UNDIFFERENTIATED CONNECTIVE TISSUE DISEASE-ASSOCIATED INTERSTITIAL LUNG DISEASE: A CASE REPORT.

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Abstract

There is insufficient data on Undifferentiated Connective Tissue Disease-Associated Interstitial Lung Disease (UCTD-ILD) in Nigeria as most may have been diagnosed as pulmonary tuberculosis and treated as such. It is uncommon for UCTD to present initially with ILD, therefore a high index of suspicion is required in making a diagnosis of any connective tissue disease as there are no diagnostic tests for any. We report a case of UCTD-ILD in a 28-year-old teacher who presented in right heart failure.

Keyword: Undifferentiated Connective Tissue Disease, Interstitial Lung Disease.

Introduction

Patients defined as having undifferentiated connective tissue disease (UCTD) are those who although do not fulfill a current diagnostic criteria for a specific connective tissue disease (CTD) present with features suggestive of a CTD. There is currently no classification criteria for UCTD, some of these patients (10%) will over time evolve into specific CTD such as systemic sclerosis, rheumatoid arthritis and spondyloarthropathy. A very common pattern of presentation is new onset Raynaud’s phenomenon with abnormal capillaroscopic features.

Few reports have described lung disease as the initial presentation of UCTD, and none have been reported in Nigeria. In this report we discuss the case and review the associated literature of UCTD.

Case report

A 28-year-old teacher who presented with a two year history of non-productive cough which was associated with easy fatigability, dyspnoea on exertion, wheezing, non-plueritic chest pain and orthopnoea. Three months prior to presentation, she developed progressively worsening pedal oedema and abdominal distension. There were also complaints of palpitation, malaise and weight loss. She was previously managed for
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pulmonary tuberculosis at a peripheral centre based on chest x-ray findings. She had no past history of smoking or chronic exposure to dust particulate matter. She did not meet the diagnostic criteria for any of the connective tissue diseases. General examination revealed a lady in respiratory distress, with an SPO2 of 85% on room air, cyanosis and bilateral pitting oedema of the lower limbs up to the thighs. She had an S3 gallop rhythm with a pulse rate of 122 beats per minute, a loud pulmonary component of the second heart sound, coarse crepitation in middle and lower lung zones bilaterally, a tender hepatomegaly and ascites. An initial diagnosis of idiopathic pulmonary fibrosis with right heart failure was made.

Computed tomography (CT) scan of the lungs revealed ground-glassing with reticular densities in the entire left lung with an area of opacity and air bronchogram in the lung base. There was also patchy ground-glassing involving the right lung associated with reticular densities (Fig.1). Pulmonary function test showed a restrictive pattern (FVC: 1.99, 59% of predicted; FEV1: 1.54, 52% of predicted; FEV1/FVC: 92%) (Fig.2). She had a positive connective tissue disease screen, and a positive antinuclear antibody assay with a titre of 1:1280 (speckled pattern).

Echocardiography showed a pulmonary acceleration time of 40msecs (normal >130 msecs) which was in keeping with pulmonary hypertension. Sputum for acid-fast bacilli and geneXpert for Mycobacterium tuberculosis were both negative. At this point her diagnosis was reviewed to undifferentiated connective tissue disease associated interstitial lung disease complicated by right heart failure.

She was commenced on mycophenolate mofetil, prednisolone (which was tapered over 3 months), tadalafil, spironolactone, torsemide and long-term oxygen therapy with the use of oxygen concentrator at home.
On follow-up, she improved clinically with resolution of symptoms and a SPO2 of >92% on room air.

**Discussion**

The term undifferentiated connective tissue disease is used to describe clinical features of CTD in patients who do not fulfill current criteria for a specific CTD. No diagnostic criteria exist for UCTD but proposals have been made by Mosca et al.

The preliminary criteria for UCTD include the following: at least one clinical manifestation of CTD, a positive anti-nuclear antibody test and disease duration of at least 3 years. Typical features of UCTD are Raynaud’s phenomenon, arthralgia, arthritis, sicca symptoms, alopecia, photosensitive rash, malar rash, sclerodactyly, leucopenia, thrombocytopenia and anemia. Raynaud’s phenomenon occurs in majority of patients. This was however absent in our patient.

The respiratory system is frequently affected in CTD and may involve every component including the lung parenchyma. Interstitial lung disease may complicate any connective tissue disease with the pathology dominated by inflammation, fibrosis or both, with distinct radiologic and histopathologic patterns. Interstitial lung disease may precede the development of other rheumatic symptoms in UCTD. Due to the presentation of chronic cough in UCTD-ILD, it is common for physicians to consider pulmonary tuberculosis in our environment. However, in the absence of positive acid fast bacilli on microscopy and negative geneXpert, ILD should be considered.

Although most patients continue to have a diagnosis of UCTD, some later develop additional symptoms and subsequently fulfill...
the criteria for a specific CTD. This emphasizes the need for continued follow up of patients. Laboratory investigations are necessary to identify markers which may suggest an autoimmune disease. A seven-centre cross sectional study found out that over 94.5% of patients with UCTD are female and virtually all of them were positive for ANA.\textsuperscript{5} Investigations required depends on the clinical manifestations. For those patients with ILD, a high resolution CT scan, pulmonary function test, lung biopsy and serology are sufficient to make a diagnosis. Echocardiography and/or cardiac catheterization is required for pulmonary hypertension. The non-specific interstitial pneumonitis (NSIP) pattern occurs in over 80% of patients with UCTD-ILD.\textsuperscript{6}

To date, no effective treatment exist for the management of any form of connective tissue disease-associated ILD. Non-specific interstitial pneumonitis is the commonest pulmonary manifestation, hence immunosuppressive drugs like corticosteroids, cyclophosphamide, azathioprine and mycophenolate mofetil are generally used. These have shown some modest benefit although none have resulted in a cure.

**Conclusion**

We have discussed a case of UCTD-ILD in a patient who had twice received full course of anti-koch’s treatment. A high index of suspicion and a multidisciplinary approach in diagnosis and management is required. CTD should always be a consideration in young females with chronic ill-health. A limitation encountered was our inability to have a histologic diagnosis via lung biopsy.

**REFERENCES**


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