A CT emphysema pattern in a non-smoker classical guitar teacher

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ABSTRACT

Background: Hypersensitivity pneumonitis (HP) is a complex syndrome due to an inflammatory response of the lungs to many aerosolized antigens. HP can be divided into acute, subacute, and chronic phenotypes, which frequently overlap.

Case presentation: We describe the case of a non-smoker man admitted to the hospital for flu-like symptoms who presented emphysema as an evolution of HP. The diagnosis of HP was proposed by a multidisciplinary team based on the clinical/functional data and CT findings; two chest CTs were compared: a CT was performed during the hospitalization of the patient and a CT had been performed 5 years before. The diagnosis was confirmed through a lung biopsy. Examining the patient history, a domestic exposition to pigeons and poultry was revealed.

Conclusion: HP can lead to emphysema instead of fibrosis. This atypical evolution of HP should be known by clinicians and radiologists in order to suspect it and promptly start the correct diagnostic workup.

Keywords: Hypersensitivity pneumonitis, pulmonary emphysema, multidisciplinary group of interstitial lung disease consult, high-resolution computed tomography, surgical lung biopsy.

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Background

Hypersensitivity pneumonitis (HP) is a complex syndrome caused by repeated inhalation of and sensitization to numerous aerosolized antigens which cause an immune response in the lung parenchyma [1].

Clinical presentation and disease progression are highly variable and depend on factors such as intensity and length of exposure to the causative antigen and host factors[1-5].

Chronic HP generally leads to pulmonary fibrosis, but rarely, pulmonary emphysema can be a consequence of chronic HP and can be assessed on radiological examinations such as High-resolution Computed Tomography (HRCT). This possible evolution should be known by the radiologists and clinicians and chronic HP should be suspected when other clinical or radiological findings of the disease are present, especially in non-smokers.

This case report shows a CT emphysema pattern in consequence of HP.

Case Presentation

A 60-year-old Caucasian man was admitted to our hospital with dyspnea, fever, productive cough with purulent sputum, and hypoesthenia. He did not complain about chest pain.

The patient did not have a significant occupational history, since he was a classic guitar teacher and he had never smoked.

Neither arthritic symptoms nor skin or mucosal lesions were present.

Physical examination revealed a hyperinflated chest with rare bibasilar inspiratory crackles on auscultation. There was no cyanosis or finger clubbing.

The respiratory rate of the patient was 22 breaths/min while his heart rate was within normal limits.

The arterial blood gas test showed: pO_2 = 58 mmHg, pCO_2 = 40 mmHg, and pH = 7.4.

Pulmonary function tests (PFTs) demonstrated a pattern of mild obstruction and low diffusing capacity of the lungs for carbon monoxide (DLCO = 38%).

Immunological laboratory tests revealed a positive Rheumatoid Factor, while IgE and α1-antitrypsin were negative.

Chest radiographs were normal (Figure 1).

An HRCT was performed and conspicuous centrilobular emphysema, prevalent in the upper lung regions, and diffuse ground-glass opacities, in the inferior lung regions, were seen (Figure 2).

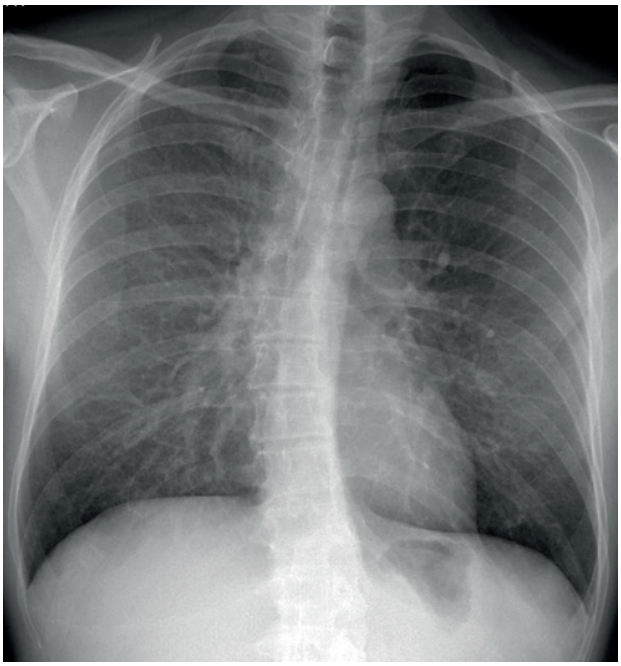

Figure1. *Chest radiography, postero-anterior view. No abnormalities at chest radiography.*

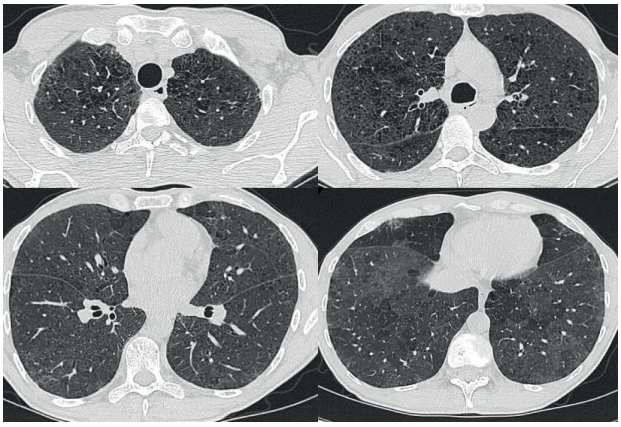

Figure 2. *Chest CT, axial image, lung window. A large amount of centrilobular emphysema is visible. It is more evident in the upper lobes and is associated to diffuse ground-glass opacities in the inferior lobes.*

The patient also brought us an HRCT performed 5 years before, which showed bilateral diffuse ground-glass opacities and a mosaic perfusion pattern, without centrilobular emphysema (Figure 3).

The case was discussed by a Multidisciplinary Group of Interstitial Lung Disease consult, composed of pulmonologists, radiologists, and pathologists.

They evaluated the patient's clinical and functional data and both the CT scans. Confirmation through a surgical lung biopsy of the lingula was asked.

Based on the radiological findings of the oldest HRCT, in particular, the presence of bilateral areas of ground-glass attenuation and centrilobular nodular opacities, two diseases were taken into consideration in the differential diagnosis: respiratory bronchiolitis-interstitial lung disease (RB-ILD) and chronic HP. However, the anamnesis

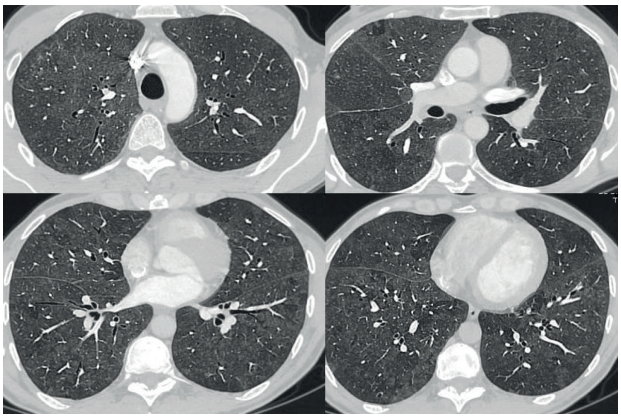

Figure 3. *Five years earlier chest CT, axial image, lung window. Bilateral ground-glass opacities and mosaic perfusion are present.*

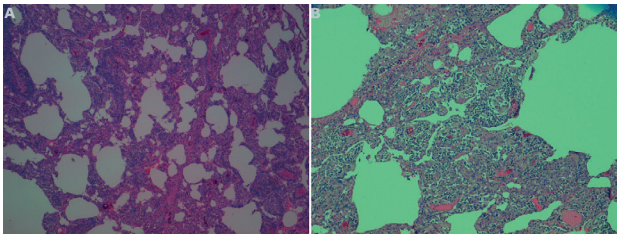

Figure 4. *Lingular biopsy of the lung, histological findings (hematoxylin and eosin stain). (A) Diffuse chronic interstitial inflammatory infiltrates with peribronchiolar accentuation and emphysema (magnification 4×). (B) Chronic interstitial inflammation and poorly formed granulomas (magnification 10×).*

of the patient did not reveal a smoking history or an exposure to aerosolized antigens which could cause HP. A surgical lung biopsy was necessary to reach the final diagnosis.

Histological findings were consistent with chronic HP: diffuse chronic interstitial inflammatory infiltrates, mainly lymphocytes, and plasma cells with peribronchiolar distribution, poorly circumscribed interstitial non-necrotizing granulomas, individual giant cells containing cholesterol, foamy macrophages, and scattered areas of organizing pneumonia. There was also a prominent component of centrilobular emphysema (Figure 4).

The interstitial inflammatory infiltrates were responsible for the ground-glass opacities which are visible on the HRCT; the mosaic perfusion pattern that was visible in the oldest HRCT was a consequence of the peribronchiolar inflammation which caused air-trapping.

Emphysema was interpreted as an evolutive pattern of chronic HP since the patient had never smoked.

During a more accurate anamnesis, the patient revealed a 5-year domestic exposure to pigeons and a 10-year domestic exposure to poultry.

The symptoms which led the patient to the emergency department could be referred to as an acute exacerbation.

The patient was treated with glucocorticoids, bronchodilators, and levofloxacin, improved in 6-7 days and was discharged. He was encouraged to avoid avian exposure.

Discussion

HP is a complex syndrome caused by the repeated inhalation of and sensitization to aerosolized antigens (primarily organic dust of animal or vegetable origin, rarely chemicals) which cause a delayed immune response in the lung parenchyma [1,2]. Thermophilic Actinomycetes species in moldy hay (responsible for farmer's lung) and proteins in avian feces and feathers (which induce bird fancier's lung) are the most common [3].

Historically HP has been classified as acute, subacute, and chronic; however, the clinical presentation of HP and its course are variable, depending on the intensity and duration of the exposure to the causative antigen, the nature of the antigen, and host factors [1-5]. Smoking is considered a protective factor [2].

Acute HP is characterized by the brusque onset of flu-like and respiratory symptoms, a few hours after exposure. Symptoms last for about a week [1-2]. In subacute and chronic HP clinical findings are subtle: cough, dyspnea, fatigue, and weight loss [1,2,3,5]. The subacute disease develops in weeks/months, the chronic disease takes months/years [1], with episodic exacerbations [2];

PFTs can reveal a restrictive or mixed ventilatory defect; DLCO is often reduced [2]. The serum can be assayed for precipitating IgG antibodies (precipitins) against potential antigens. A positive test supports the diagnosis of HP, but a negative test does not exclude it [2,3].

Chronic HP may progress to end-stage fibrosis with pulmonary hypertension and chronic respiratory failure [1]. Rarely, patients (even non-smokers) develop widespread centrilobular emphysema instead of fibrosis, especially in chronic Farmer's Lung or Bird fancier's disease, as in our case [2,3,5]. The pathogenesis of emphysema in HP is not known [5].

The chest radiograph is often normal. HRCT demonstrates abnormalities in more than 90% of cases [3].

Typical HRCT findings of HP include bilateral and symmetric ground-glass opacities predominant in the middle and lower parts of the lung, poorly defined centrilobular small nodules (smaller than 5 mm), and air trapping [2-5]. Ground-glass opacities reflect the presence of interstitial lung inflammation, the small centrilobular nodules reflect cellular bronchiolitis and air trapping is due to bronchiolar obstruction [3-5].

Lung consolidations can be a consequence of organizing pneumonia or a superimposed infection [5].

When fibrosis develops, HRCT shows reticulation, traction bronchiectasis, and sometimes honeycombing with a middle-lung distribution (lower lobe predominance is not indicative of chronic HP) [3] and with lobar volume loss [2,3,5].

Rarely in chronic HP, emphysema in the upper lobes can be present [2,3,5].

HP has a broad spectrum of histological manifestations. Since most patients who have lung biopsies are in the chronic phase, little is known about the histological findings of acute HP [5]. Usually, it is characterized by a neutrophilic infiltrate in the alveoli and respiratory bronchioles, and non-specific interstitial pneumonia is commonly seen [5]. Subacute HP shows lymphocytic interstitial pneumonitis, granulomas, organizing pneumonia, and fibrosis [5]. Chronic HP can have a distinctive histological appearance consisting of chronic bronchiolitis, diffuse chronic interstitial inflammatory infiltrates, poorly circumscribed interstitial non-necrotizing granulomas, scattered areas of organizing pneumonia with intraluminal bronchiolar polyps, individual giant cells in the alveoli or interstitium, and in advanced cases of fibrosis [3-5]. Subtle cases require clinical correlation and the diagnosis may remain tentative [3-5].

Conclusion and Learning Point

The diagnosis of HP can be challenging, in particular, when consequent emphysema is present. A possible linkage of emphysema to HP should be considered, especially when there is no association to smoking history or α1-antitrypsin deficit and typical CT findings of HP are present. In uncertain cases, a lung biopsy can provide a definitive diagnosis.

What is new?

The importance of this case report is raising the attention of the clinicians on a rare and atypical presentation of Hypersensitivity Pneumonitis, which is not seen frequently in clinical practice.

List of Abbreviations

DLCO	Diffusing capacity of the lungs for carbon monoxide
HP	Hypersensitivity Pneumonitis)
HRCT	High-resolution computed tomography
PFTs	Pulmonary function tests
RB-ILD	Respiratory bronchiolitis-interstitial lung disease

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Conflict of interests

The authors declare that there is no conflict of interest regarding the publication of this article.

Consent for publication

Written consent was obtained from the patient.

Ethical approval

Ethical approval is not required at our institution to publish an anonymous case report.

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Summary of the case

1	**Patient (gender, age)**	Male, 60Y
2	**Final diagnosis**	Chronic Hypersensitivity Pneumonitis
3	**Symptoms**	Dyspnea, fever, productive cough with purulent sputum and hypoesthenia
4	**Medications**	Glucocorticoids, bronchodilators and levofloxacin
5	**Clinical procedure**	Multidisciplinary group of lung disease - HRCT – Lung Biopsy
6	**Specialty**	Radiology and imaging – Pulmonolgy - Pathology