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# Pneumomediastinum as a primary manifestation of chronic hypersensitivity pneumonitis

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## Summary

**Background:**

Patients with chronic hypersensitivity pneumonitis (HP) can present with an insidious onset of their disease without typical fluctuating flu-like symptoms, and there are only signs of chronic respiratory failure caused by the progressive fibrotic lung disease.

**Case Report:**

A 45-year-old man with a pneumomediastinum and interstitial lung disease was referred for further investigations and therapy. No traumatic event or interventional procedure had occurred prior to referral. The patient had been working in farming for almost 20 years and was exposed in childhood by his father to pigeon breeding from childhood until 20 years ago. He reported dyspnea on exercise for the previous 2 years. High-resolution CT of the lung showed a pneumomediastinum and a fibrotic interstitial lung disease without dominating ground-glass opacities. Specific IgG antibodies were markedly elevated against molds and avian antigens. Bronchoalveolar lavage demonstrated a slightly lymphocytic and neutrophilic alveolitis. After recovering from the pneumomediastinum, an open lung biopsy was performed and a UIP-pattern was detected. An inhalative challenge with hay from the work-place was positive. A diagnosis of chronic farmer's lung was made.

**Conclusions:**

Pneumomediastinum has been described in other fibrotic lung diseases, but until now it has not been described as a primary manifestation of chronic fibrotic HP. Particularly in cases of concurrent antigen sources, an inhalative challenge could be done, even in a chronic course of HP.

**key words:**

farmer's lung • hypersensitivity pneumonitis • pneumomediastinum

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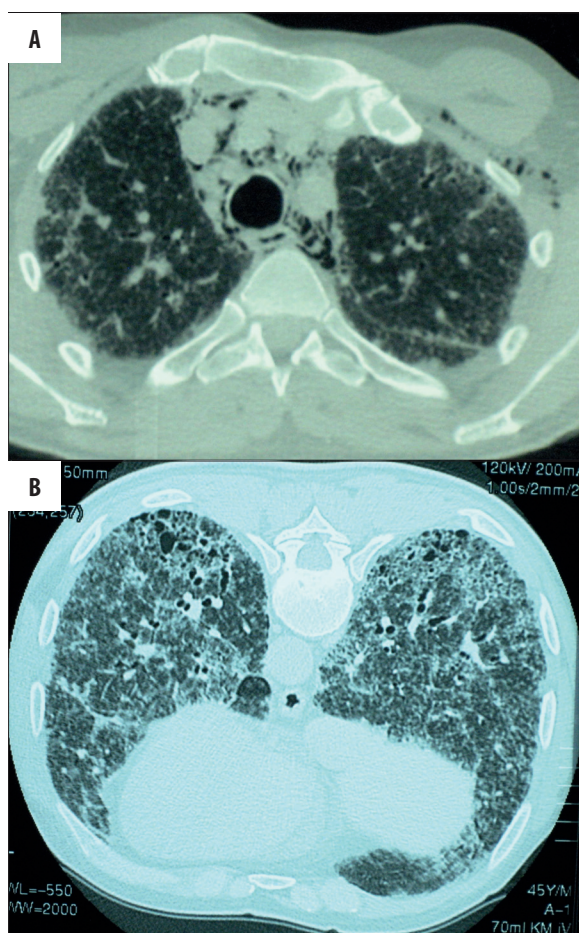
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## BACKGROUND

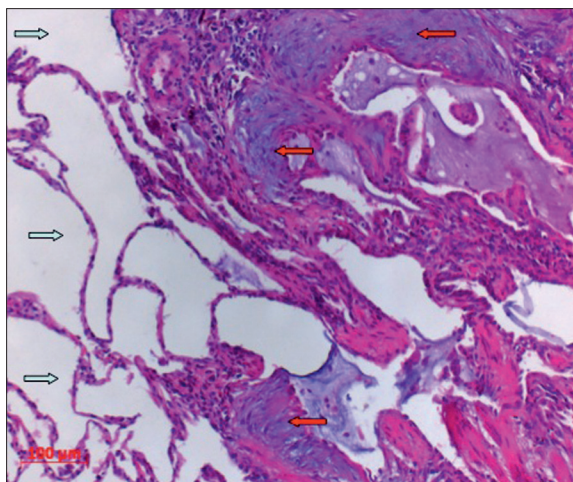
Chronic hypersensitivity pneumonitis (HP) can be grouped into 2 types – recurrent and insidious chronic HP. Particularly in insidious chronic HP, patients have no history of acute episodes with typical flu-like symptoms, but instead have slowly progressive chronic respiratory disease [1].

Pneumomediastinum, also called mediastinal emphysema, is the presence of free air around mediastinal structures and can be divided in spontaneous pneumomediastinum, without any obvious primary source, and secondary pneumomediastinum, with a specific responsible pathologic event such as trauma, intrathoracic infections or trauma to the aerodigestive tract [2].

Until now, pneumomediastinum has only been described in a patient with acute HP [3] and in a patient with HP during steroid treatment [4]. To the best of the authors' knowledge, this is the first report of pneumomediastinum as the primary manifestation of HP in a patient with a well-documented chronic insidious course of farmer's lung.



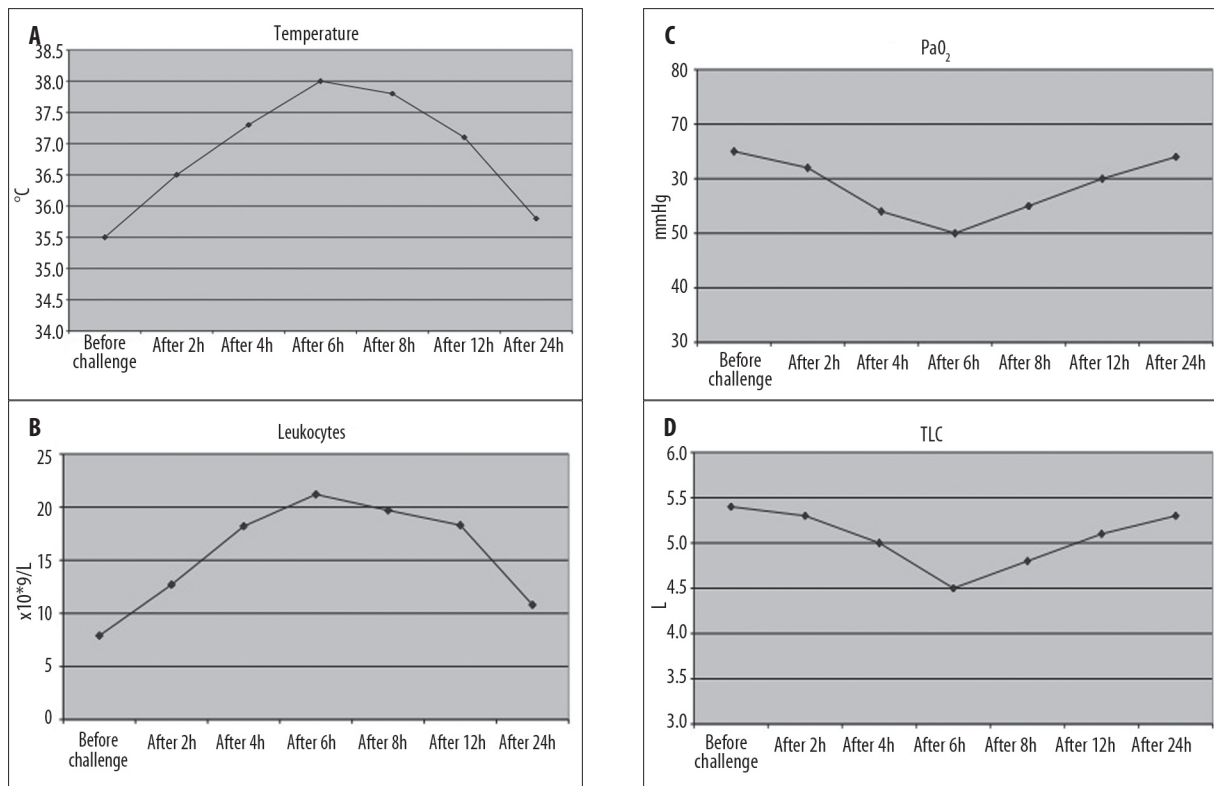
**Figure 1.** High-resolution computed tomography (CT) of the chest demonstrated a) a marked pneumomediastinum and b) an interstitial lung disease with reticulation in all lobes, but basal predominance, honeycombing, traction bronchiectasis, and mild to moderate ground-glass opacities.



**Figure 2.** Surgical lung biopsies showed lung fibrosis in a patchy pattern with architectural distortion and fibroblast foci resembling usual interstitial pneumonia (UIP). Green arrows: normal lung parenchyma. Red arrows: fibroblast foci.

## CASE REPORT

A 45-year-old nonsmoking male was transferred to the Fachkrankenhaus Coswig Centre for Respiratory Medicine for further investigation and management of pneumomediastinum and interstitial lung disease. He had worked in farming for almost 20 years and was exposed in childhood by his father to pigeon breeding until 20 years ago. He reported dyspnea on exercise and weight loss of 10 kg in the previous 2 years. Two days prior to admission at our center, he had complained of chest pain. No traumatic event or interventional procedure had occurred previously. On physical examination, the patient presented with tachypnea and was afebrile. Auscultation of the lungs revealed inspiratory crackles in both lung bases. Blood gas analysis showed mild respiratory failure at rest, with a PaO<sub>2</sub> of 68 mmHg, PaCO<sub>2</sub> of 40 mmHg and pH of 7.44. Laboratory tests showed no abnormalities. Specific IgG antibodies were highly elevated for different molds, particularly *Aspergillus fumigatus* and moderately elevated for different avian antigens, particularly pigeon antigens and goose feathers. A high-resolution CT scan of the lung (Figure 1A,B) demonstrated a marked pneumomediastinum and interstitial lung disease with reticulation in all lobes, but basal predominance, honeycombing, traction bronchiectasis, and mild-to-moderate ground-glass opacities. Pulmonary function tests revealed moderate restriction (TLC 4.5 L [59% predicted], FVC 3.0 L [56% predicted], FEV<sub>1</sub> 2.9 L [72% predicted]), FEV<sub>1</sub>/FVC 98%, and a diffusion capacity of 2.20 mmol/min/kPa (19% predicted). Bronchoscopy with BAL and transbronchial biopsies were performed. The BAL differential cell count showed 19% lymphocytes, 72% macrophages and 9% neutrophils. Flow cytometric analysis of the BAL lymphocyte population revealed a CD4/CD8 ratio of 0.5. Because the transbronchial biopsy specimens revealed no conclusive pathological pattern, an open lung biopsy was performed and demonstrated a UIP-pattern without any granulomas (Figure 2). After complete resolution of the pneumomediastinum and slight improvement of the lung function with temporary prednisolone therapy for 6 weeks, an inhalative



**Figure 3.** Inhalative hay challenge over one hour resulted in fever of 38°C, a leucocytosis in the peripheral blood, hypoxemia with a PaO<sub>2</sub> of 50 mmHg and restrictive lung function pattern with a decrease of TLC by 15%, 4–6 hours after stopping the test. The patient complained of dyspnea, mild shivering and chest tightness.

challenge with tossing hay from the workplace over a 1-hour period was performed in a special provocation chamber of the hospital. The inhalative challenge was positive, with systemic and pulmonary reaction 4–6 hours after stopping the exposure (Figure 3A–D). A definitive diagnosis of a chronic farmer's lung was made.

## DISCUSSION

In this report we present a well-documented case of a patient with chronic farmer's lung and a pneumomediastinum as the primary manifestation of the disease.

Pneumomediastinum results from rupture of the alveoli due to marked increase in intra-alveolar pressure. In patients with interstitial lung diseases, subpleural or paracardiac blebs may form due to the distortion of lung architecture, and the rupture of paracardiac blebs may also lead to air leakage in the mediastinum [5]. In a retrospective analysis of 74 patients with pneumomediastinum, 28 had a spontaneous form and 2 (7%) had an idiopathic pulmonary fibrosis (IPF) as a predisposing condition [2]. The frequency of pneumomediastinum in patients with IPF was investigated in a study by Franquet et al, who found 4 (5%) pneumomediastinums in 78 patients with confirmed IPF who underwent a computed tomography of the chest [6]. Another study found a pneumomediastinum in 5 of 34 patients with pulmonary fibrosis (15%) on computed tomography of the chest. Honeycombing and violent cough were considered to be predisposing factors of this complication [7].

There is 1 case report of a pneumomediastinum in a patient with acute HP. An open lung biopsy revealed a granulomatous inflammation of the lung, resulting in obstruction or narrowing of respiratory bronchioles and emphysematous surrounding alveoli [3]. It was postulated that this obstructive bronchiolitis, which is a common pathologic feature in acute and chronic courses of farmer's lung, assumed an important role in the development of the pneumomediastinum. Another case report presented a patient with non-fibrotic chronic farmer's lung complicated by bilateral pneumothorax and mediastinal emphysema under treatment with steroids. The authors postulated that the steroid administration may have caused these complications [4].

In chronic insidious bird fancier's lung, a hypersensitivity pneumonitis caused by avian antigens, honeycombing was observed on HRCT scans in most cases. Only a mild lymphocytosis was observed in BAL fluid, and UIP-like lesions could be revealed from surgical lung biopsies [1,8]. Particularly, an inhalative challenge could help to differentiate chronic bird fancier's lung from ILF [9,10]. In our patient there is therefore no doubt about the diagnosis of chronic farmer's lung, because of the history of farming over nearly 20 years with highly elevated specific IgG-antibodies against molds, particularly *Aspergillus fumigatus*, and the positive inhalative hay challenge.

## CONCLUSIONS

In conclusion, pneumomediastinum is a rare complication of interstitial lung diseases. To the best of the authors'

knowledge, this is the first report of a pneumomediastinum associated with chronic fibrotic HP in a well-documented case of farmer's lung, and particularly as the primary manifestation of the disease.

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