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Case report

Emphysema mimicking interstitial lung disease: Two case reports

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A B S T R A C T

Honeycombing in general is a sign of severe end-stage fibrosis. Here we present two cases, where the combination of emphysema, acute inflammation and pulmonary embolism gave an appearance of honeycombing seen in pulmonary fibrosis. HRCT interpretation in the evaluation of acutely ill patients with pulmonary infection is a challenge. Our case reports emphasize the importance of a multidisciplinary approach, when it comes to patients with suspected complicated pulmonary diseases. At the same time they give very realistic examples of the challenges found in diagnosing patients with simultaneous acute and chronic pulmonary diseases.

Introduction

Honeycombing is defined as clustered cystic airspaces, usually with a diameter in the order of 0.3–1.0 cm [1]. Honeycombing is localized in the subpleural areas of the lung and is characterized by well-defined walls, which are often thick [2]. Honeycombing can be seen in a variety of interstitial lung diseases. The appearance of honeycombing and/or traction bronchiectasis predominantly located in the upper or mid fields in general is a sign of severe end-stage fibrosis. When a reticular pattern and honeycombing predominantly is localized in the inferior aspects definite UIP most probably is the cause [1]. Other pathologies in the lung parenchyma as cystic lung diseases and emphysema, especially the paraseptal type may mimic honeycombing, and sometimes it is a challenge by CT to make a definitive diagnosis [3]

Therefore, a multidisciplinary evaluation that combines the medical history, clinical appearance, high resolution computered tomography (HRCT) and histopathology is mandatory for the classification of interstitial lung diseases.

We present two case stories, in which the combination of emphysema, acute inflammatory changes and pulmonary embolism were erroneous described as the honeycombing seen in pulmonary fibrosis.

Case story 1

A 69-year old woman, previous smoker (30 pack years (PY)) presented with dry cough, headache, dyspnoea at rest and general malaise. The symptoms appeared 2½ weeks prior to admission, and. Six days treatment with antibiotics and corticosteroids did not have any effect. Before admission, the patient had never experienced pulmonary symptoms. At admission, the patient had a temperature of 39.5 °C, normal blood pressure, saturation of 92% and basal crackles at lung auscultation. Blood samples showed a mild anaemia, elevated C-reactive protein 174.5 mg/ml (CRP) and normal leucocytes (LKC). No deep venous thromboembolism was present on clinical examination. Echocardiography showed a normal left ventricular ejection fraction of 60%, a borderline dilated and hypertrophic left ventricle and no signs indicating increased pulmonary pressure. Tests for atypical pneumonia, tuberculosis and fungal infections were negative.

Intravenous broad-spectrum antibiotics were initiated but due to continuing fever and an elevated CRP, a chest X-ray and an HRCT were performed. On the chest radiograph, blurry bilateral less than 1 cm large consolidations were present predominantly on the left side.

HRCT revealed diffuse ground glass opacities, thickening of interlobular septae, traction bronchiectasis, localized subpleural honeycombing, mosaic perfusion pattern in the apical areas of the lung parenchyma and mediastinal lymphadenopathy.

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Radiologically, a diagnosis of pulmonary fibrosis and infection was made (Fig. 1A and 1B). CT angiography did not demonstrate pulmonary embolisms. Chronic eosinophilic pneumonia (CEP) or fibrotic non-specific interstitial pneumonia (NSIP) was suggested as an underlying interstitial lung disease with simultaneous infection.

Prophylactic low-molecular-weight-heparin was administered together with oxygen, intermittent continuous positive airway pressure (CPAP) and high dose corticosteroid. Despite treatment, the patient's condition deteriorated and respiratory support including high frequency oscillation and NO treatment was started at the intensive care unit, where, as a consequence of multi-organ failure, she succumbed after 21 days of hospitalisation.

A post mortem autopsy showed widespread pulmonary infarction of different age with haemorrhagic necrosis. There were limited areas with signs of previous infection, and diffuse extensive emphysema, but no fibrosis. The cause of death was determined to be caused by haemorrhagic pulmonary infarction, caused by widespread bilateral pulmonary thrombosis.

Case story 2

An 81-year old male, previous smoker (100 PY) with known paroxysmal atrial fibrillation was admitted because of cough, haemoptysis, myalgias, arthralgias and skin suggilations. The patient presented with a temperature of 38.8 °C and basal crackles at lung auscultation. Arterial blood gas analysis showed P_aO_2 6.0 kPa, SaO2 87%, compensatory hyperventilation with P_aCO_2 3.8 kPa, pH 7.47 and lactate 2.0 mmol/L. CRP was elevated to 57 mg/L, LKC $16.1 \cdot 10^9$/L, INR >10 International units (IU) and a low haemoglobin of 5.4 mmol/L. An electrocardiogram found a high frequency atrial fibrillation (132 beats/min).

Chest X-ray showed bilateral extensive consolidation, most pronounced at the left side. A contrast enhanced CT of the chest performed at the local hospital was interpreted as a combination of infection and extensive honeycombing (Fig. 1C) and a diagnosis of infection and pulmonary fibrosis was made.

Treatment with intravenous vitamin K and blood transfusions 2nd generation cephalosporin and macrolide antibiotics, inhaled bronchodilators, corticosteroid and furosemide were initiated.

After 3 days, the patients' general condition improved and the fever dropped but there was still minor haemoptysis and desaturation to 85–90% in spite of oxygen supplementation with 8 L/min.

After 17 days, the patient was discharged from the hospital with long term oxygen treatment and referred to a tertiary interstitial lung disease centre for management of the suspected interstitial lung disease.

At an MDT conference re-evaluation of the CT showed emphysema, pleural plaques, and bilateral extensive consolidations. No signs of honeycombing could be seen. A new HRCT showed complete regression of the consolidated areas and extensive emphysema (Fig. 1D). Spirometry confirmed that the patient had chronic obstructive pulmonary disease with a forced expiratory volume in 1 s (FEV1) of 1.4 L (54% of expected), and a FEV1/forced vital capacity (FVC) ratio of 45%.

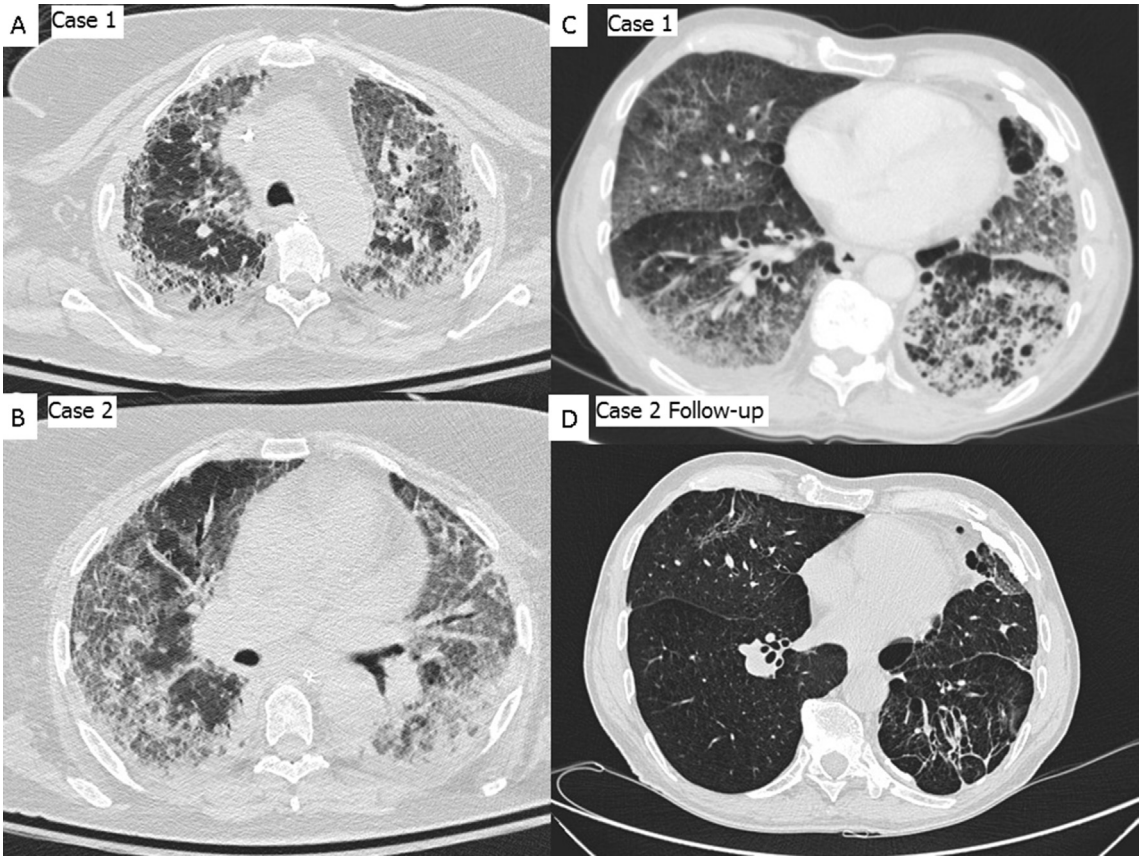

Fig. 1. A and B. CASE 1. Computed tomography upper and lower image. Mosaic perfusion pattern is seen in the upper areas of the lung (a). Diffuse areas with ground glass appearance, especially in the peripheral part of the lungs is evident. Thickening of the intra and interlobular septae and traction bronchiectases is seen, which was the main reason for suggesting ILD. Several consolidated areas mostly localized subpleural may represent abscesses or infarctions. In the spared parenchyma emphysematous changes appeared, and changes mimicking honeycombing probably represent paraseptal emphysema. C. CASE 2. Computed tomography. Emphysema can be seen in the spared lung parenchyma. Furthermore, large consolidated areas and peribronchial thickening is present. Bronchiectasis most evident in the left lower lobe probably was interpreted as honeycombing. D. CASE 2 FOLLOW-UP. At follow-up HRCT showed extensive emphysema. Bronchiectasis in the left lower lobe is evident.

Discussion

Fibrotic interstitial lung diseases are rare, and the usual symptoms with slowly progressive dyspnoea and cough are rather unspecific. Less than 5% of the patients initially present with an acute exacerbation [1]. HRCT is performed as part of the acute diagnostic set-up in these patients, while lung biopsies often are found too risky due to the severity of the disease and the risk of eliciting or worsening of an acute exacerbation of idiopathic pulmonary fibrosis (IPF). Under these circumstances, the diagnosis rests on a combination of the radiologic and the clinical evaluation. In the presented case stories the patients clinically presentation and CT findings were suggestive of severe pulmonary infection. However, initial CT findings did that fibrosis was also suspected.

The combination of severe emphysema and infection or pulmonary infarction mimicking honeycombing has only seldom been described, but the erroneous description of honeycombing was the primary and sole reason for the ILD diagnosis in our two cases.

Thickening of the intra- and interlobular septae due to infectious disease or lung parenchyma infarction may mimic honeycombing, although definite honeycombing never is presented. Honeycombing defined as clustered cystic airspaces with well-defined walls is typically located in the subpleural areas [1]. Normally, honeycombing is a sign of permanent fibrotic changes of the lung and thus reflects end-stage parenchymal destruction. It is not only seen in the fibrotic idiopathic interstitial pneumonias, but also in diseases such as hypersensitivity pneumonitis, asbestosis and sarcoidosis [1,4,5].

CEP and NSIP were suggested as possible underlying interstitial lung disease in the first patient. The typical HRCT pattern of NSIP is bilateral reticular abnormalities with a basal predominance, subpleural sparing and traction bronchiectasis and while honeycombing is rare, areas with ground glass attenuation may be extensive [1]. HRCT findings in CEP is classically described as peripheral localized areas with ground glass appearance, the so-called “reverse batwing appearance” [4]. The same pattern can also be seen in cryptogenic organizing pneumonia, pulmonary vasculitis, aspiration, pulmonary contusion and pulmonary infarction [4,6]. Venous thromboembolism could not be identified on CT angiography case 1. No doubt the patient at the time when CT was performed also had a severe pulmonary infection. Infections may also result in mosaic perfusion pattern, and the time delay between the CT scan and the autopsy may explain the different findings. Some of the pulmonary embolus or infarction seen at autopsy can have arrived during the stay in the intensive care unit.

Hunninghake et al. [7] examined the diagnostic value of HRCT for IPF and NSIP diagnosis. Four chest radiologists ranked the HRCT as “certain, uncertain, or unlikely” consistent with IPF. When the radiologists felt confident in the IPF diagnosis, the positive predictive value (PPV) and specificity was 96% and 95% respectively. However when the uncertain cases were not excluded, the PPV and specificity were lower. Lower lobe/basal predominant honeycombing and upper irregular lines were the findings most closely associated with a histopathologic pattern of UIP [7,8].

Morgenthau compared Hunninghake's results with studies concerning the accuracy in HRCT diagnosis of other IIP's specially NSIP. These studies showed a much lower specificity of about 50–60% [5,9–11].

Akira et al., observed that concomitant emphysema complicated the radiologic distinction between UIP and NSIP. If emphysema was present, an accuracy of 44% was observed compared to an accuracy of 71% in patients without emphysema [12]. In 9/54 patients, a definite IPF diagnosis based on HRCT and transbronchial biopsies were changed to emphysema following thoracoscopic biopsy [8].

In the presented case 2 the extensive emphysematous changes although visible on CT is obscured by changes reflecting infections. The severe parenchymatous changes seen on CT in the presented case 1 almost totally obscure the dramatic emphysematous findings found at autopsy.

In conclusion, HRCT interpretation in the evaluation of the acutely ill patients with clinically and radiologically obvious pulmonary infection is a challenge. The CT changes induced by the infection may obscure even severe emphysema. Furthermore, erroneous suspicion of interstitial disease at CT performed during the acute stage is easy to make. A follow-up HRCT after the infection is treated is essential for the diagnosis of interstitial disease.

A multidisciplinary team discussion is advisable when patients with complicated pulmonary disease are considered, especially when several pulmonary diseases are present simultaneously.

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