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Medicine

Early Onset Interstitial Lung Disease with Cor Pulmonale in Rheumatoid Arthritis

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ABSTRACT

Interstitial lung Disease in Rheumatoid Arthritis is an increasing common entity and is becoming a common cause of RA related morbidity and mortality. Usually ILD is seen in patients of longstanding Rheumatoid Arthritis, with a gap of several years between the first symptoms of arthritis and ILD. Here we present a case of ILD in Rheumatoid Arthritis where the symptoms of ILD occur prior to the onset of symptoms of arthritis. This patient presented with complains of dyspnoea and dry cough prior to pain in multiple joints, after she came in our observation and after due workup she was found to be a patient of Rheumatoid Arthritis with concurrent ILD

KEYWORDS

Introduction:

Rheumatoid Arthritis is one of the most common auto immune diseases inflicting the world population and with an increasing prevalence in India as well. One of the extra articular features of this disease is pulmonary involvement in the form of Interstitial Lung Disease. ILD is amongst not so uncommon manifestation of RA and may present as either Idiopathic Pulmonary Fibrosis or Non Specific Interstial Pneumonia among the more common patterns of involvement. Although ILD associated with RA usually confers a poor prognosis the Non Specific Interstitial Pneumonia type of presentation has a relatively better prognosis. ILD usually sets in years after the onset of articular symptoms. The course is usually progressive and ends up in mortality.

Case Summary:

This female aged 60 years came to our medical emergency. She had presented with complains of increasing shortness of breath for the last few days which had severely hampered her day to day activity and she was unable to carry out her daily activities. On detailed history taking she revealed that her complains had started around one year back. She started complaining of cough that was non-productive with no diurnal or postural variation. After sometime she started complaining of shortness of breath that was exertional in nature, relieved on rest .Her complains of shortness of breath gradually increased in intensity such that for the last few days prior to admission in our hospital she experienced shortness of breath even on rest. That was when her attendants decided to bring her to our hospital. During the course of illness she had sought advice from local doctors as well, some of whom had given her short courses of steroids and antibiotics, however not satisfied by treatment she had discontinued the drugs.

Apart from respiratory symptoms, for the past 4 months she had complains of pain in multiple joints that included bilateral proximal inter-phalngeal joints, metacarpo-phalangeal joints, wrist, elbow, shoulders, knee, ankle and metatarsals joints. There was presence of morning stiffness. Joint pain restricted her day to day activity and caused her significant distress. She used to take NSAID's from the local pharmacy and got partial relief but the symptoms persisted. Along with her complains of pain in multiple joint. She had no previous history of Tuberculosis, Asthma or COPD or Coronary artery disease. Prior to these last 12 months she never had any complains of chronic cough or shortness of breath. There was no family history of Asthma or COPD and Ischemic Heart Disease. There was no history of any kind of smoking or any hazardous occupational exposure.

At presentation in emergency the patient was severely dyspnoeic and cyanosed (SpO2 71 %). She was first put on inhalation of Moist Oxygen and then a detailed examination was carried out once the patient was feeling better. General examination of the patient revealed cyanosis that was of central type and tenderness in all the joints pointed out in her history. Tenderness was marked in Wrist, MCP and PIP joints. There were no signs of heart failure. Rest of the general examination was unremarkable. Examination of her chest revealed tachypnea, vesicular breathing all over the chest, there were fine inspiratory crackles in the basal regions bilaterally with occasional end inspiratory rhonchi as well. Examination of Cardiovascular system revealed a loud pulmonary component of second heart sound, rest findings were non remarkable. Further systemic examination revealed nothing significant.

Immediately an ECG, ABG and Chest X-ray were ordered. Her ABG analysis showed type 1 respiratory failure. The ECG showed sinus tachycardia with P-pulmonale, no signs of any acute coronary syndrome or previous ischemic insults were seen. The Chest x-ray revealed a bibasilar reticular pattern. Next a Spirometry was done which showed a restrictive pattern with decreased TLC and FRC and minimal decrease in FEV1. Considering the possibility of an interstitial lung disease a High Resolution CT scan was ordered which revealed bilateral multiple areas of ground glass opacities suggestive of ILD – Usual Interstitial Pattern. Next day an Echocardiography was ordered which was suggestive of Cor- Pulmonale. Thus the diagnosis of ILD was established.

X-rays of wrist elbow shoulders and knee revealed no joint erosions. USG of major joints showed no collections. Based on the Revised ACR classification score, the diagnosis of Rheumatoid Arthritis was arrived at

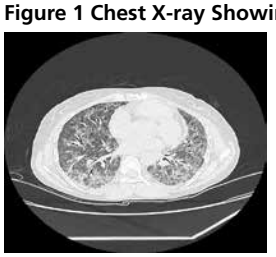
Joint Involvement	>10 joints including a small joint	5
Serology	Rheumatoid Factor > 3 times upper limit Anti CCP positive	3
Acute Phase reactants	Abnormal CRP and ESR	1
Duration	More than 6 months	1

The score was thus 10/10 and the likelihood of this being a case of Rheumatoid Arthritis was very high. However at this stage no joint erosions had developed with the disease having manifested for last 12 months.

His investigation revealed ESR 98mm, CRP:4.8mg/dl, TLC 10700, DLC N71 L25 E2,platelet 4.86 lac/ul, RBS 106 mg/dl, Blood urea 31mg% and serum creatinine 0.6mg%.RA factor 84IU/ML, anti ccp 68 IU/ML.Sputum for AFB was negative. Usg B/L wrist shows No serous joint effusions seen.spirometry shows restrictive pattern

Imaging Studies:

Figure 1 Chest X-ray Showing Basal Reticular Pattern



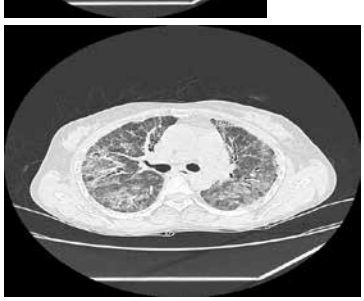


Figure 2 & 3: HRCT showing patchy areas of ground glass opacities.

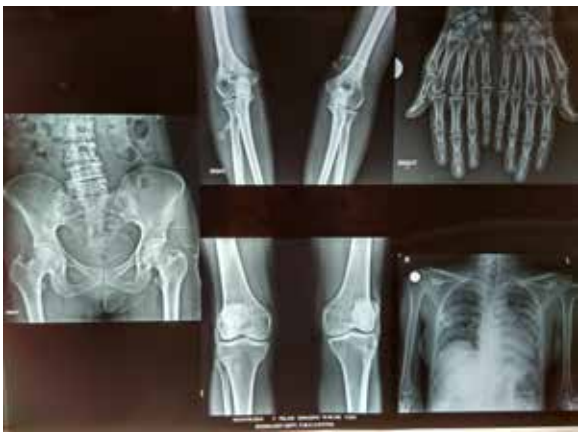


Figure 4: X-ray of Pelvis, Chest and Bilateral Wrist with Hands, Elbow and Knee

Treatment

There is not much data available about the protocol for treatment to be considered in patients of RA with concomitant ILD. It is a well known fact that Methotrexate, the primary drug used for treating Rheumatoid Arthritis itself causes ILD. Therefore the use of Methotrexate in such a patient can hardly be justified. Thus we were in a dilemma on deciding the regimen for treatment. With references to previous case reports and discussion with others involved it was finally decided to give the patient Pulse dosing of Methyl Prednisolone with Cyclophosphamide. After four pulse doses and concurrent administration of Cyclophosphamide the patient presented in our outdoor. She had shown significant improvement, her quality of life had improved with abetment in dyspnea (she was able to maintain a SpO2 of 98 %), her arthritis had subsided significantly. The patient is still under our follow up and seems to be doing well.

Discussion

Rheumatoid arthritis is a systemic inflammatory disorder that most commonly affects the joints, causing progressive, symmetric, erosive destruction of cartilage and bone, which is usually associated with autoantibody production. Rheumatoid arthritis affects around one percentage of the population in developed countries. The incidence and prevalence of rheumatoid arthritis in developing countries is thought to be lower, but is difficult to quantify[1]. Although joint disease is the main presentation, there are a number of extra-articular manifestations including subcutaneous nodule formation, vasculitis, inflammatory eye disease and lung disease

Respiratory symptoms in rheumatoid arthritis can be due to a variety of conditions that affect the parenchyma, pleura, airways or vasculature. Complications may arise directly from rheumatoid arthritis involvement or may occur secondary to immune-modulating medications used to treat rheumatoid arthritis. ILD is one of the most common pulmonary manifestation of rheumatoid arthritis lung disease, although the exact prevalence varies depending on the population studied and the diagnostic modality used to define the disease. ILD is heralded by symptoms of dry cough and progressive shortness of breath.ILD and is generally found in patient with higher disease activity. However it may be diagnosed in up to 3.5% of the onset of joint symptoms.[2]

Diagnosis is readily made by High Resolution Chest computed Tomography scan. Pulmonary function test shows a restricted pattern with a reduced diffused capacity for carbon monoxide. Different types of histopathology patterns of ILD has been associated with different types of connective tissue disorders. The predominant histopathology pattern in patients with scleroderma, polymyositis, sjogren syndrome and undifferentiated CTD is NSIP. In contrast, the currently available data show that, among RA-ILD patients, there is a higher proportion of patients with UIP pattern [when] compared with patients with

other CTDs. UIP to be the most common histopathologic pattern in RA-ILD patients. This was followed by NSIP and organizing pneumonia .the presence of ILD confers a poor prognosis[3] .The prognosis is not quite as poor as that of idiopathic pulmonary fibrosis because ILD secondary to RA respond more favourably than idiopathic ILD. The risk of developing ILD was higher in RA patients who were older at the time of disease onset, in male patients, and in individuals with more severe RA. The risk of death for RA patients with ILD was 3 times higher than in RA patients without ILD. Median survival after ILD diagnosis was only 2.6 years. ILD contributed approximately 13% to the excess mortality of RA patients when compared with the general population.[4]

Relative frequency of different histological subtypes of RA-ILD and their radiological pattern

Subtype	percentage	pattern
Usual interstitial pneumonia (UIP)	66%	Fibrosis, traction bronchiectasis and honey combing
Non-specific interstitial pneumonia (NSIP)	24%	Lower lobe volume loss and extensive ground glass opacity, reticular abnormality.
Cryptogenic organising pneumonia (COP)	4%	Multifocal peripheral consolidation, ground glass opacities, small nodular opacities and bronchial wall thickening and dilation.
Overlap syndrome (OS)	6%	mixed

The most striking feature of our case was the early onset of ILD in this patient of rheumatoid arthritis prior to the manifestation of joint symptoms. Of all literature available in the normal course of Rheumatoid Arthritis ILD usually present after long duration after the first symptoms of joints disease manifest. No credible explanation for this aberrancy was available in currently available literature. However, further understanding of the case based on molecular and pathological studies could give a clue on factors playing an important role in development of ILD in RA.

References:

1. Megan Shaw, Bridget F. Collins, Lawrence A. Ho, Ganesh Raghu Rheumatoid arthritis-associated lung disease European Respiratory Review 2015 24: 1-16;
2. Ankoor Shah, E.William St.Clair. Rheumatoid arthritis. In: Longo DL, Kasper DL, Jameson JL, Fauci AS, Hauser SL, Loscalzo J,19th eds. Harrison's Principles of Internal Medicine. USA: The McGraw-Hill Companies Inc., 2015:2136.
3. Esther Chan SpR in Rheumatology and General Medicine1,2 Kate Chapman SpR in Rheumatology1 clive Kelly consultant Physician and Rheumatologist1,2 1Department of Rheumatology, 2Department of Acute Medicine, Queen Elizabeth Hospital, Gateshead Interstitial lung disease in rheumatoid arthritis: a review arthritis research UK Issue 3 (Topical Reviews Series 7) Summer 2013
4. Bongartz T1, Nannini C, Medina-Velasquez YF, Achenbach SJ, Crowson CS, Ryu JH, Vassallo R, Gabriel SE, Matteson EL. Incidence and mortality of interstitial lung disease in rheumatoid arthritis: a population-based study Arthritis Rheum. 2010 Jun;62(6):1583-91