Original Research Article

A study on clinical profile of systemic sclerosis and outcome

Pazhamalai K*

Chief Civil Surgeon, Government Headquarters Hospital, Kallakurichi, Tamil Nadu, India
*Corresponding author email: **pazhamalaikandasamy541@gmail.com**

International Archives of Integrated Medicine, Vol. 8, Issue 4, April, 2021.
Available online at http://iaimjournal.com/
ISSN: 2394-0026 (P) **ISSN: 2394-0034 (O)**
Received on: 01-03-2021 **Accepted on:** 18-03-2021
Source of support: Nil **Conflict of interest:** None declared.
How to cite this article: Pazhamalai K. A study on clinical profile of systemic sclerosis and outcome. IAIM, 2021; 8(4): 24-30.

Abstract

Introduction: Systemic sclerosis is a chronic multisystem disorder of unknown etiology characterized clinically by thickening of the skin caused by the accumulation of collagen and by structural and functional abnormalities of visceral organs including the gastrointestinal tract, lungs, heart, and kidneys. Systemic sclerosis is often of tragic consequence to patients. Survival is determined by the severity of the visceral disease, especially involving the lungs, heart, and/or kidneys.

Aim of the study: To study the clinical spectrum of patients presenting with Systemic Sclerosis (SSc) and to evaluate the internal organ involvement.

Materials and methods: Patients who attended the General Medicine Department of Government Headquarters Hospital, Kallakurichi, Tamil Nadu from January 2019 to September 2019 were taken up for the study. Systemic sclerosis was diagnosed in 25 patients as per the American College of Rheumatology.

Results: The initial presentation was skin manifestation (52%) Arthralgia (40%), Raynaud's (8%) Visceral symptom (4%), Cumulative manifestations included skin (Cutaneous) 100%, Diffuse skin Involvement was seen in (4%), Arthralgia (80%). No one presented with fully developed CREST syndrome. Calcinosis – Nil, Reynaud's was 28%. Joint involvement in the form of arthralgia was seen in 80%. Gastro-Intestinal system was involved in 72%. The respiratory system was involved in 16%. The cardiovascular system was involved in 16%. The renal system was not involved even in one case.

Conclusion: Sclerodactyly and pigmentary changes are more common. Raynaud's phenomenon is very much less probably due to climatic causes. A fully developed CREST was not present in this study. Resorptions of terminal phalanges were less common. Gastrointestinal involvement is more than in other studies. Respiratory involvement was low compared to other studies. Cardiac involvement is less like other studies. Renal involvement is NIL in this study.

Key words

Systemic sclerosis, Skin lesion, Organ involvement, Clinical profile.

Introduction

Systemic sclerosis is a chronic multisystem disorder of unknown etiology characterized clinically by thickening of the skin caused by the accumulation of collagen and by structural and functional abnormalities of visceral organs including the gastrointestinal tract, lungs, heart, and kidneys. Systemic sclerosis is often of tragic consequence to patients. Survival is determined by the severity of the visceral disease, especially involving the lungs, heart, and/or kidneys. Even though current clinical and diagnostic utilities have led to a better understanding of the disease, its pathogenesis remains unknown [1]. Scleroderma is a heterogeneous disease with a wide range of clinical manifestations ranging from mild skin fibrosis with minimal internal organ disease to severe skin and organ involvement. The three main pathological events that are involved in scleroderma pathogenesis are mainly endothelial damage, fibrosis, and autoimmune dysregulation [2]. Etiopathogenesis of scleroderma is characterized by fibroproliferative alterations, cellular and humoral immune abnormalities resulting in a severe and often progressive fibrotic process. Scleroderma can also be subdivided according to different criteria, such as involvement of organs and the presence of specific antibodies which are hallmarks of the disease [3]. These autoantibodies are disease-specific and usually mutually exclusive and correlate with the extent of skin involvement and associated disease manifestations. The most common is DNA topoisomerase (anti-Scl70), anti-centromere antibodies (CENP A and/or B protein) [4]. These autoantibodies are marker antibodies for relatively distinct clinical phenotypes of SSc where anti-Scl70 antibodies are a marker for dcSSc and SSc patients with clinically significant pulmonary fibrosis with a poor prognosis whereas anti-centromere antibodies typically are associated with lcSSc, uncommon pulmonary fibrosis, and late-onset of pulmonary hypertension but generally are associated with an overall good prognosis [5, 6].

Materials and methods

Patients who attended the General Medicine Department of Government Headquarters Hospital, Kallakurichi, Tamil Nadu from January 2019 to September 2019 were taken up for the study. Systemic sclerosis was diagnosed in 25 patients as per the American College of Rheumatology.

Major criteria: Sclerodermatous skin change in any location proximal to the Metacarpophalangeal joints.

Minor criteria: Sclerodactyly, Digital pitting scars of fingertips (or) Loss of digital finger pad substance, and Bibasillar pulmonary fibrosis. One Major criteria (or) two or three minor criteria. The sensitivity of these criteria was 97% and the specificity 98% by applying this American College of Rheumatology Criteria. All 25 patients were analyzed from clinical, Biochemical, Immunological, and radiological parameters.

Results

The initial presentation was skin symptoms in 13 patients (52%). Arthralgia in 10 patients (40%) and Raynaud's phenomenon in 2 patients (8%) and visceral symptoms (Dyspnoea on exertion) in one patient (4%). Majority of patients had their symptoms between 30 – 50 years of age (**Table – 1**).

25 patients had skin changes (100%), 20 patients had Arthralgia/ Arthritis (80%), 7 patients had Raynaud's phenomenon (28%), 7 patients had visceral symptoms (28%) as per **Table – 2**.

Skin involvement was seen in all patients. Pigmentary changes were more common (68%). Sclerodactyly – next common manifestation

(40%) as per **Table - 3**. Vasospastic changes were present in 7 patients. This is comparatively less than other studies (**Table – 4**). Joint manifestation was seen in 20 patients (80%) as per **Table - 5**.

Out of 25 patients 18 patients presented with Dysphagia and Regurgitation. All the symptomatic patients underwent Ba Swallow and upper Gastro-Intestinal Endoscopy. Patients presented with Dysphagia showed dilated esophagus in Barium Swallow studies (48%). Patients presented with Regurgitation showed gastroesophageal reflux disease on upper gastrointestinal endoscopy (24%) as per **Table – 6**.

Out of 25 patients 4 had a symptom of Respiratory system in the form of Exertional Dyspnea for which all patients were investigated with chest x-ray, pulmonary function test, and CT chest. Out of 4 patients who had symptoms of Exertional Dyspnea, the X-ray chest was normal for 2 patients. X-ray chest: showed basal (B/L) Haziness for another 2 patients. PFT: showed mild to moderate Restriction airway disease for 2 patients who had X-ray normal.PIT: showed moderate to severe restriction airway disease for 2 patients who had x-ray B/l basal Haziness.CT Chest: showed Interstitial lung disease for 2 patients who had X-ray findings and PFT showed moderate to severe restriction airway disease. Another 2 patients CT normal study (**Table – 7**).

Table – 1: Symptom analysis.

Symptoms	**No. of patients**	**%**
Skin	13	52%
Arthralgia	10	40%
Raynaud's Phenomenon	2	8%
Visceral symptoms	1	4%

Table – 2: Cumulative manifestations.

Symptoms	**No. of patients**	**%**
Skin	25/25	100%
Joint	20/25	80%
Raynaud's phenomenon	7/25	28%
Visceral manifestation	7/25	28%

Table – 3: Skin involvement (cutaneous).

Clinical features	**No. of patients**	**%**
Limited cutaneous Sclerodactyly Pigmentary changes (Hyper Pigmentation & Pepper salt pigmentation)	 10 17	 40 68
Calcinosis	0	0
Ulceration, Stellate scars	15	60
Telangiectasia	0	0
Resorption Hand	1	4
CREST	0	0
Vasculopathy ulcer	1	4
Diffuse cutaneous	1	4

Table - 4: Vasospastic changes (raynaud's phenomenon).

Features	No. of patients	%
Vasospastic changes	7/25	28%
Severity		
Mild	3/25	12
Severe	4/25	16
Involvement		
Unilateral	3/25	12
Bilateral	4/25	16

Table – 5: Joint involvement.

Joint involvement features	No. of patients	%
MinorJoints	**13/25**	**52**
Major joints	**7/25**	**28**
Deformity	**5/25**	**20**

Table – 6: Gastrointestinal tract manifestations.

Symptoms	No. of patients	%	Ba Swallow	Upper gastroIntestinal Endoscopy
Dysphagia	**12/25**	**48%**	**Dilated esophagus**	**Normal**
Regurgitation	**6/25**	**24%**	**Normal**	**Gastroesophageal Reflux disease**

Table – 7: Respiratory system.

Patient No.	Symptom	Chest X-ray	P.F.T.	CT Chest
Patient No.1	Exertional dyspnea	B/L basal Haziness	Mod to severe restrictive airway disease	Interstitial lung disease
Patient No.2	Exertional dyspnea	B/L basal Haziness	Mod to severe restrictive airway disease	Interstitial lung disease
Patient No.3	Exertional dyspnea	Normal study	Mild to moderate restrictive airway disease	Normal study
Patient No.4	Exertional dyspnea	Normal study	Mild to moderate restrictive airway disease	Normal study

Out of 25 patients symptoms attribute to the Cardiovascular system were present in 4 patients (16%) for which they were evaluated for cardiac involvement (**Table – 8**).

Out of 25 patients Renal involvement was absent in all patients. Renal involvement was screened in the form of Proteinuria (more than 200 mg/24 hrs) and Hypertension and renal function tests. Acute onset of malignant hypertension followed by rapidly progressive renal insufficiency is termed as Scleroderma renal crisis which is low in Indian patients (**Table – 9**).

Normocytic Normochromic anemia in 16%.ANA in low titer detected in 56%. Rheumatoid Factor was positive 16%.CRP was positive at 40% (**Table – 10**).

Table – 8: Cardiovascular manifestations.

No. of patients	Symptoms	X-Ray chest	ECG	Echo
Patient No.1	Chest pain Dyspnea on exertion	Enlarged cardiac silhovette sign	Low voltage QRS complex and increase of T waves	Moderate pericardial effusion
Patient No.2	Chest pain and palpitation	Normal	Normal	Mitral Valve prolapse disease
Patient No.3	Chest pain Dyspnea on exertion	Enlarged cardiac silhovette sign	Increase of Twaves	Mild to moderate pericardialeffusion
Patient No.4	Dyspnea on exertion	Basal Haziness	Normal	Pulmonary Hypertension secondary to ILD

Table – 9: Renal system.

No. of patients	%	Renal Function Test	24 Hours proteinuria	B.P.
25	0/25	Normal	Normal	Normal

Table – 10: Investigations.

Investigations	No. of patients	%
Hb < 10 gm%	4/25	16%
ESR > 20 mm/hr	17/25	68%
Proteinuria(> 200 mg/24 hrs)	0/25	0%
Rheumatoid Factor	4/25	16%
ANA	9/25	56%
CRP	8/25	40%
Skin Biopsy	23/25	92%

Discussion

The various clinical features of systemic sclerosis of 25 patients have been analyzed in this study. The results are compared with the previous South Indian Study, North Indian Study [12], West Indian Study [14] and Western country study. Out of 25 patients studied there were 2 males and 23 females with a sex ratio of 1: 11.5 with female preponderance. Similar to the universal picture the mean age of onset in this study was 40 years similar to other studies. The peak age of onset occurred in the 3rd and 4th decade in this study this is also comparable with other studies. It was very similar to other studies [7]. Raynaud's phenomenon was the most common presenting symptom in North Indian, West Indian, and Western Country study. But in this study and South Indian study, Raynaud's phenomenon was low incidence. In this study, Raynaud's presentation was 28%. The lower incidence of Raynaud in this study is probably due to the hot climate prevailing in South India. Vascular involvement in the form of vasculitic ulcers is seen in 1 patient [8]. One patient had resorption of terminal changes of fingers in the radiograph of hands probably due to ischemia of the digits. In this study skin (cutaneous lesions) (100%) and arthralgia (80%) were the most common presenting manifestations which are similar to previous South Indian studies and other studies [9]. One of the patients in this study had diffuse skin involvement and internal organ involvement affecting lungs as Interstitial lung disease and pericardial effusion. CREST syndrome was not noticed in any patient. Pigmentary changes were present in as high as 68% and this is comparable with the West Indian study where the incidence of pigmentation was 76.2% and this may be

because of the geographical location of these two zones which are closer to the equator [10]. Sclerodactyly (40%) was seen to occur more commonly in our patients. Skin Biopsy was done for 23 patients of the 21 (84%) had features of Scleroderma, Biopsy showed epidermal skin appendages atrophy and collagen fibers in the reticular dermis appear broad and hyalinized. A loss of space between collagen bundles is noted. Mononuclear cells, mostly T cells form a variable perivascular infiltrate in the deep dermis and subcutis [11]. Joint manifestations were seen in 20 of the 25 patients. Small joints involvement was seen more often (52%) similar to South Indian studies. Major joint involvement is 36.5%. Similar to South Indian study [12]. Gastrointestinal involvement was present in 72%, compared to 40.8% in South Indian, 50.5% in North Indian, 45% in Western Country studies. Dysphagia was the commonest symptom, 12 of 25 patients presented with Dysphagia. 6 of 25 patients presented with Regurgitation. Gastro Intestinal tract involvement was confirmed by Ba swallow and upper gastroduodenal endoscopy. Ba swallow showed Dilatation of the esophagus in 12 cases who presented with Dysphagia. Upper Gastro-Intestinal endoscopy confirmed gastroesophageal reflux disease who presented with regurgitation. Exertional Dyspnea with bibasilar crepitations was present for four patients (16%). Owens reported a 70% incidence of lung involvement [13]. For all 4 patients, a chest X-ray was taken two of them had normal X-ray chest. Another two showed bilateral basal Haziness. For all 4 patients, a pulmonary function test was done. For those who showed normal X ray's had mild to moderate restrictive airway disease [14]. Those who showed bilateral haziness had moderate to severe restrictive airway disease. Computed tomography was done for all 4 patients. CT chest was normal for whom X-ray was normal [15]. CT showed interstitial lung disease for whom X-ray showed bilateral haziness and pulmonary function test showed severe restrictive airway disease [16]. Symptoms attributable to the cardiovascular system were present in 4 patients (16%). Two of them had ECG changes of T wave abnormality. Two of them ECG was normal [17]. Echocardiography showed moderate pericardial effusion for 2 patients and MVP Mitral Valve Prolapse in one patient. The fourth patient developed moderate pulmonary hypertension secondary to Interstitial Lung Disease (ILD) [18, 19, 20].

Conclusion

Female preponderance (Female: Male 11.5: 1). The highest incidence was seen in the 3rd and 4th decade. Limited cutaneous type was commoner than the diffuse SSc. Skin and joint manifestations were predominant. Sclerodactyly and pigmentary changes are more common. Raynaud's phenomenon is very much less probably due to climatic causes. Fully developed CREST was not present in this study. Resorptions of terminal phalanges were less common. Gastrointestinal involvement is more than in other studies. Respiratory involvement was low compared to other studies. Cardiac involvement is less like other studies. Renal involvement is NIL in this study.

References

1. Kasper DL, et al. Harrison's Principles of Internal Medicine, 16th Edition Volume II, McGraw-Hill, 2004.
2. Marc C. Hoch Berg, Alan J Silman. Rheumatology, 3rd Edition, Volume II, Elsevier, 2018, p. 1463.
3. Kelley's Text Book of Rheumatology, 6th edition, Volume II, Elsevier, 2012.
4. Laing TJ, Gillespie BW Toth MB, et al. Racial difference in scleroderma among women in Michigan. Arthritis Rheum, 1997; 40: 734.
5. Mayes M.D. Epidemiology of systemic sclerosis and related disease cum open Rheumatol., 1997; 9: 557.
6. Gilliand BC, Mannik M. Harrison's principles of internal medicine. 10th Edition, New York, McGraw Hill, 1983, p. 2002-6.
7. Malavya AN, Adhar GI, Pasricha V S. Connective tissue disease in India, III clinical and immunological profile of

PSS. J Assoc. Physician India, 2009; 27: 395 – 7.

8. Krishnamoorthy V, Chandrasekaran AN, Porkodi R, Ramakrishnan S, Rajendran, CP, Madhavan R. Achudhan K, Parthiban M, PSS in South India J.Assoc. Physicians India, 1991; 39: 254 – 57.
9. Ashokkumar, Malavya AN, Tiwari SC, Singh RR, Abbay Kumar Pande JN. Clinical and laboratory profile of Systemic sclerosis Northern Indian. J. Assoc. Physicians India, 1990; 38: 765-68.
10. Gupta HL, Balakrishna. Clinical profile of PSS in Women. J Indian Med. Assoc. (JIMA), 1987; 85(2): 38-40.
11. Yajna Desai, Ghanekar MA, Singnera RD, Joshi VR. Renal involvement in Scleroderma. J Assoc Physicians India, 1990; 38: 768-70.
12. Frank KH, Fussel M Conrad K, et al. Differential distribution of microsatellite in anti-topoisomerase 1 responder among Scleroderma patients with and without exposure to quartz/metal dust. Arthritis Rheum., 1998; 41: 1306.
13. Whiteside TI, Medsger TA, Roadnaw GP. Studies of HIA antigens in PSS. In Black CM Myeres AR) eds. Systemic sclerosis, 1st Edition, New York Gower Medical, 1985, p. 89 – 96.
14. Lyrics C.J., Singh G, Whiteside T.L., et al. Histocompatibility antigens in PSS. J Clin Immunol., 1982; 2: 314.
15. Briggs D, Stephens C, Vaughan R, et al. A molecular and semilogic analysis of the major histocompatibility compliancy and complement component C4 in systemic sclerosis. Arthritis Rheum., 1993; 36: 943.
16. Rothfield NF, Roadnan GP. Antinuclear Antibodies in PSS. Arthritis Rheum., 1968; 11: 607.
17. Haustein UF, Herrmann K. Environmental Scleroderma. Clin Dermatol., 1994; 12: 467-473.
18. Haustein UF, Ziegler V. Environmentally induced systemic sclerosis-like disorders. Int. J.Dermatol., 1985; 24: 147-51.
19. Haustein UF, Ziegler V, Herrmann K, Pseudosklerodermien. Hautnah "Dermatologie", 1991; 1: 67-71.
20. Malaviya AN, Adha GC, Pasricha JS. Connective tissue diseases in India III. Clinical and immunological profile of progressive systemic sclerosis. J Assoc Physicians India, 1979; 28: 395-400.