Concurrent Onset of Pulmonary and Articular Symptoms - Rare Manifestation of Rheumatoid Arthritis Associated Interstitial Lung Disease: A Case Report

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Abstract

Lung is the most common extra-articular site of involvement in rheumatoid arthritis (RA). It can involve pleura, lung parenchyma and pulmonary vasculature. Among many diseases, most debilitating one is RA associated interstitial lung disease (RA-ILD). Here, we describe a middle aged Bangladeshi lady who presented because of progressively deteriorating non-productive cough, exertional dyspnea and polyarthritis of 5 month duration. Diagnostic workup confirmed RA-ILD and she responded well with prednisolone, hydroxychloroquine and azathioprine.

Key words: articular, concurrent, interstitial lung disease, pulmonary, rheumatoid arthritis.

Introduction

Rheumatoid lung is a common condition, occurring in upto 54% of rheumatoid arthritis (RA) cases. Pulmonary diseases in RA is the second leading cause of death. Among the various patterns of pulmonary diseases, interstitial lung disease (ILD) represents the most severe one, accounting for 20-30% cases. Risk factors for RA associated ILD (RA-ILD) includes male sex, smoking, high titres of rheumatoid factor (RF) and anti-CCP antibody, environmental exposures, methotrexate and other disease modifying anti-rheumatic drugs etc. Usually RA-ILD is suspected when long standing RA patients present with non-productive cough and exertional dyspnea. Rarely patients may present with cor pulmonale. In some cases, RA manifest later in established ILD cases. But simultaneous onset of pulmonary and articular symptoms are rare. Here, we present such a case.

Case Report

A 65-year-old diabetic lady presented with 5 month history of progressively deteriorating non-productive cough, exertional dyspnea and polyarthritis involving all small and large joints including hand joints with significant morning stiffness. She did not have any chest pain, palpitation or rash. She lost 3 kg in weight during this period.

The lady was moderately anaemic. There was no rash or lymphadenopathy. She had bilateral crepitation upto the midchest. Her metacarpophalangeal and proximal interphalangeal joints were swollen and tender and there was swan neck deformity in 3rd and 4th fingers bilaterally. Her limb movements were restricted because of pain. Her haemoglobin was 10.4 gm/dl, normochromic normocytic, total and differential white cell count was normal, platelet count was 4,79,000/cmm, ESR- 100 mm in 1st hour, CRP- 24 mg/dl. Her fasting blood glucose was 11.8 mmol/L, HbA1c- 8.9%.

Chest x-ray PA view revealed increased reticular shadowing in both bases and periphery suggesting
interstitial lung disease (Figure 1). X-ray hand was normal. Arterial blood gas analysis showed mild alkalosis. Pulmonary function test revealed mild restrictive disease.

So, the patient was finally diagnosed as a case of RA-ILD and diabetes mellitus (DM). Treatment was started with ibuprofen, prednisolone, hydroxychloroquine and azathioprine. For DM she was receiving premixed insulin and metformin.

Discussion

Pulmonary fibrosis and pleural effusion are two most common pulmonary manifestation of RA. Pathologically, five different groups of disorders are identified; rheumatoid pulmonary nodule, usual interstitial pneumonia (UIP), non-specific interstitial pneumonia (NSIP), bronchiolitis obliterance with organizing pneumonia (BOOP), lymphoid hyperplasia and cellular interstitial infiltrates. Usually pulmonary fibrosis results from a slowly progressive interstitial disease (UIP/NSIP). Open lung biopsy, which carries a significant morbidity, can give a definitive diagnosis, but the advent of HRCT scan has largely replaced it now a days. In our case, ILD was diagnosed by HRCT of chest and lung biopsy was not done.

Typically RA-ILD develops in male patients in 4th to 5th decades. Smoker with high RF and anti-CCP antibody titres are more vulnerable, implies that there might be some pulmonary reaction to autoantibodies produced in RA. Conversely, in cases of ILD without RA, presence of autoantibodies specific for RA may imply that environmental factors exposed to lung may initiate the immunological reaction that may ultimately target synovial joints to produce RA. It is obvious that, concurrent onset of pulmonary and articular symptoms mandate careful exclusion of sarcoidosis, vasculitis, adverse drug reactions and other differentials. Our patient was a female in her 50s, non-smoker with high titres of RF and anti-CCP antibody and we also excluded the differentials.

Lung function tests in ILD typically show restrictive pattern with reduced carbon-monoxide transfer factor. In our case, spirometry revealed moderate restrictive disease.

Treatment includes proper control of RA and ILD with corticosteroids and immune suppressive agents. The initial response in our case with prednisolone, hydroxychloroquine and azathioprine looked good. Overall prognosis is disappointing, median survival after diagnosis is 2.6 years. However, aggressive immune suppression with rituximab, heart lung transplantation
and other experimental agents may improve the overall outcome in future.6

Conclusion
Though uncommon, it is not impossible to present simultaneous onset of respiratory and joint symptoms in RA-ILD. High index of suspicion and careful exclusion of other differentials are necessary.

References