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(RESEARCH ARTICLE)

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Gougerot-Sjögren Syndrome Associated with Hypersensitivity Pneumonitis: Report of Two Cases"

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Abstract

Chronic hypersensitivity pneumonitis (HP) is a rare interstitial lung disease caused by environmental antigen exposure. This report describes two cases of chronic HP associated with Sjögren's syndrome. The first case involved a 55-year-old woman exposed to pigeon droppings, presenting with dyspnea and alveolitis confirmed by bronchoalveolar lavage and positive anti-pigeon antibodies. The second case was a 41-year-old woman exposed to mold, presenting with basal fibrosis and positive anti-SSA/SSB antibodies. Both cases demonstrated salivary gland lymphocytic sialadenitis consistent with Sjögren's syndrome. Treatment included antigen avoidance, corticosteroids, and azathioprine. These cases underscore the importance of thorough diagnostic evaluation and a multidisciplinary approach for optimal management of HP and associated autoimmune diseases.

Keywords: Hypersensitivity pneumonitis; Sjögren's syndrome; Autoimmune diseases; Antigen exposure; Corticosteroids; Immunosuppressive therapy

1. Introduction

Hypersensitivity pneumonitis (HP) is an immune-mediated interstitial lung disease caused by repeated exposure to specific environmental antigens. Among its forms, "domestic lung," associated with indoor mold exposure, and "bird fancier's lung," linked to avian protein inhalation, are well-documented. These conditions can lead to chronic pulmonary inflammation and, if untreated, progress to irreversible fibrosis [1].

Sjögren's syndrome, a systemic autoimmune disorder primarily affecting exocrine glands, is known to cause a variety of pulmonary manifestations, including interstitial lung disease. While its association with HP is rare, it presents unique diagnostic and therapeutic challenges due to overlapping clinical and pathological features [2].

Here, we report two cases of HP associated with Sjögren's syndrome, each involving a distinct etiology: one due to mold exposure "domestic lung" and the other related to avian antigen exposure "bird fancier's lung". These cases underscore the importance of considering environmental triggers in patients with autoimmune diseases and highlight the complexities involved in diagnosing and managing such overlapping pathologies.

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2. Case presentation

2.1. Case 1

A 55-year-old female, with a history of exposure to pigeon droppings, presented with a 7-month history of progressive dyspnea and intermittent dry cough. She reported xerophthalmia as an extrathoracic symptom. The pleuro-pulmonary examination was normal.

Chest CT revealed a mosaic attenuation pattern in both lungs, multifocal ground-glass opacities predominantly in the subpleural regions, and associated septal and non-septal thickening (Figure 1).

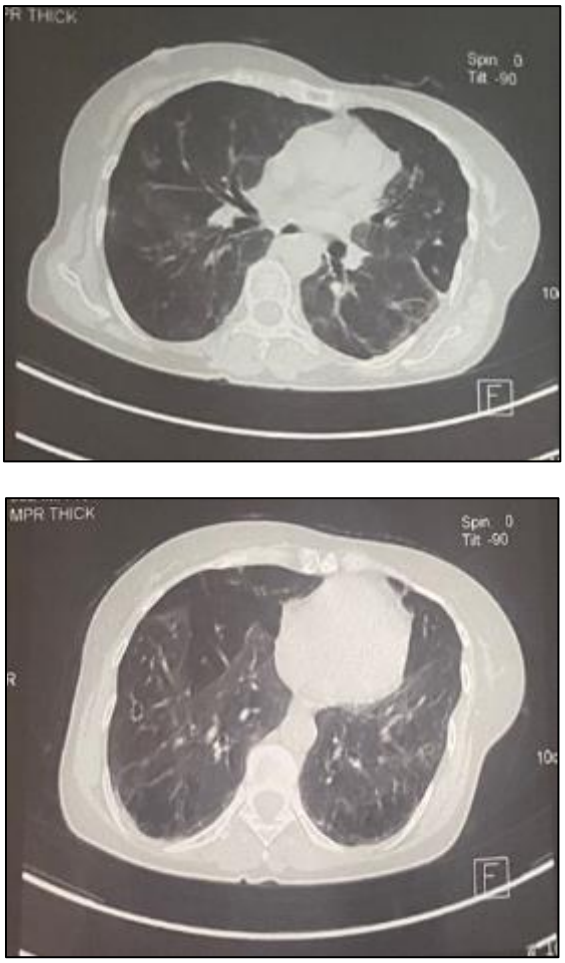

Figure 1 Chest CT scan in the parenchymal window showing a mosaic aspect

Bronchoalveolar lavage showed neutrophilic alveolitis (55%) and eosinophilia (13%). Lung biopsy revealed non-specific chronic inflammatory lesions.

Avian precipitins testing was positive, with anti-pigeon antibodies measured at 109 mg/L, while the immunological workup was negative. Additionally, a biopsy of the salivary glands demonstrated grade 3 sialadenitis, and the Schirmer test was positive, confirming associated xerophthalmia. Based on the clinical, radiological, and histological findings, a diagnosis of bird fancier's lung (hypersensitivity pneumonitis) associated with Sjögren's syndrome was established.

The patient was advised to avoid exposure to pigeons and was started on corticosteroids and immunosuppressive therapy with azathioprine to manage the associated autoimmune component.

2.2. Case 2

A 41-year-old female, with a history of domestic mold exposure and no prior treatment for pulmonary tuberculosis or recent contact, presented with a 7-year history of dyspnea and productive cough with whitish sputum. She also reported xerostomia as an extrathoracic symptom. The condition evolved in an afebrile state with no systemic signs of significant weight loss or asthenia.The pleuro-pulmonary examination revealed bilateral basithoracic dry crackles. Chest CT showed bilateral basal ground-glass opacities with advanced fibrosis, more pronounced in the basal regions, suggestive of chronic hypersensitivity pneumonitis (CHP)

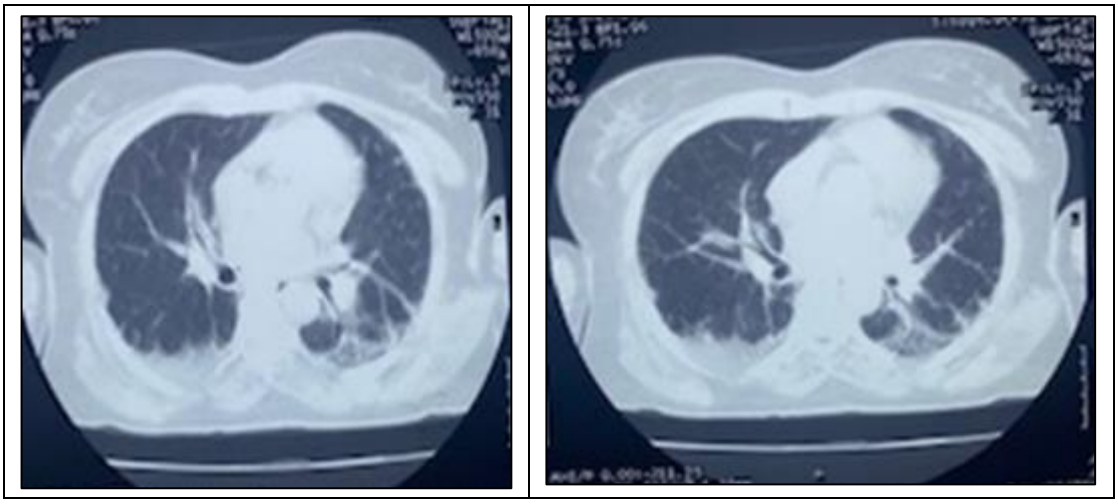

Figure 2 Chest CT showed bilateral basal ground-glass opacities

Bronchoalveolar lavage (BAL) revealed an inflammatory fluid rich in epithelioid histiocytes. Domestic precipitins testing, were positive for Dermatophagoides pteronyssinus indicating significant antigen exposure.

Biopsy of the salivary glands demonstrated lymphocytic sialadenitis, stage 4, consistent with Sjögren's syndrome. Immunological workup confirmed the presence of anti-SSA and anti-SSB antibodies.

Both patients were advised to avoid further exposure to their respective environmental antigens (pigeon droppings and domestic mold) to prevent ongoing antigenic stimulation. Treatment was initiated with corticosteroids, along with immunosuppressive therapy using azathioprine to manage the associated autoimmune component.

This combined approach aimed to control pulmonary inflammation, prevent disease progression, and manage the systemic manifestations of Sjögren's syndrome. Regular follow-up was scheduled to monitor therapeutic response, assess lung function, and detect potential side effects of the treatment.

3. Discussion

Hypersensitivity pneumonitis (HP) is an interstitial lung disease caused by repeated exposure to inhaled organic antigens, leading to immune-mediated inflammation. Chronic HP, as observed in both cases, can progress to pulmonary fibrosis, a condition associated with significant morbidity and mortality [3].

In the first case, exposure to pigeon antigens led to bird fancier's lung, a well-documented form of HP. The second case involved exposure to domestic molds, a less common but recognized cause of HP. Both cases highlight the importance of detailed environmental exposure histories in diagnosing HP, as the clinical presentation can overlap with other interstitial lung diseases [4,5].

Radiological findings in both cases were consistent with fibrotic hypersensitivity pneumonitis, characterized by features such as mosaic attenuation with air trapping, ground-glass opacities, and basal-predominant fibrosis. According to the most recent ATS/JRS/ALAT guidelines, high-resolution CT plays a central role in diagnosis by identifying a combination of fibrosis and small-airway disease patterns suggestive of antigen-induced lung injury [6].

The association of HP with Sjögren's syndrome in both cases underscores the potential overlap between autoimmune diseases and interstitial lung diseases. Sjögren's syndrome can independently cause lymphocytic interstitial pneumonia,

but the coexistence of chronic HP suggests a multifactorial pathophysiology. The detection of anti-SSA and anti-SSB antibodies in both patients strengthens the diagnosis of Sjögren's syndrome and highlights the importance of a thorough immunological evaluation [7,8].

Treatment for chronic HP focuses on antigen avoidance and immunosuppression. Both patients were started on corticosteroids to reduce inflammation and azathioprine to manage autoimmune components and prevent further fibrosis. This approach is consistent with current management guidelines for HP and connective tissue disease-associated interstitial lung diseases [9,10].

The prognosis of chronic HP depends on the extent of fibrosis and ongoing antigen exposure. Early recognition and intervention are crucial to improve outcomes. These cases emphasize the importance of a multidisciplinary approach, involving pulmonologists, immunologists, and radiologists, for optimal patient care [11].

4. Conclusions

These cases highlight the diagnostic complexity of chronic hypersensitivity pneumonitis (HP) and its association with autoimmune conditions like Sjögren's syndrome. Accurate diagnosis relies on detailed environmental history, imaging, and immunological evaluation. Early intervention with antigen avoidance, corticosteroids, and immunosuppressive therapy is crucial to prevent disease progression and fibrosis. This underscores the need for a multidisciplinary approach to optimize outcomes in such complex cases.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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