



Case report

Successful treatment of suspected organizing pneumonia in a patient without typical imaging and pathological characteristic: A case report



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ABSTRACT

Introduction: Organizing pneumonia (OP) is a clinicopathological entity characterized by granulation tissue plugs in the lumen of small airways, alveolar ducts, and alveoli. Diagnosis of OP needs the combination of clinical features, imaging and pathology. But it occurs often that there are no typical pathological features to support the diagnosis, which poses a challenge for clinicians' diagnosis and treatment. We diagnosed a case of OP without typical imaging and pathological characteristic and treated successfully. Finally we confirmed the pathological diagnosis.

Conclusions: Not every OP case is supported by pathological evidence and typical imaging changes. It is important for us to judge and decide the diagnosis according to clinical experience.

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1. Introduction

Organizing pneumonia (OP) is a clinicopathological entity characterized by granulation tissue plugs in the lumen of small airways, alveolar ducts, and alveoli [1]. The study demonstrated a significant increase in the incidence of organizing pneumonias over the last 20 years [2].

The disease usually develops after a prodrome of a flu-like syndrome associated with fever, fatigue, nonproductive cough (70%), mild dyspnea (65%), and weight loss (60%), which lasts for several weeks [3].

There is no specificity in the imaging features of OP. The most common radiological pattern of COP (cryptogenic organizing pneumonia) is peripheral, bilateral, diffuse alveolar opacities, which is often confused with infectious pneumonia or cancer. Air bronchograms and bronchial dilatation are often observed. Besides, solitary pulmonary nodule and diffuse pulmonary infiltration are found in some cases [4].

Although the typical clinical and radiological features may

suggest the diagnosis of OP, histological confirmation is essential for the diagnosis. So the diagnosis of OP needs the combination of clinical features, imaging and pathology [5,6].

However, it occurs often that there are no typical pathological features to support the diagnosis because of the limitation of biopsy methods. This poses a challenge for clinicians' diagnosis and treatment.

We report a case of suspected OP without typical imaging and pathological characteristic, and treated successfully.

2. Case presentation

A 65-year-old man presented with a one-month history of cough, sputum and chest pain. He was a current smoker (20 cigarettes a day for 40 years) and reported no occupational exposure. His medical history was unremarkable. Symptoms persisted without medication.

At admission, temperature was 36.5 °C, respiratory rate was 18/minute, and moist rales were heard in left lung.

Arterial blood gases at 21% FiO₂ (fraction of inspiration O₂) showed PaO₂ (partial pressure of oxygen) 69 mmHg, PaCO₂ (partial pressure of carbon dioxide in artery) 36 mmHg, pH 7.43.

Laboratory parameters were as follows:

White blood cell count of 7.99×10^9 cells/l [normal range (NR), $4-10 \times 10^9$ cells/l], differential neutrophil count of 69% (NR, 40–70%) and a C-reactive protein (CRP) level of 36.38 mg/l (NR, <10 mg/l). Serum electrolytes, renal and liver function tests were normal, as well as serological tests for tumor markers, Legionella

Abbreviations: OP, Organizing pneumonia; COP, Cryptogenic organizing pneumonia; CRP, C-reactive protein; CT, Computed tomography; PET-CT, Positron emission tomography-computed tomography; FiO₂, Fraction of inspiration O₂; PaO₂, Partial pressure of oxygen; PaCO₂, Partial pressure of carbon dioxide in artery.

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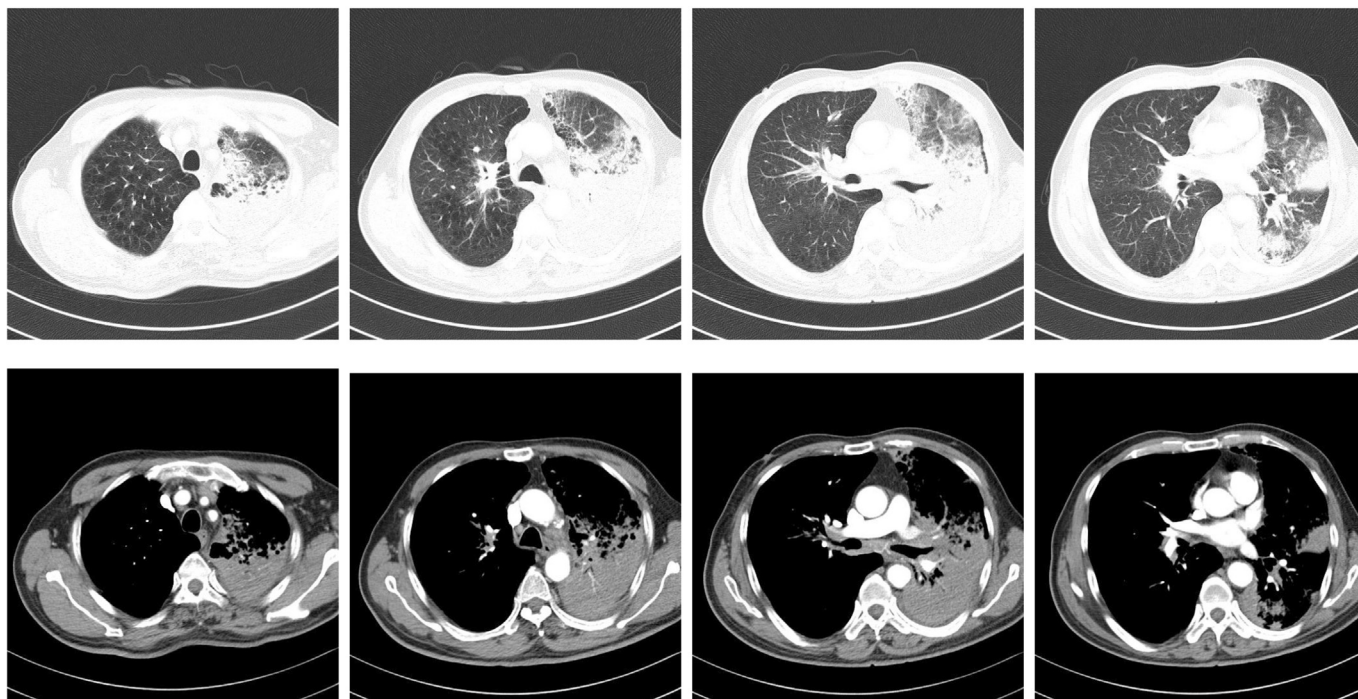


Fig. 1. CT images at admission. At admission, CT scan showed demonstrated extensive shadows of high density in left lung, inburst into mediastinum, with bronchial ventilation sign, pleural thickening, and enlargement of lymph nodes in hilum and mediastinum. There was no obvious abnormal enhancement.

pneumonia, Mycoplasma, Chlamydia, Coxiella psittaci, hepatitis C and B viruses, and HIV. Antinuclear antibody, perinuclear anti-neutrophil cytoplasmic antibody (P-ANCA), cytoplasmic-ANCA, and cyclic citrullinated protein antibody tests were negative.

High-resolution computed tomography (CT) demonstrated extensive shadows of high density in left lung, with bronchial ventilation sign, pleural thickening, and enlargement of lymph nodes in hilum and mediastinum. No abnormal enhancement was showed (Fig. 1). Lung cancer was considered by imaging diagnosis, without the exception of lymphatic metastasis.

Bronchofibroscope showed mucosa thickening and tracheobronchial stenosis in left lung (Fig. 2). Transbronchial biopsy indicated interstitial fibrous tissue hyperplasia and chronic inflammatory cell infiltration (Fig. 3A).

After 10 days antibiotic therapy with amoxicillin/clavulanic acid and roxithromycin, the symptom improved, but there was no significant change of CT sign. Percutaneous lung biopsy was arranged. Percutaneous biopsy from the left lower lobe also indicated interstitial fibrous tissue and chronic inflammatory cells without specificity (Fig. 3B).

One month later, the patient completed PET-CT (positron emission tomography-computed tomography) scan outside the hospital. The results showed high density shadow in left lung with increased metabolism, without obvious change in the range than before (Fig. 4). PET-CT indicated inflammation, without the exception of adenocarcinoma. Percutaneous biopsy was suggested again. The second percutaneous biopsy results was the same as above, showed interstitial fibrous tissue and chronic inflammatory cells.

Next, the patient visited Shanghai Chest Hospital, did EBUS-TBNA (endobroncheal ultrasonography-transbronchial needle aspirations) check. Pathology still suggested inflammatory. There were no characteristic pathological changes and definite diagnosis.

After four times pulmonary biopsies, there was no definitive pathological evidence. Also, there was no characteristic imaging finding. Other infectious diseases were less likely to be diagnosed based on the patient's clinical characteristics, tumor was the main differential diagnosis, but no tumor cells were found after multiple biopsies. The patient and his family defused to do further open lung biopsy, and the treatment could not be delayed. According to the

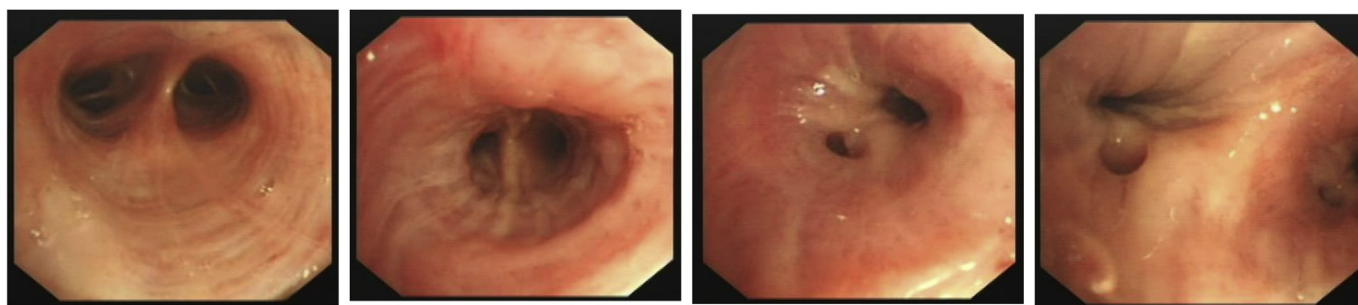


Fig. 2. Bronchofibroscope images. Bronchofibroscope was performed after CT scan. Bronchofibroscope images showed mucosa thickening and tracheobronchial stenosis in left lung. No neoplasm and caseous necrosis were found in the lumen.

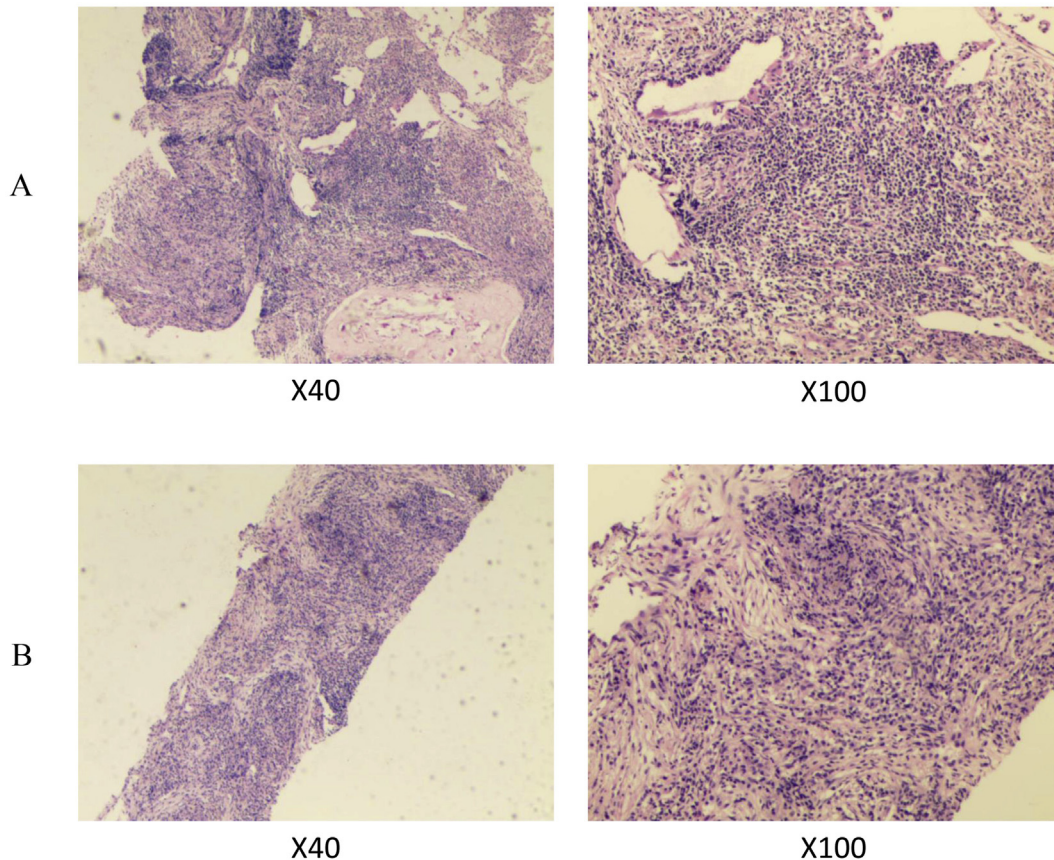


Fig. 3. Pathological images (A) Pathological images of biopsy by Bronchofibroscope, indicated interstitial fibrous tissue hyperplasia and a large number of chronic inflammatory cell infiltration. (B) Pathological images of biopsy by percutaneous lung puncture from the left lower lobe, also indicated interstitial fibrous tissue and many chronic inflammatory cells without specificity.

clinical features, we diagnose OP in this case after excluding other diseases and gave corticosteroid therapy.

Oral corticosteroid therapy was started at 0.75mg/kg/day. Improvement of CT sign was observed after 1 month treatment, the shadow had dramatically decreased. Steroids were tapered over 6 months and lung CT scans significantly improved (Fig. 5). But new small shadow appeared, as the sign of recurrence (Fig. 5).

Once again, we persuaded the patient to do another biopsy. The patient underwent percutaneous biopsy again, and finally, the pathological manifestation indicated OP by finding granulation tissue plugs in the lumen of alveoli (Fig. 6). So far, the patient has been followed up for one year.

Informed consent was signed by the patient.

3. Discussion

Usual clinical and imaging presentations of OP have been well characterized in large series in the literature. The clinical course is usually subacute in 50- to 60-year-old patients. The disease usually develops after a prodrome of a flu-like syndrome associated with fever, fatigue, nonproductive cough, mild dyspnea, and weight loss, which lasts for several weeks [7]. Sparse crackles and bronchial breath sounds can be heard in areas of air space consolidation.

We found there was no specificity on OP's symptoms; it's hard to suspect the diagnosis of OP through cough, fever, fatigue or dyspnea. The clinical features are easily confused with inflammation. So anti-infection is the usual early treatment.

The common CT scan signs of OP are patchy air space

consolidation areas with a migratory course and ground glass opacities predominantly in the lung periphery [8,9].

However, many other appearances have been reported, consisting of solitary or multiple nodules, focal pneumonia [10], peribular consolidations, curved bands of consolidation, atoll sign [11,12].

In this case, extensive shadows of high density were showed, with bronchial ventilation sign, pleural thickening, and enlargement of lymph nodes in hilum and mediastinum. It was different to distinguish OP from pneumonia or cancer through CT scan signs. And there were no obvious imaging changes in short time.

The prominent histological finding in OP is patchy involvement of the pulmonary parenchyma by fibromyxoid, polypoid plugs of granulation tissue (also known as Masson bodies) within the alveoli and occasionally within the bronchioles (in which case the term bronchiolitis obliterans organizing pneumonia may be applied) [13,14].

The pathology results indicated interstitial fibrous tissue and chronic inflammatory cells without specificity. After four times pulmonary biopsies, there was no definitive pathological evidence. At first, we could not make a definite diagnosis of OP. The patient and his family defused to do further open biopsy, and the treatment could not be delayed. This is the difficulty for us in this case. Finally the last pathological evidence confirmed the diagnosis of OP.

The standard treatment for OP is corticosteroids. Corticosteroid therapy results in complete recovery in up to 80% of patients within a few weeks to 3 months [15]. The disease is persistent in the remainder [16].

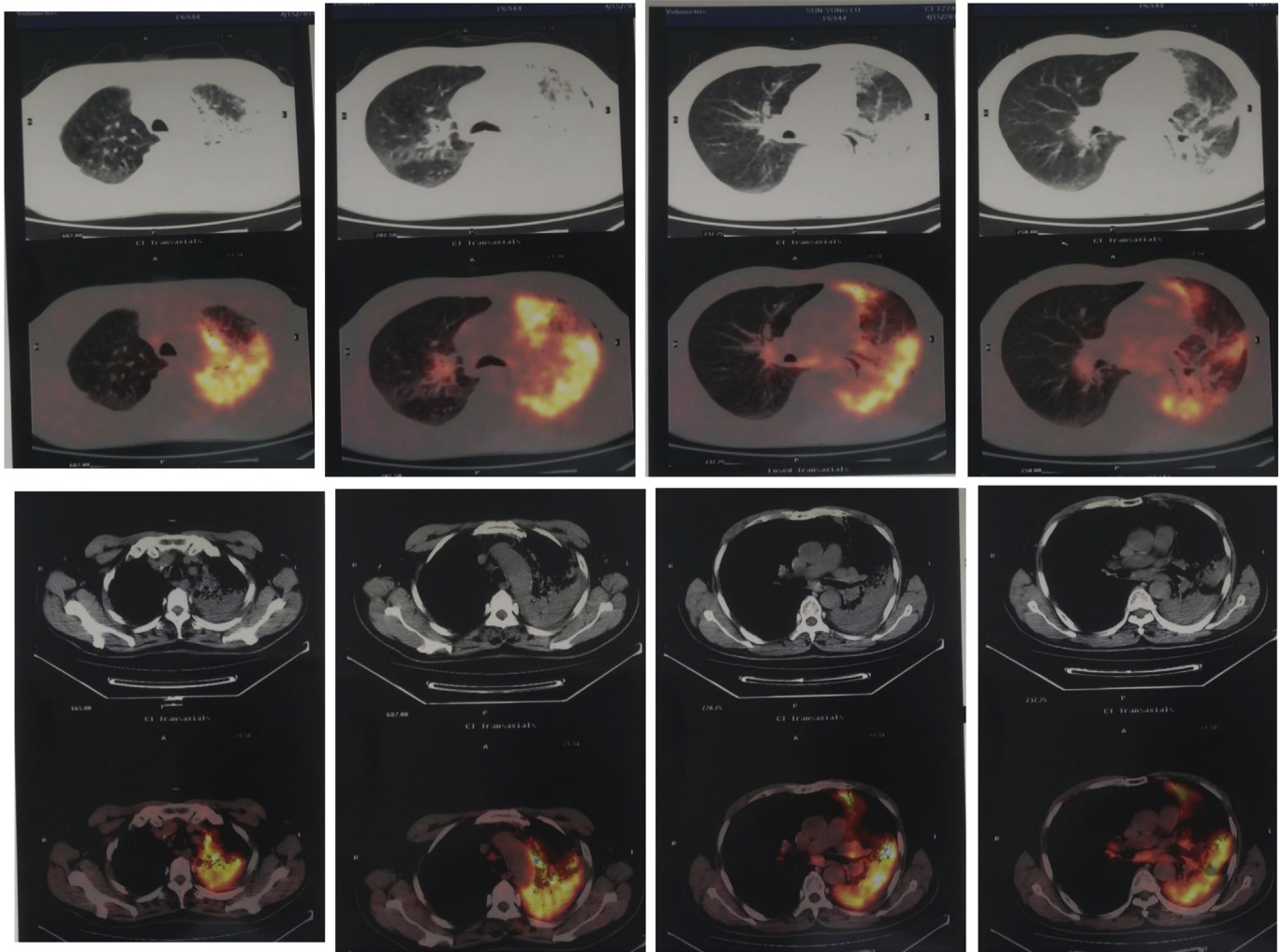


Fig. 4. PET-CT images after one month. After 10 days antibiotic therapy with amoxicillin/clavulanic acid and roxithromycin, the symptom improved, medication discontinued. PET-CT scan was arranged one month after admission. PET-CT showed high density shadow in left lung with increased metabolism, without obvious change than before.

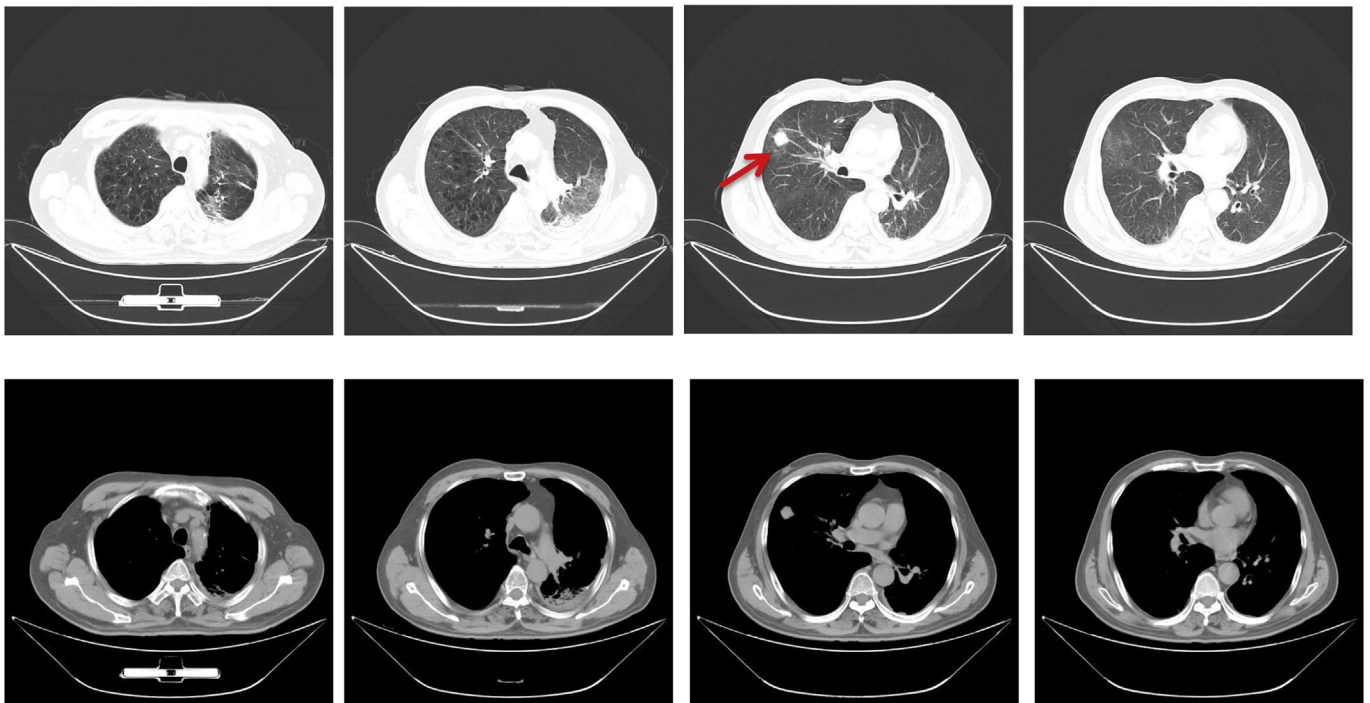


Fig. 5. CT images after corticosteroid treatment. After one month oral corticosteroid therapy, the shadow of left lung had dramatically decreased than before. New small shadow appeared in right lung (red arrow). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

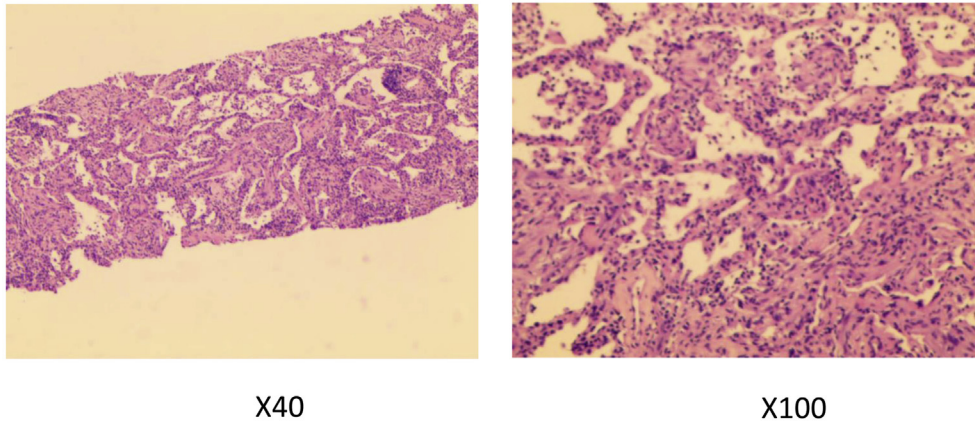


Fig. 6. Pathological images. Biopsy by percutaneous lung puncture from the right upper lobe (the new lesion appeared in Fig. 5), interstitial fibrous tissue and many chronic inflammatory cells, granulation tissue plugs in the lumen of alveoli.

Relapses are common, ranging from 13% to 58%, and are usually associated with tapering or withdrawal of corticosteroid treatment [17,18].

In this case, the lesions were significantly improved after oral corticosteroid therapy. Also, new small shadow showed the sign of recurrence.

In conclusion, the clinical, imaging and pathological characteristics of organizing pneumonia are now well established, the diagnosis of OP needs the combination of clinical features, imaging and pathology. But not every OP case is supported by pathological evidence and typical imaging changes. It is important for us to judge and decide the diagnosis according to clinical experience.

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