(International Print/Online Journal)

SJIF IMPACT FACTOR: 5.565
PUBMED-National Library of Medicine ID-101739732

ISSN (Print): 2209-2870
ISSN (Online): 2209-2862

International Journal of Medical Science and Current Research (IJMSCR)
Available online at: www.ijmscr.com
Volume 5, Issue 4 , Page No: 175-179
July-August 2022

IJMSCR

Systemic Sclerosis Sine Scleroderma Associated Interstitial Lung Disease

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Type of Publication: Original Research Paper
Conflicts of Interest: Nil

Abstract

Background:
Evidence of ILD can be found in upto 65% of SSc patients by HRCT, clinically significant ILD develops in 16-43%.ILD is one of the 1st clinical manifestations of systemic sclerosis sine scleroderma.

Description:34/F housewife r/o Panvel presented to opd with c/o breathlessness on exertion MMRC II and fatigue since 2 years. No skin tightening,digital ulceration,tanning of skin.

Conclusion: Decision to treat SSc sine scleroderma patients with ILD is strongly influenced by FVC and DLCO, extent of disease on HRCT and duration of disease. Early decline in FVC is strongly predictive of severe ILD which develops in first 4 years.

Keywords: Systemic sclerosis sine scleroderma, intertital lung disease

Introduction

Systemic sclerosis is a complex and clinically heterogeneous orphan disease with protean clinical manifestations, a chronic and frequently progressive course causing significant disability, disfigurement and mortality. Systemic sclerosis sine scleroderma (ssSSc) was first described in 1962 by Rodnan and Fennel [3]. ssSSc is defined by specific organ involvement in the absence of skin thickening. Systemic Sclerosis sine scleroderma is a variant of Systemic sclerosis which shares visceral, serological and vascular manifestations but lacks skin thickening. Systemic Sclerosis sine scleroderma still remains a rarely reported subtype of Systemic Sclerosis unlike commonly reported limited cutaneous and diffuse cutaneous types.

While evidence of ILD can be found in upto 65% of SSc patients by HRCT,clinically significant ILD develops in 16-43%.

Case Report:

34/F housewife r/o Panvel presented to opd with c/o breathlessness on exertion MMRC II and fatigue since 2 years. No skin tightening,digital ulceration,tanning of skin.

No history of photosensitivity/malar rash/recurrent oral ulcers/fever/dryness of mouth or eyes/Raynaud's phenomenon/digital ulcers/cough/joint pain/restricted joint movements

No GI/urinary complaints

H/o similar complaints in sister since 3 years.

No other co morbidities

O/E -

Pallor- present,

Ingram sign and Barnett's sign-negative

Mouth opening-3 fingers,

Nail fold capilloaroscopy-dilated giant capillaries present.

RS-B/l basal fine crepts in inframammary, infra axillary and infra scapular region

	Dec	March
FVC	45.4%	56%
RV/TLC	157.4	143.4
DLCO	26%	37.3%

Restrictive ventilatory impairment with air trapping with severe diffusion defect.

2D Echo- No PAH, LVEF-60%

Rest S/E-Normal.

Diagnosis

Hb-11.2

Tlc-11,000

Pt-2.7lac

Cr-0.62

Sgot/Sgpt-20/12

ESR-45

Urine r/m-nad

Urine protein-3.48

24hr urinary protein-0.1012g/24hr

ANA positive, Scl 70 positive.

HRCT s/o multifocal patchy areas of subpleural ground glass attenuation in all lobes, reticular opacities, traction bronchiectasis present in poster basal segment of B/l lobes s/o NSIP pattern.

PFT s/o

Management: Patient was started on Wysolone 10mg OD f/b tapering doses and Azathioprine 50mg OD over 3months.

F/u PFT shows significant improvement in FVC and FEV1

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Figure 1 : CXR -Normal

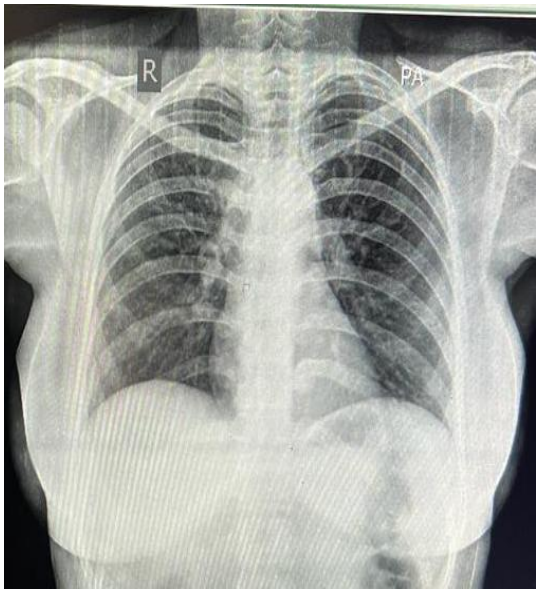

Figure 2 : Restrictive ventilatory impairment on PFT

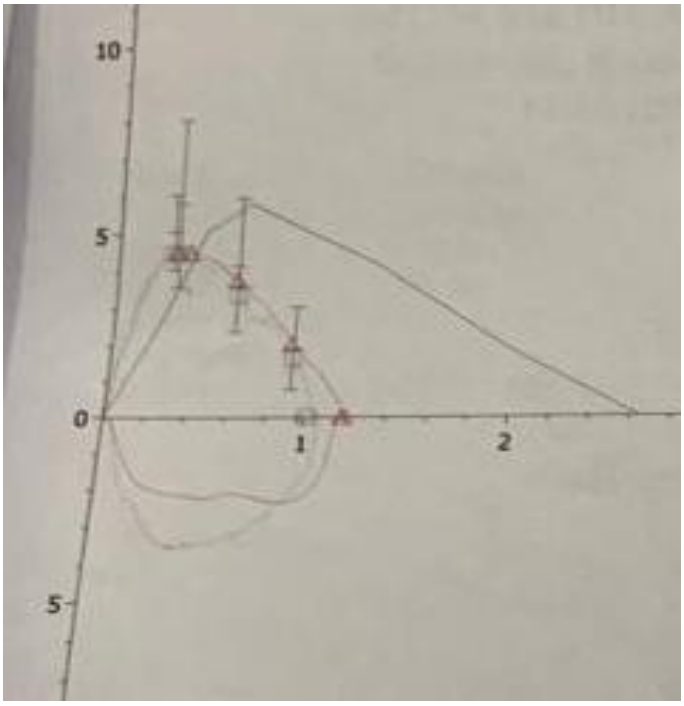

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Figure 3: HRCT : Sub pleural ground glass attenuation with reticular opacities

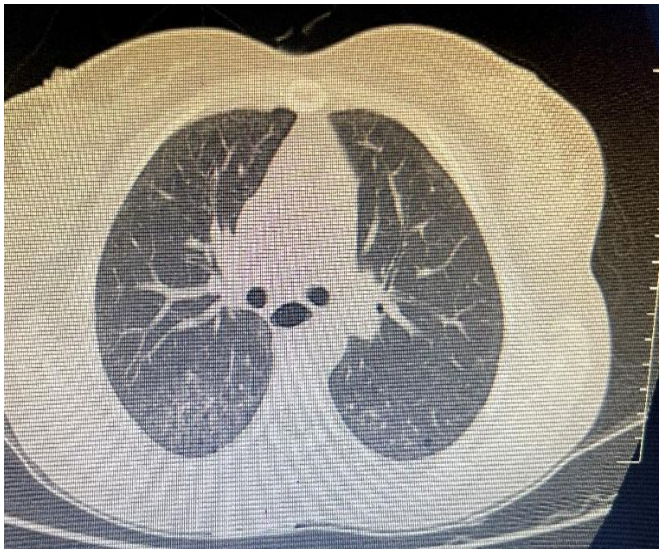
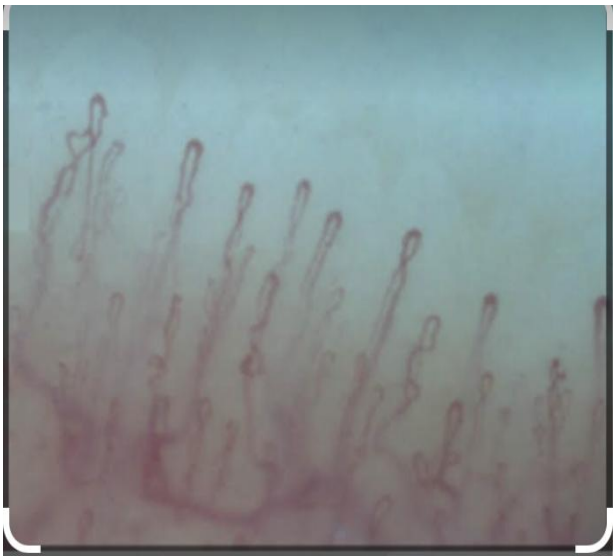


Figure 4: Nail fold capillaroscopy Showing giant and dilated capillaries



Discussion:

The prevalence of lung disease in SSc sine scleroderma depends on method used for detection. Dysnea is present in roughly 55% of patients .Cough ,a less frequently reported symptom tends to be dry and nonproductive. Hemoptysis is rare but may complicate carcinoma or bronchial telangiectasia. Lung disease maybe the first manifestation of SSc sine scleroderma. Exercise lung function testing increases ventilation perfusion mismatching and also increases diffusion abnormalities resulting in hypoxemia and widening of alveolar arterial oxygen difference.BAL may indentify neutrophilic alveolitis which predicts a more progressive disease.However, an increase in neutrophils reflects an increase in extent of disease on CT, particularly of reticular pattern, thus this increase is likely a marker of more extensive disease rather than an independent index of progressive disease. Many patients with apparently normal BAL findings may exhibit progressive disease. Hence BAL not recommended routinely for diagnosis or monitoring of patients with ILD.Surgical lung biopsy is virtually never required in diagnosis of SSc sine scleroderma interstitial lung disease.Pathologically the most prevalent pattern is NSIP with alveolar wall thickening mixed with inflammatory cells,connective tissue matrix cells and proteins combined with type II pneumocyte proliferation and vascular obliteration.

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Conclusion:

Although crude mortality are 3.9%/year for men and 2.6%/year for women,lung disease remains the most common cause of death in patients with SSc.Thus decision to treat SSc sine scleroderma patients with ILD is strongly influenced by FVC and DLCO, extent of disease on HRCT and duration of disease. Early decline in FVC is strongly predictive of severe ILD which develops in first 4 years.Low dose steroid theraphy remains justified as an invaluable adjunct to the treatment of lung disease.

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