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Farmer's Lung in a Case after Bullectomy

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Established Facts

- Patients with chronic hypersensitivity pneumonitis, particularly farmer's lung, may develop lung emphysema and not lung fibrosis.

Novel Insights

- A single giant bulla, and not only lung emphysema, can be the primary and single manifestation of chronic hypersensitivity pneumonitis.

Key Words

Farmer's lung · Hypersensitivity pneumonitis · Giant bullae · Lung emphysema

Abstract

We present a case of farmer's lung (FL) with the primary presenting feature of a large bulla in the lung. A 70-year-old nonsmoking woman with dyspnea on exercise was referred for surgical resection of a large bulla in the lung. The postoperative evaluation of the lung tissue revealed a follicular lymphocytic alveolitis and loosely formed granulomas suspicious for hypersensitivity pneumonitis (HP). The patient had worked in farming since her youth. Dyspnea on exercise was the only symptom, but it was related to the large bulla. No other radiologic features of HP were shown in a high-resolution CT of the lung. Specific IgG antibodies against typical antigens of FL were detected, bronchoalveolar lavage dem-

onstrated no lymphocytic alveolitis but an inhalative challenge with own hay was positive. A diagnosis of chronic FL was made. Despite lung emphysema being a possible reaction in FL, giant bullae as primary and single manifestation of this disease have not been reported before.

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Introduction

Hypersensitivity pneumonitis (HP), also known as extrinsic allergic alveolitis, represents an immunologically mediated diffuse interstitial and alveolar lung disease. It is caused by repeated inhalation of various antigens, such as organic dust particles or low-molecular-weight chemicals [1]. Chronic HP can be subgrouped into two kinds – recurrent and insidious. Particularly in insidious chronic HP, patients have no history of acute epi-

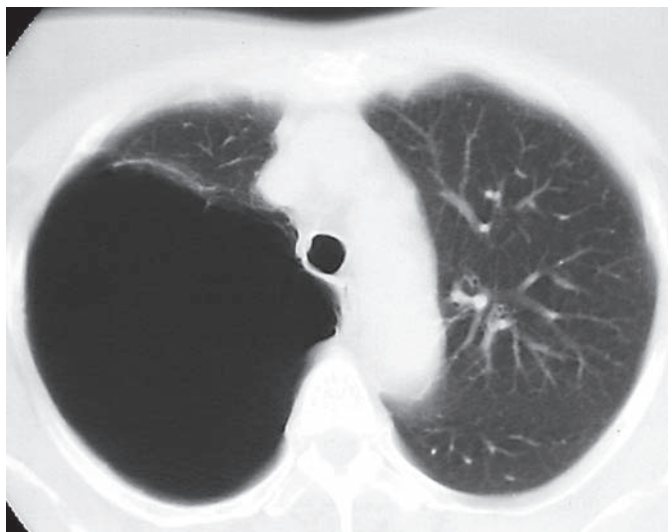
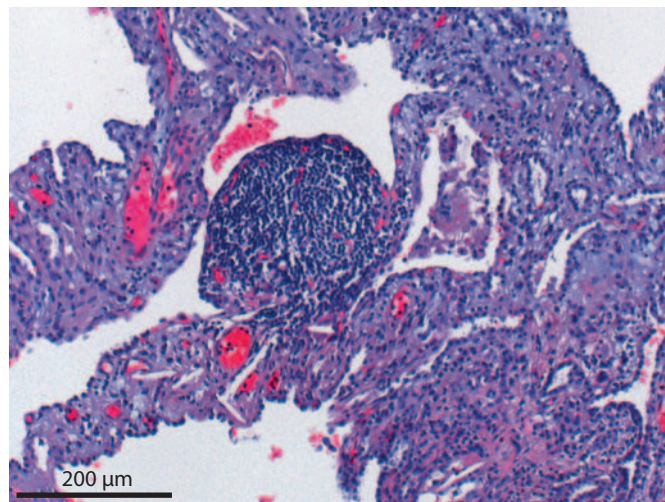


Fig. 1. A CT scan of the lung demonstrated a large bulla in the right upper lobe, occupying nearly half of the right hemithorax and compressing the adjacent normal lung tissue.



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Fig. 2. The histopathological examination of the resected bulla demonstrated interstitial follicular lymphocytic alveolitis in the surrounding lung parenchyma, poorly formed nonnecrotizing granulomas and occasional cholesterol clefts suspicious for HP.

Table 1. Pulmonary function tests before and after bullectomy

Variable	Before bullectomy	After bullectomy
FVC, l	2.6 (102)	2.9 (114)
FEV ₁ , l	1.9 (91)	2.2 (105)
FEV ₁ /FVC, %	74	76
TLC, l	5.6 (112)	5.4 (110)
RV/TLC, %	54.5 (127)	47.1 (110)
DL _{CO} , % predicted	70	82

Values in parentheses are % predicted. RV = Residual volume; DL_{CO} = diffusing capacity of the lung for carbon monoxide.

sodes with typical flu-like symptoms, but have slowly progressive chronic respiratory disease [2].

Chronic HP may progress to pulmonary fibrosis, but particularly in chronic farmer's lung (FL), emphysema seems to be more frequent [3, 4]. The pathogenesis of emphysema in these patients is not definitively known.

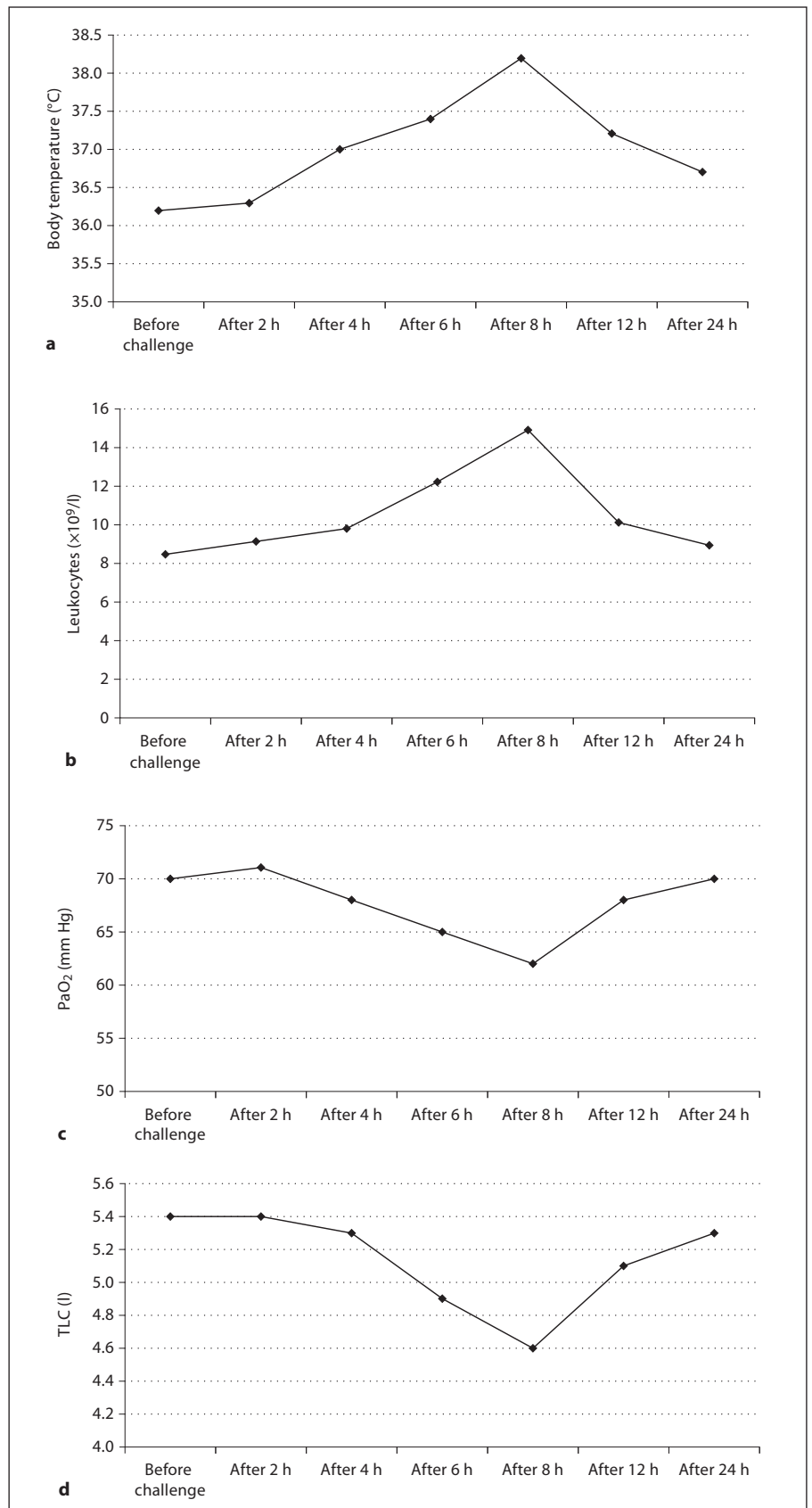
Case Report

A 70-year-old nonsmoking woman was transferred to the Fachkrankenhaus Coswig, Centre for Respiratory Medicine for resection of a large bulla in the lung. She reported increased coughing and dyspnea on exercise in the previous year. Physical

examination, particularly auscultation of the lungs, revealed no abnormalities. Blood gas analysis at rest was normal and showed mild hypoxemia on exercise with a PaO₂ of 65 mm Hg, PaCO₂ of 39 mm Hg and pH of 7.45. Pulmonary function tests (table 1) revealed normal lung volumes [total lung capacity (TLC) 5.6 liters (112% predicted), forced vital capacity (FVC) 2.6 liters (102% predicted), forced expiratory volume in 1 s (FEV₁) 1.9 liters (91% predicted)], FEV₁/FVC of 75% and a diffusion capacity of 5.80 mmol/min/kPa (70% predicted). A CT scan of the lung (fig. 1) revealed a large bulla in the right upper lobe, occupying nearly half of the right hemithorax and compressing the adjacent normal lung tissue. No features of a diffuse emphysema or interstitial lung disease were detected. A bullectomy was performed and the histopathological examination of the resected bulla demonstrated interstitial follicular lymphocytic alveolitis in the surrounding lung parenchyma, poorly formed nonnecrotizing granulomas and occasional cholesterol clefts suspicious for HP (fig. 2).

Further investigation revealed that the patient had worked in farming since her youth but had never reported typical recurrent flu-like symptoms. Ten years ago she had stopped active working but was still living in the farmhouse. Specific IgG antibodies were elevated for *Aspergillus fumigatus*, *Aspergillus niger*, *Penicillium chrysogenum* and *Saccharopolyspora rectivirgula*. A bronchoscopy with BAL was performed and the differential cell count showed 10% lymphocytes with a decreased CD4/CD8 ratio (0.61), 84% (foamy) macrophages, 2% eosinophils and 3% neutrophils. An inhalative challenge with tossing own hay from the former workplace over a 1-hour period was performed. The inhalative challenge was positive with systemic and pulmonary reactions 4–8 h after stopping the exercise (fig. 3a–d). A definitive diagnosis of a chronic residual FL was made. The patient was followed up for more than 2 years without any further clinical, functional or radiological deterioration.

Fig. 3. Inhalative hay challenge over 1 h resulted in fever of 38.2°C (a), a leukocytosis in the peripheral blood (b), hypoxemia with a PaO₂ of 62 mm Hg (c) and restrictive lung function pattern with a decrease of TLC by 14% (d), 4–8 h after stopping the test. The patient complained of dyspnea, coughing, mild shivering and chest tightness.



Discussion

In this report we present the case of a patient with FL and a giant bulla as the primary and single manifestation of the disease.

Patients with chronic HP may develop diffuse lung fibrosis, but emphysema seems to be more frequent in FL. Emphysema has been found significantly more often in FL patients than in control farmers [3]. Radiological evidence of emphysema in high-resolution computed tomography images was detected in 31 out of 95 FL cases, and of these 17 had never smoked, suggesting that emphysema is a direct consequence of HP [4].

In our patient no emphysema was found in CT but a giant bulla of the right upper lobe was present. In the management of bullous emphysema it is important to differentiate between a simple giant bulla and vanishing lobes, which means a lobe destroyed by bullae. Resection of giant bullae is a well-known procedure, but the vanishing type of bullous emphysema is not a good indication for elective operation because the preoperative situation reappears in the remaining emphysematous parenchyma [5]. Most patients with giant bullae are smokers, but they can also occur in nonsmokers [6] as seen in our patient. We can only assume that the giant bulla may be due to an obstructive bronchiolitis, which is a common pathologic feature in acute and chronic courses of FL [7].

The histopathological findings in the surrounding lung parenchyma of the resected bulla in our patient

demonstrated interstitial follicular lymphocytic alveolitis, poorly formed nonnecrotizing granulomas and occasional cholesterol clefts, and were highly suspicious for HP [8]. In the BAL no lymphocytosis was shown, which usually makes a diagnosis of HP uncertain [1], but not in residual disease. This clinical course represents late emphysematous or fibrotic sequelae of the disease in which the typical alveolar lymphocytosis has disappeared [9, 10]. In our patient there is no doubt about the diagnosis of FL, given the patient's history of farming since youth, that she still lives in the farmhouse, the elevated specific IgG antibodies against typical antigens of FL, the highly suspicious pathologic features in the lung parenchyma and the positive inhalative hay challenge. There was no other differential diagnosis for the patient's disease. CT scans showed no signs of other interstitial lung diseases and no medical history for drug-induced pneumonitis existed. In patients with organic dust toxic syndrome, a noninfectious, febrile illness that occurs after heavy organic dust exposure, no granulomatous or lymphocytic alveolitis is detectable in lung biopsies and positive specific IgG antibodies are not typical [11]. Therefore, we would classify the clinical course of our patient as a nonprogressive or residual chronic HP according to the classification of Fink [12].

In conclusion, to the best of our knowledge, this is the first report of a giant bulla associated with chronic HP in a case of chronic farmer's lung, particularly as the primary manifestation of the disease.

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