Scholars Journal of Medical Case Reports **ISSN 2347-6559 (Online)**
Sch J Med Case Rep 2015; 3(9B):928-932 **ISSN 2347-9507 (Print)**

(An International Publisher for Academic and Scientific Resources) DOI: 10.36347/sjmcr.2015.v03i09.042

Early Pulmonary Manifestations of Diffuse type of Systemic Sclerosis: Case Report

Dr. C. Sumalata[1], Dr. P. Ajoy Kumar[2], Dr.S.B.Lal[3],
[1]Medical Officer, District Tuberculosis Office, Nalgonda, Telangana, India
[2]Post-Graduate, Department Of Pulmonology, Siddhartha Medical College, Vijayawada, Andhra Pradesh, India
[3]Registrar,Dr.NTRUHS,Vijayawada. Ex-Professor and HOD ,Department of Pulmonology, Siddhartha Medical College,

***Corresponding author**
Dr. C. Sumalata
Email: drsumachittiboyina@gmail.com

Abstract: Systemic sclerosis (SSc) is a multisystem disease that is associated with inflammation, fibrosis and vasculopathy. There are two major subsets of systemic sclerosis, limited cutaneous SSc and diffuse cutaneous SSc (dcSSc). Respiratory symptoms are common in patients with SSc, but physical examination often fails to establish if the underlying cause is interstitial lung disease (ILD), pulmonary arterial hypertension (PAH), impaired locomotion due to systemic disease or loss of fitness. Impaired lung function is usually evident on pulmonary function testing, with the pattern of functional impairment often discriminating usefully between ILD and PAH. The presence of PAH can be indicated by echocardiography and must be confirmed by right heart catheterization. Whereas chest radiographs detect established ILD, high-resolution CT can identify earlier or very mild inflammatory changes. Here we present a case, where a young lady presented with diffuse type of systemic sclerosis. She was given a multidisciplinary approach, and a thorough systemic examination of Dermatology, Cardiology, Pulmonology, and Gastroenterology was done. She was investigated and advised accordingly.
Keywords: Respiratory complaints, interstitial lung Diseases, Systemic sclerosis, diffuse type.

INTRODUCTION

Lung involvement in systemic sclerosis was first described at the end of the last century (Lewin and Heller, 1894) [1]. Patients with SSc may exhibit proliferative small artery and obliterative micro vascular disease, plus inflammation and fibrosis affecting the skin, oesophagus, respiratory tract and other target organs. Pulmonary involvement is common in patients with SSc and most often comprises fibrosis or interstitial lung disease (ILD), and pulmonary vascular disease leading to pulmonary arterial hypertension (PAH). Pulmonary manifestations are the leading cause of disease-related morbidity and mortality in patients with SSc [2]. ILD in SSc may account for 16% of the deaths [3].

CASE REPORT

A 28 year old female from Vijayawada who is a home-maker came to pulmonary medicine OP with chief complaints of cough and shortness of breath on exertion since 20 days. She neither complained of sputum nor haemoptysis nor chest pain. Her parents marriage was not consanguinous. She had complaints of pigmentation of skin and thickening of dorsal surfaces of hands. She was moderately built and nourished. She had microstomia. On cutaneous examination she showed shiny light hidebound skin over extensor aspects of forearm and front of the legs. Salt and pepper appearance over both ears. Digital pitting scars were also seen. ENT Examination was normal. On examination of chest she had coarse crepitations over basal regions. Cardiovascular examination was unremarkable. Per abdomen was soft. CNS was normal. Vitals were in normal limits. Reynaud's phenomenon was not elicited.

She was subjected to few investigations and results were as follows:

Hb-10.4 g/dl, TC-7,500/cc, DC- polymorph-55 lymphocytes-40 eosinophils-3 monocytes -2

Complete Haemogram showed mild degree of microcytic hypochromic anaemia

ESR-15mm/Hr, RBS-84mg/dl, Serum creatinine-0.6mg/dl,

Liver Function Tests: Serum bilirubin 0.6mg/dl, SGOT-18IU/L, SGPT-20IU/L.

ANA profile: ANA-1:40 titres. Anti Scl70 Ab--2.43.Anti centromeres were negative.

Skin Biopsy suggestive of Systemic Sclerosis.

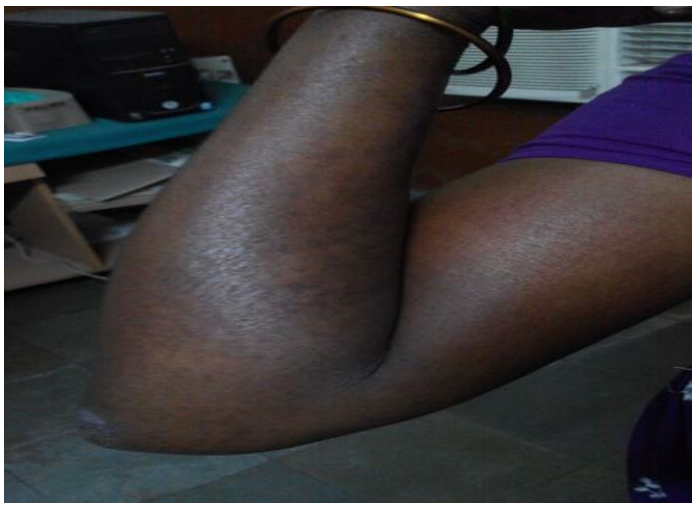


Fig 1: Skin lesions over forearm

Fig 2 &3: Digital pitting scars were also seen

Chest X-Ray shows as follows:

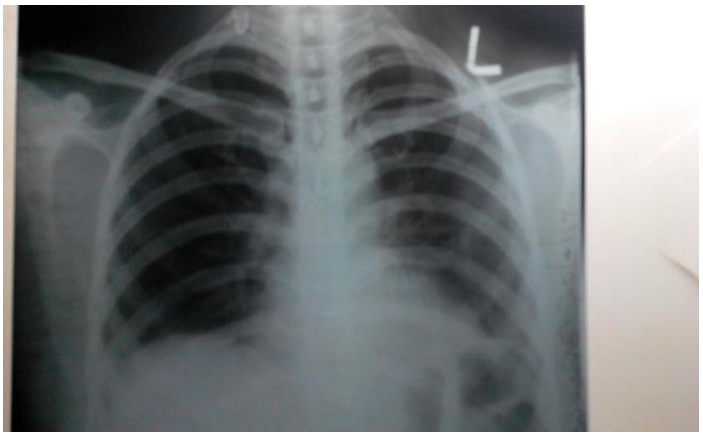


Fig 4: Hazy shadowing seen in left lower zone, there is no obliteration of Costophrenic angle

Computed Tomography of Chest shows:

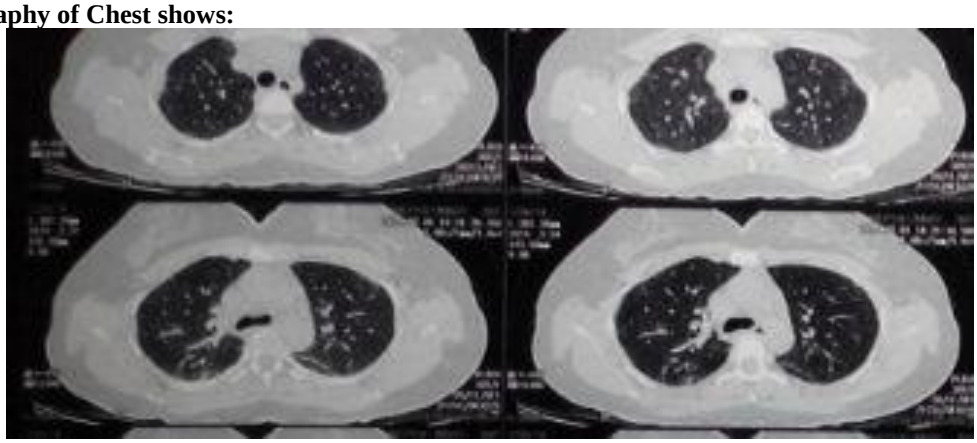
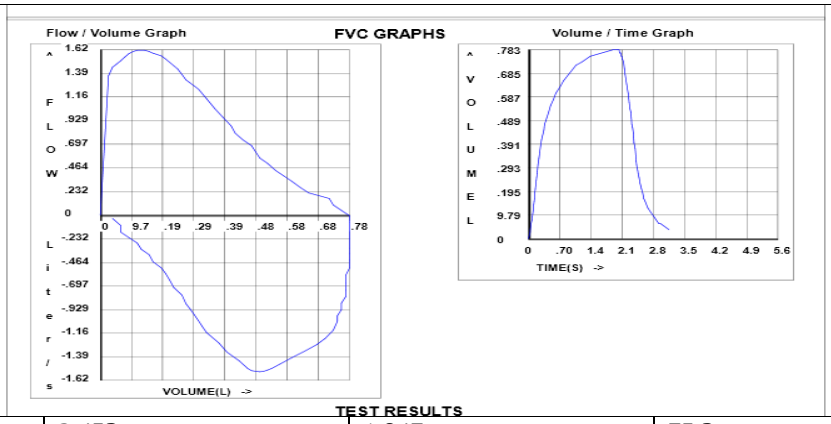


Fig 5: Small nodular shadows at the base of lung, Ground glass opacity was also seen

Pulmonary function Tests shows:



FVC	2.453	1.847	75.3
FEV1	2.118	1.68	79.6

Fig 6: Pulmonary Function Test showing obstructive and restrictive Pattern

Spo2 was maintained at 92% at rest, Bronchoscopy was done, and Broncho alveolar lavage findings were not significant.

ECG was as follows:

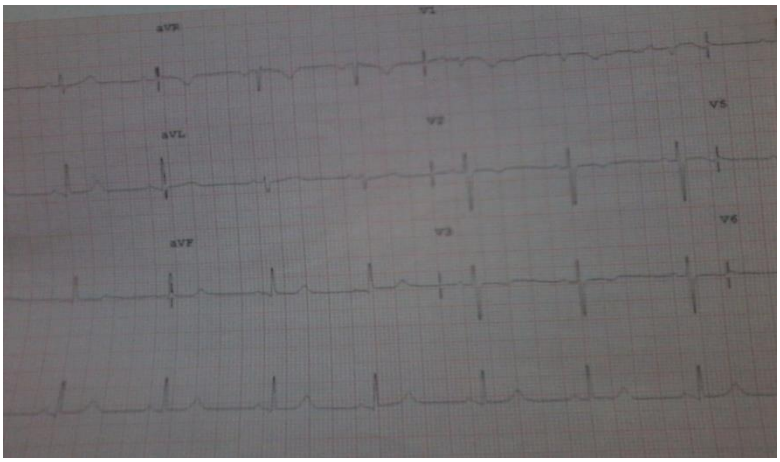


Fig-7: Echocardiography

Echocardiography was normal.

Barium Swallow showed:

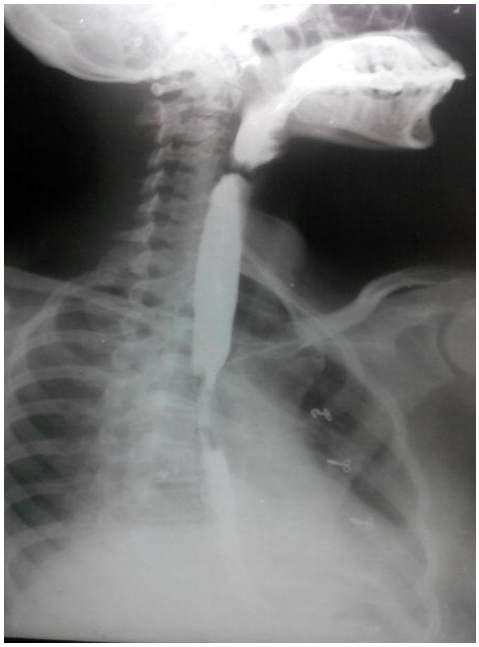


Fig-7: Showed narrowing at the lower 2/3 rd of esophagus, Delayed emptying of barium.

Upper GI endoscopy showed mild antral gastritis. The case was diagnosed as diffuse type of Systemic Sclerosis. She was given steroids which were tapered slowly. Her symptoms improved. She was advised for a regular follow up.

DISCUSSION

Lung Manifestation of systemic sclerosis included Intestinal lung Diseases and Pulmonary Artery Hypertension. The case in our report had complains of cough and shortness of breath. The general examination revealed cutaneous manifestations. All systems were examined to rule out any early manifestations like oesophageal dysmotility, Reynaud's phenomenon, etc. The diagnosis was made diffuse type because of involvement of skin of extremities, face and digital pitting scars, along with the respiratory involvement in the form of Interstitial Lung Disease[ILD}.Limited form usually manifests as Pulmonary Artery Hypertension [PAH] rather than ILD. The diagnosis was confirmed after looking ANA profile .Cutaneous lesions along with systemic manifestations should always raise the suspicion of connective tissue disorders. Connective tissue disorders involve all systems and therefore detailed examination of each and every system is mandatory and investigations are to be advised accordingly. The case here was young women who showed not much findings on examination of Chest except for crepitations in the basal areas. The radiological investigations revealed early nodular shadows in the basal areas, especially in a HRCT chest which prompted to consider the possibility of Connective Tissue disorders. The case here showed early manifestations. Her pulmonary function Tests showed both obstructive and restrictive pattern. However BAL did not reveal any results. She was explained the same and was asked for a check up at cardiology and Gastroenterology. Barium swallow revealed narrowing at upper end of oesophagus. Along with the same she was advised for ANA profile.

There is always a challenge for the respiratory clinicians to determine the underlying cause when patient complains of cough, chest pain, fatigue, non-productive cough. Early signs of ILD include septal and sub pleural line opacities, ground-glass opacities and subpleural cysts. As fibrotic lung disease progresses, isolated areas of opacity are replaced by apparent honeycomb or micro-honeycomb formation and/or traction bronchiectasis/bronchiolectasis[4]. The history and the findings in the case initially made us to put a differential Diagnosis of Basal Pneumonia and Pulmonary Edema which were ruled out with subsequent workup of the case.

The respiratory involvement of systemic sclerosis is in interest because of the consequences it would lead to and finally landing in catastrophic respiratory ailments. Respiratory symptoms can be non-specific, and early-stage pulmonary fibrosis can progress undetected. However, the identification and staging of pulmonary manifestations is of paramount importance to the management of patients. Lung involvement is defined as 'normal' when FVC and DLCO are >80% of the predicted values adjusted for age, sex and height. It has been proposed that disease severity in patients with SSc be defined as mild, moderate or severe when these measures are 70–79, 50–

69 and <50 of the predicted values, respectively [5]. Decreases in DLCO over3 years are closely associated with increased mortality [6]. The disease could affect chest muscles and may restrict the work of breathing which in turn may lead to respiratory failure. This case was been brought to discussion because the possibility of Connective Tissue Disorders was often missed as routine investigations may mimic infective aetiology and early manifestations may be overlooked with other differential diagnosis. An examination of all systems becomes mandatory when such suspicion arises. A multidisciplinary team consisting of dermatologist, gastroenterologist, Cardiologist, respiratory physician, radiologist is needed to make a correct diagnosis and help patient in preventing irreversible functional changes.

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