=8.9) but inflammatory exudate could be observed around the mass. We gave antibiotics and performed CT-guided percutaneous lung biopsy. The result showed that there were a large number of macrophages in the alveolar cavity, alveolar epithelial type II cells hyperplasia in some areas and chronic inflammation was noted in the interstitium. Then the desquamative interstitial pneumonia was diagnosed. We gave the patient glucocorticoid therapy, then the patient's symptoms disappeared gradually and the lung CT showed that the pulmonary mass was disappeared.

Conclusions: Our report illustrates the rare presentation of DIP. Furthermore, for the pulmonary mass, even if appeared hypermetabolic on PET/CT, it is still necessary to make a definite pathological diagnosis before operation, so as to reduce the damage to patients.

Background

Desquamation interstitial pneumonia (DIP) is a rare type of idiopathic interstitial pneumonia and the incidence only accounts for about 10–17%[1]. The vast majority (>90 percent) of patients with DIP are smokers, a small percentage of cases are associated with rheumatic diseases[2]. High resolution computed tomography (HRCT) shows ground glass opacities without the peripheral reticular opacities characteristic of usual interstitial pneumonia (UIP)[3]. Here we report a case of DIP presenting as a solid mass in the lung.

Case Report

A 77-year-old man presented with dry cough and dyspnea on exertion for one month was admitted to our hospital. The lung CT scan showed there was a pulmonary mass in right lower lung and the patient was scheduled for surgery. He was a sculpture artist and non-smoker. The previous medical history included only mandatory spondylitis without other connective tissue diseases. His admitted physical examinations revealed vital signs were within normal limits. There were no cyanosis on lips, no clubbing and no superficial swollen lymph node being palpable. Breath sounds were decreased on the right lower side. Physical examinations of cardiovascular system and abdomen were normal. Blood chemistry showed

almost normal white cell count, procalcitonin (PCT) and C-reactive protein (CRP). Erythrocyte sedimentation rate was 77 mm/1st h. The human leukocyte antigen-B27 (HLA-B27) antibody was mildly elevated. Among the autoimmune disease markers (C3, C4, rheumatoid factor, antinuclear antibody, anti-neutrophil cytoplasmic antibodies, anti-cardiolipin antibody, anti-dsDNA, anti-Sm, anti-SSA, anti-SSB, anti-SCL-70, anti-Jo-1, and anti-RNP/Sm antibodies) and immunoglobulin G, A, and M were all normal. Tumor markers (CA199, f-PSA, ProGRP, PSA, CEA, SCC, CYFRA21-1, CA242, CA125, NSE, CA153, CA724, CA50) were only SCCA 12.6 ng/ml (range, 0-2.5 ng/mL). Pulmonary function tests showed that the patient had mild restrictive ventilatory dysfunction and mild diffusion dysfunction. Nine days post-last CT, the FDG PET/CT demonstrated the mass in the right lower lung was significantly hypermetabolic (SUV value=8.9) but inflammatory exudate could be observed around the mass. We gave antibiotics and performed PET/CT-guided percutaneous lung biopsy. The result showed that there were a large number of macrophages in the alveolar cavity, alveolar epithelial type II cells hyperplasia in some areas and chronic inflammation was noted in the interstitium (see Figure 2). Then the desquamative interstitial pneumonia was diagnosed. Methylprednisolone intravenous drip of 40 mg per day for 2 weeks were performed. After that, the treatment was converted to 40 mg of oral prednisone per day. The patient's symptoms disappeared gradually and the lung CT showed that the pulmonary mass was decreased in size (see Figure 1). The patient's symptoms were resolved, and he was discharged with 40 mg of oral prednisone per day. The dose of prednisone was reduced by 5 mg weekly until to 20 mg. Then the dose was reduced by 5 mg monthly until to 10 mg and maintained for three months. The total course of treatment was half a year. The lung mass significant shrink was observed after discharged about one month and eventually disappeared. (see Figure 1).

Discussion

The exact etiology of the DIP is uncertain but 58-91% of DIP cases are linked with cigarette smoking[4, 5]. In addition, the DIP may be associated with harmful irritating substances from environmental/occupational (such as asbestos, talc, graphite, silica and aluminum), systemic disease virus infection and drugs [6, 7]. The main histopathological feature of DIP is a large number of macrophages in alveoli, which were usually diffusely distributed throughout the pulmonary acini within alveoli, and proliferation of pneumocytes over the course of alveolar septa [8]. Although there may be mild infiltration of inflammatory cells in the alveolar septum, the alveolar structure generally remained normal. The infiltrating inflammatory cells include a small number of eosinophils, accompanied by fibroblast proliferation. The rare changes are mild fibrosis, but severe fibrosis and honeycomb like lesions are very rare.

Pathological diagnosis is the "gold standard" for the diagnosis of DIP. Methods of lung biopsy included transbronchial lung biopsy (TBLB), CT-guided percutaneous needle biopsy, transbronchial cryobiopsy and surgical lung biopsy. Lung specimens from TBLB are too small, which can't fully reflect the scope and degree of lung lesions and satisfy the diagnosis of interstitial lung disease. The specimens from surgical lung biopsy can meet the diagnostic requirements of interstitial lung disease, but the surgical lung biopsy is costly and traumatic, which is not easy for patients to accept. Transbronchial cryobiopsy or CT-guided

percutaneous lung biopsies have fewer healthcare costs, fewer traumatic, and the specimens can meet the diagnostic requirements of interstitial lung disease, which is easy for patients to accept. This patient underwent percutaneous lung biopsy and the result showed that there were a large number of macrophages in the alveolar cavity and chronic inflammation was noted in the interstitium, so it could be diagnosed as DIP. At the same time, the patient had no history of smoking or second-hand smoke, which was rare in the literature. The patient was a sculptor. Because the environmental or occupational exposure may be one of an etiological risk factors, it is speculated that dust inhalation may be one of the inducing factors in this case. However, because the specimens were relatively small, it was impossible to know whether there was dust deposition in other lesions regions.

The special feature of this case was lung CT, which showed a single solid mass in the right lower lung, and then rapidly progressed with peripheral infection. However, the typical imaging manifestation of DIP is diffuse ground glass shadow in both lungs, which is located under the pleura. The lower lung is more than the upper lung, and some are accompanied by linear opacities and reticulation in the base [9]. Honeycomb appearance is uncommon. Therefore, the imaging findings of this case were different from the typical imaging findings of DIP, which were rarely reported in the literature. These imaging findings were often misdiagnosed as pulmonary malignant tumor because of PETCT demonstrated hypermetabolism and the patients were usually treated by surgery. Therefore, for the pulmonary mass, even if PETCT indicates hypermetabolic lesions and highly suspected tumor, the pathological diagnosis of preoperative mass is still very important to avoid irreversible damage to patients.

The main treatment is to quit smoking and glucocorticoid therapy and some cases become spontaneous remission after quitting smoking. Glucocorticoids have been reported to be beneficial in case series, although improvement may be temporary [3]. Environmental/occupational factors are also suspicious etiology of DIP [6, 7]. That DIP improved in two cases after leaving the workplace without steroid treatment was reported by Loughheed et al [7]. Therefore, It is reasonable to terminate any possible environmental risk factors. Initial glucocorticoid therapy consists of 40-60 mg/day for 6 weeks, then decreased gradually and stopped within 6-9 months. Glucocorticoid therapy of this case: intravenous methylprednisolone 40mg for 2 weeks, oral prednisone 40mg for 3 weeks, and then decrease 5 mg weekly until to 20 mg. Then the dose was reduced by 5 mg monthly until to 10 mg and maintained for three months. The total course of treatment was half a year. The case clinical symptoms and radiology were significant improvement after steroid treatment.

Conclusions

The imaging findings of such a case are rarely described in the literature. There is no smoking history and it is easy to be misdiagnosed as tumor. However, all of these clinical characteristics mentioned above are uncommon. Therefore, we still know little about this rare interstitial lung disease, and we still need more clinical cases, more complete clinical imaging and pathology to enrich our understanding of the disease. At the same time, for the pulmonary mass, even if appeared hypermetabolic on PET-CT, it is still

necessary to make a definite pathological diagnosis before operation, so as to reduce the damage to patients.

Abbreviations

DIP: Desquamation interstitial pneumonia; HRCT: High resolution computed tomography; UIP: usual interstitial pneumonia; PCT: procalcitonin; CRP: and C-reactive protein; HLA-B27: human leukocyte antigen-B27; TBLB: transbronchial lung biopsy;

Declarations

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Authors' contributions

LYH designed the study and revised the manuscript. LW conducted clinical data collection and analysis and drafted the manuscript. WWY, SYH and LF conducted clinical data collection and analysis. The authors read and approved the final manuscript.

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Ethics approval and consent to participate

Not applicable

Consent for publication

Written informed consent was obtained from the patient for publication. All authors are in agreement for the publication of the study.

Competing interests

The authors declare that they have no competing interests.

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Figures

Figure 1

The patient's symptoms disappeared gradually and the lung CT showed that the pulmonary mass was decreased in size

Figure 2

The result showed that there were a large number of macrophages in the alveolar cavity, alveolar epithelial type II cells hyperplasia in some areas and chronic inflammation was noted in the interstitium

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