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Case Report

Desquamative Interstitial Pneumonia in a 63-Year-Old Non-Smoker

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Abstract

Desquamative interstitial pneumonia is an interstitial lung disease most associated with smoking [1]. It was first described by Liebow in 1965 and the name originated from the belief that the dominant histological feature was the desquamation of epithelial cells. We report the case of a 63-year-old man who presented at the medical outpatient department of UNTH with two years history of recurrent cough and shortness of breath on exertion and a transbronchial biopsy histological finding of foci showing intra-alveolar accumulation of macrophages with interstitial fibrosis and mild lymphocytic infiltrates. This patient had no history of smoking nor exposure to occupational dust but was involved in old currency handling for over 25 years as a bank worker. Thus, DIP is not always associated with exposure to tobacco smoke, occupational exposure to other agents should also be considered to avoid disregarding an occupational aetiology.

Keywords: Desquamative Interstitial Pneumonia, Non-Smoker, Trans-Bronchial Biopsy, Interstitial lung Disease

Background

Desquamative interstitial pneumonia is an interstitial lung disease most associated with smoking [1]. It is a rare form of idiopathic interstitial pneumonia characterized pathologically by extensive alveolar infiltration with macrophages followed by interstitial inflammation and fibrosis. It was first described by Liebow in 1965 and the name originated from the belief that the dominant histological feature was the desquamation of epithelial cells. Even though a vast majority of patients are heavy smokers (90%) with an average smoking history of 18 pack-years, other predisposing factors will include autoimmune/systemic disorders eg rheumatoid arthritis, systemic sclerosis, infections eg HIV, Occupational exposures to asbestos, beryllium. It is also common among farmers, painters, mine workers and rock blasters. The prevalence of DIP is not exactly known, there is paucity of epidemiologic data available for rare forms of IIP such as DIP. We hereby report the case of a 63-year-old man who presented at the medical out-patient department of UNTH with a two-year history of recurrent cough and shortness of breath on exertion and a transbronchial biopsy histological finding of foci showing intra-alveolar accumulation of macrophages with interstitial fibrosis and mild lymphocytic infiltrates. This patient had no history of smoking nor exposure to occupational dust but was involved in old currency handling for over 25 years as a bank worker.

Case Presentation

Our patient is a 63-year-old man who presented with two years history of recurrent cough, and shortness of breath on exertion. Cough was of insidious onset, mostly dry but occasionally

productive of whitish sputum. There was associated non-pleuritic, non-radiating central chest pain and unintentional weight loss. Shortness of breath was first noticed with moderate exertion and progressed to be present on mild exertion. He had no history of smoking nor exposure to occupational dust though he reared poultry for a period of 2 years about 23 years prior to presentation, he was also involved in old currency handling for over 23 years as a bank worker. He was not known to have hypertension, diabetes or asthma.

Physical examination revealed middle aged man who was tachypnoeic with a low oxygen saturation (Spo2) of 80% and bilateral inspiratory crepitations in all lung zones which was worse on the left. He had first and second heart sounds with a loud P [2]. Spirometry result showed a mild restrictive ventilatory pattern Chest radiography showed diffuse reticulonodular opacities with background cystic changes involving all visualized lung fields. Chest computed tomography scan showed widespread and bilateral ground glass opacities in both lung fields with slight upper lobe preponderance. There were associated ground glass nodules, profuse bronchial wall thickening and reticulation with resultant honeycombing effect, small traction and emphysematous bronchiectatic changes are seen bilaterally with left sided preponderance. Bronchoalveolar lavage showed moderate neutrophilic cellular pattern. Trans bronchial biopsy histology result showed pulmonary parenchyma with preserved architecture, foci showing intra-alveolar accumulation of macrophages with interstitial fibrosis and mild lymphocytic infiltrates. Blood and urine work up were essentially normal. A diagnosis of Desquamative interstitial pneumonia was made. He was treated with Tablet Prednisolone 40mg mane, Tablet Azithromycin 500mg daily, Syrup Broncholyte 10 mls tds, Supplemental intranasal oxygen at 3L/min over 2 hours stat, then PRN to aim at oxygen saturation above 94% on room air. Patient made a good clinical recovery and was subsequently discharged and is being followed up in our out-patient clinic.

Discussion

Desquamative interstitial pneumonia is a rare form of idiopathic interstitial pneumonia which comprises a group of diffuse lung diseases of unknown aetiology, that primarily involve the pulmonary interstitium as well as the septal and broncho vascular tissues which make up the fibrous framework of the lung [3]. These primarily interstitial processes frequently also involve the airways, the vasculature and alveolar airspaces. It is characterized pathologically by extensive alveolar infiltration with macrophages followed by interstitial inflammation and fibrosis. Risk factors for the disease include smoking especially heavy smokers (90%) with an average smoking history of 18 pack-years, other predisposing factors will include autoimmune/systemic disorders eg. rheumatoid arthritis, systemic sclerosis, infections eg HIV, and occupational exposures to asbestos, and beryllium. Farmers, painters, mine workers and rock blasters are also affected [2]. Thus, DIP is not always associated with exposure to tobacco smoke; occupational exposure to other agents should also be considered to avoid disregarding an occupational aetiology.

Desquamative interstitial pneumonia Together with respiratory bronchiolitis-associated interstitial lung disease and primary Langerhans cell histiocytosis are collectively grouped as smoking related ILDs [4]. Studies have shown that it typically affects cigarette smokers in their 4^{th} and 5^{th} decades of life; [5,6] a small number of cases are noted to be associated with autoimmune and systemic disorders like rheumatoid arthritis, use of drugs like sirolimus, nitrofurantoin, and sulfasalazine, infections like hepatitis C, and cytomegalovirus and surfactant dysfunction in children [2]. It generally presents in the 5^{th} decade of life and there is a slight male preponderance. Most patients complain of exertional dyspnea and dry cough, crackles are the most frequent remarkable findings at physical examination [7]. Pulmonary function shows a restrictive pattern with decreased DLCO. A diagnosis of DIP cannot be reliably established based on clinical and radiological features alone and ideally requires a lung biopsy. Histology shows diffuse accumulation of pigmented macrophages in alveolar airspaces [4]. Smoking cessation is considered an essential component of management in smokers. Corticosteroids are often used with high response rates [8,9] A few patients have been treated with additional immune suppressive agents [10,11]. Including Azathioprine and Cyclophosphamide although response to these agents is not described [5]. Prognosis is good, improvement in ground glass opacity on HRCT may correlate with response to treatment [12,13]. The overall survival is 70-88% after 10 years. Fluctuating course with remissions and relapses may occur [8,14,15] (Figure 1-4).

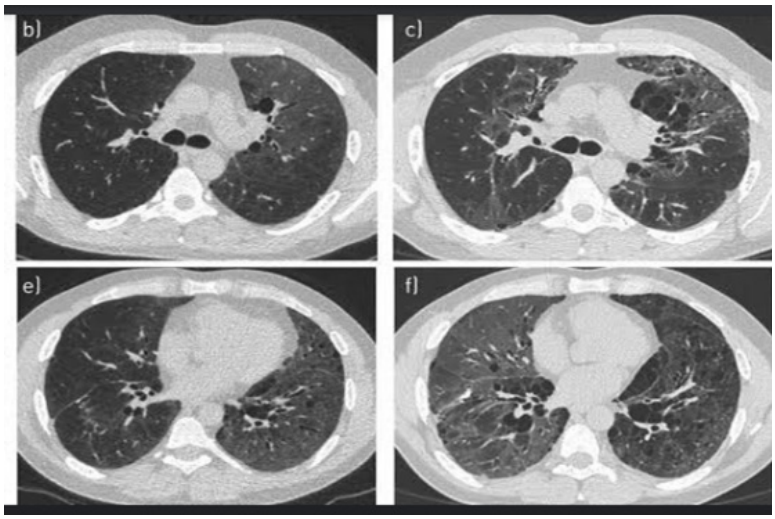

Figure 1

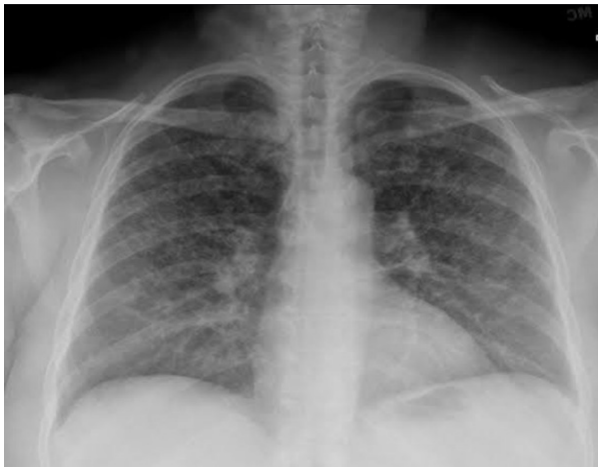

Figure 2: Chest radiograph showing reticulonodular opacities with background cystic changes.

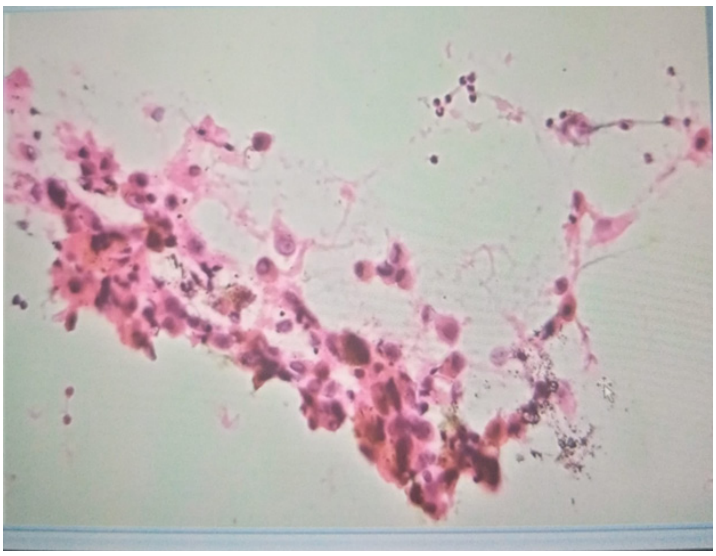

Figure 3: Histology picture of BAL fluid showing neutrophilic cellular pattern.

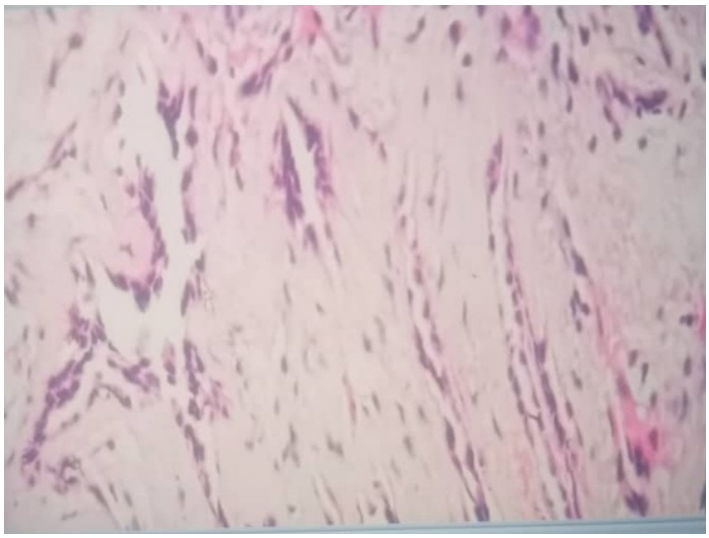

Figure 4: Histology picture showing intra-alveolar accumulation of macrophages.

Acknowledgement

None.

Conflict of Interest

None.

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