CASE REPORT

CONNECTIVE TISSUE DISEASE ASSOCIATED INTERSTITIAL LUNG DISEASE – AN ACUTE EXACERBATION IN EMERGENCY ROOM

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Interstitial lung disease (ILD) is a pathology involving lower respiratory tract, causing damage to the alveolar walls and vascular bed, resulting in decreasing functional alveolar units. Two third cases of ILD are idiopathic and one third cases of ILD are related to connective tissue disease (CTD). Clinical course of ILD is chronic and progressive in nature, CTD-ILD has a better prognosis compared to idiopathic ILD. Acute exacerbations are life-threatening and require close monitoring and immediate treatment, though CTD-ILD initial clinical manifestation at first clinical encounter is rarely found to be life threatening or acute exacerbation of ILD. I am reporting a case of CTD-ILD presenting to the Emergency Room of Memon Medical Institute Hospital (MMIH) manifesting as acute exacerbation of underlying ILD.

Keywords: Acute exacerbation; Interstitial lung disease; CTD-ILD; Connective tissue disease

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INTRODUCTION

Interstitial Lung Disease (ILD) consists of a group of disorders of the lower respiratory tract characterized by damaging of the alveolar walls and declining of functional alveolar capillary units. More than 100 agents can cause ILD but in two third cases, no cause can be identified.[1] Connective Tissue Disease associated ILD has a stable and chronic course, the most common histopathological pattern is non-specific interstitial pneumonia (NSIP) except for the rheumatoid arthritis in which Usual Interstitial Pneumonia (UIP) is the most common.[2] Systemic lupus erythematosus, rheumatoid arthritis, systemic sclerosis and polymyositis/dermatomyositis are the common CTD associated with the ILD.[3] The pharmacological treatment of CTD-ILD is not yet well defined and there are no global guidelines to follow.[4] Surgical biopsy, bronchoalveolar lavage and HRCT stay as the most important diagnostic investigations.[2]

CASE REPORT

A 35-year-old female patient with no known co-morbid presented to the ER triage counter on 16th April 2018 with the complaint of occasional shortness of breath for last 6 months and difficulty in breathing for last 15 days. Her vital signs were heart rate 150 beats/min (regular), transcutaneous oxygen saturation of 90% on room air, respiratory rate 35/min and temperature 38 C. In family history, her mother and sister were known cases of Systemic Lupus Erythematosus and patient had a generalized small joint pain for last one year. On examination, patient was grade IV (modified medical research counsel dyspnoea scale) dyspnoeic, tachypnoeic (35/min) and tachycardic (150/min), breathing pattern was regular but laboured, lung auscultation revealed diffuse crept throughout the lung fields bilaterally. Her health record showed positive anti-citrullinated cyclic protein (Anti-CCP) and Anti-Nuclear Antibody (ANA). After initial assessment, provisional diagnosis of Acute Pulmonary Embolism versus CTD-ILD was made, management started accordingly. Initial investigations revealed pH 7.231, pCO2 50.7 mmHg, pO2 193.1 mmHg, HCO3 21.5, Pro-BNP 5844, CRP 117 mg/L, WBC 16.7 /Cumm, Haemoglobin 14.3 gm/dl, platelets 437 /Cumm, Na 146 mEq/L, K 4.9 mEq/L, HCO3 20 mEq/L, Calcium 10.1 mg/dl, Creatinine 0.7 mg/dl, Magnesium 2.0 mg/dl, Urea 45 mg/dl.

Patient was taken on Bi-PAP followed by the blood gases report and admitted to the Intensive Care Unit. Chest X-ray showed diffuse bilateral reticulonodular changes and prominent vascular and bronchial markings, echocardiography revealed dilated right sided chambers, normal size left ventricle with normal systolic function, mild tricuspid regurgitation and mild pulmonary artery hypertension. Serial blood gases in ICU revealed progressively declining pH and GCS deteriorated from 15/15 to 10/15, patient was intubated for impending respiratory failure (type II). CT pulmonary angiogram was done to differentiate between acute pulmonary embolism and CTD-ILD, CT scan showed extensive honey combing in superior and basal segments of both lungs lower lobes with associated traction bronchiectasis, no evidence of pulmonary embolism, findings were most likely due to usual interstitial pneumonia (UIP). Bronchoalveolar lavage and PFTs were planned once patient become stabilized.

Patient developed acute hypotension and anuria on 20th April 2018, patient was given fluids but didn't respond, patient was started on Norepinephrine to maintain blood pressure and perfusion to the vital organs, subsequently Creatinine increased to 2.67 and pH declined to 7.187 with pCO2 of 50.7 mmHg and HCO3 of 19.4 mEq/L. On 21st April 2018, patient made about 300 ml, blood gases revealed severe respiratory acidosis (pH 6.919, pCO2 107.5 mmHg, pO2 75.0 mmHg, HCO3 22.2 mEq/L) along with the Haemoglobin of 9.0 g/dl, WBC 14.82/Cumm and Platelets 300/Cumm, patient developed asystole in the evening, despite performing CPR for 10 mins, patient died of cardiopulmonary arrest. As pulmonary embolism was ruled out, CTD-ILD acute exacerbation diagnosis was made based on history, serological markers and HRCT.

DISCUSSION

Connective tissue disease (CTD) associated ILD has a better prognosis compared to non-classifiable ILD and has a chronic course.[3] Acute exacerbations are life-threatening and need an immediate identification and treatment. Diagnosing and managing any CTD-ILD in the early course helps to avoid life-threatening exacerbations and poor outcomes. Since the optimal time for intervening is not defined, acute exacerbations remain a big risk.[2,5]

This patient presented with a history of difficulty breathing and prolonged generalized small joint pain including a strong family history of SLE and positive anti- CCP and ANA titers, it strongly indicated an underlying CTD. UIP on HRCT and positive anti-CCP titers make rheumatoid arthritis more likely as an underlying CTD in the appropriate clinical context, since UIP is only found to be the most common histopathological pattern related to RA compared to any other CTD [2]. On the other hand, strong family history of SLE and positive ANA titers increase the likelihood of SLE as an underlying CTD and anti-phospholipid syndrome contributing to pulmonary embolism as well. In such setting, HRCT plays a vital role in differentiating.

Special attention must be given to the patients with a family history of connective tissue disease and positive serological markers, developing mild symptoms, to avoid life-threatening acute exacerbation. More research work is indicated to devise structured management plan and decide about the timing of intervening.

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