IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 15, Issue 1 Ver. II (Jan. 2016), PP 24-28
www.iosrjournals.org

Profile of Systemic sclerosis in Tertiary care centre, Vijayawada.

Dr.G.Purnima[1], Dr.V.GangaBhavani[1], DrT.S.P Kumar[1],
Dr.S.Uma Sankar[1]
[1] *Department of Dermatology Venereology Leprosy, Siddhartha medical college, Vijayawada, India*

Abstract:
Background & Objectives: *Systemic sclerosis(SSc)is a multisystem connective tissue disorder of uncertain etiology characterized by thickening of skin and involvement of internal organs . The aim of the study was to find the relative frequencies, characteristics of Cutaneous ,Systemic features and Laboratory profile of SSc in patients attending DVL OPD at tertiary care centre,Vijayawada.*
Methodology: *This was a cross sectional, clinical observational study. Consecutive patients of SSc attending DVL OPD of tertiary care centre Vijayawada between July 2013 – 2015 were enrolled in the present study.*
Results: *A total of 20 patients (all females; mean age 36.15±12.71 years) were evaluated. Among the cutaneous manifestations, common presenting features were skin sclerosis (90%), pigmentary abnormalities (80%), others included Raynaud's phenomenon (60%), sclerodactyly(60%) , microstomia (60%), digital pitted scars (55%).Common systemic features seen were shortness of breath (55%), GERD symptoms(45%), dysphagia (30%).Lab abnormalities noted were raised ESR (80%),ANA positive(85%), anti–Scl 70(35%),abnormalities in CXR-PA (40%), HRCTchanges(65%) ,PFT abnormalities(60%), Upper GI endoscopy abnormalities (80%) .*
Conclusion: *Skin sclerosis and pigmentary abnormalities were more frequent findings , HRCT & PFT changes and Upper GI endoscopy findings were seen in majority of patients making them mandatory in all patients of SSc.*
Key words: *Anti nuclear antibody, cutaneous features, systemic features, Systemic sclerosis*

I. Introduction

Systemic sclerosis (SSc) is a multi-system connective tissue disorder characterized by thickening of the skin and involvement of internal organs like kidney, lung, gastrointestinal system and heart. It is characterized by damage to the endothelium of the small vessels resulting in tissue ischemia, activation of the immune system and fibrosis.[1]

The degree and extent of the cutaneous and internal organ involvement are variable. With the advances in therapies particularly renal transplantation, pulmonary involvement has taken over as the leading cause of morbidity and mortality in SSc patients.[2] Two thirds of patients suffering from SSc present with pulmonary disease with effort dyspnoea being the most usual respiratory symptom.[3] Pulmonary artery hypertension and interstitial lung disease (ILD) are the two main syndromes associated with SSc.[4]

Disease of the gastrointestinal tract (GIT) occurs in approximately 90% of patients with SSc and has a major impact of life. Every part of the GIT can be involved in SSc, including the mouth (Xerostomia), Oesophagus(dysmotility,acid reflux),stomach(vascular ectasia, gastroparesis), intestines(vascular lesions, hypomotility, bacterial overgrowth, intestinal pseudoobstruction) and anorectal system (faecal incontinence)[.5,6]

II. Patients & Methods

This was a cross sectional and clinical observational study.Consecutive patients of Systemic sclerosis attending DVL OPD of tertiary care hospital of Vijayawada between July 2013 – 2015 were enrolled in the present study.Informed consent was obtained from all the patients.

Routine laboratory investigations including complete hemogram, ESR, C-reactive protein,CXR-PA view and biochemical profile were done. Special laboratory tests like estimation of anti nuclear antibody, anti – scl 70 , HRCT,Pulmonary function tests, upper gastrointestinal (UGI) endoscopy were done in all cases.Lesional histopathology and other relevant investigations were done whenever needed.

III. Observations And Results

A total of 20 patients (all females) were evaluated. The age of the patients was between 14 and 65 yrs with mean age of 36.15±12.71 years. Maximum number of SSc cases were reported in between 20 -40yrs age group. The clinical and laboratory profiles were summarized in Table no.1 and 2 respectively.

Common cutaneous symptoms with which patients presented were tightness of skin in 18(90%), Pigmentary changes in 17(85%), Raynaud's phenomenon in 10(50%), Diffuse alopecia in 8(40%), Generalized pruritus in 5(25%). Common cutaneous signs observed in SSc were Skin sclerosis in 18(90%), Salt pepper pigmentation in 17(85%), Diffuse hyperpigmentation in 11(55%), Mask like face 15(75%) [Figures 1and 2], Sclerodactyly in 12(60%), Digital pitted ulcers/scars in 12 (60%), Raynaud's phenomenon in 12(60%), Microstomia in 12(60%) [Figures 3 and 4], Diffuse alopecia in 8(40%), Nail changes in 7(35%), Hands and Feet swelling in 5(25%), Flexion contracture of digits in 5(25%), Amputation of digits in 3(15%), Digital gangrene in 2(10%), Mat like telangiectasis in 2(10%), Calcinosis cutis in one (5%) patients.

Shortness of breath was complained by 11(55%) and dry cough in 6(30%) patients. On examination crepetations were observed in 6(30%), decreased chest expansion in 7(35%) patients. CXR-PA abnormalities were detected in 8 (40%), abnormal pulmonary function tests in 12(60%) patients. HRCT abnormalities were seen in 11 (100%) symptomatic patients and 2(10%) asymptomatic patients..

GERD symptoms like, epigastric pain, belchings, regurgitation were seen in 9(45%) patients. Dysphagia was seen in 6(30%) patients. Among them upper GI endoscopy abnormalities were seen in 14(93.3%) patients. Upper GI abnormalities were seen in 2(40%) out of 5 asymptomatic patients. The abnormalities seen were Lax Lower esophageal sphincter (LES) in 10(50%) patients, lower 1/3rd esophagitis in 8(40%) patients, Fundal gastritis and prolapsed gastropathy in 5(25%) patients, Hiatus hernia in 4(20%) patients, Esophageal web in one (5%) patient.

Laboratory abnormalities seen were elevated ESR in 16(80%), Proteinuria in 2 (10%) patients. The antinuclear antibody (ANA) was positive in 17(85%) patients. Speckled pattern was the most common in 10 patients followed by homogenous pattern in 4 patients, Nucleolar pattern in 3 patients. Anti Scl 70 antibody was positive in 7 (35%) patients.(Table No.3).

IV. Tables And Figures

Table no.1 clinical profile of Systemic sclerosis

S.No.	Clinical features	No. of patients (n=20)	Percentage (%)
1	Skin sclerosis	18	90%
2	Mask like face	15	75%
2	Salt pepper pigmentation	17	85%
3	Diffuse hyperpigmentation	11	55%
4	Sclerodactyly	12	60%
5	Digital pitted ulcers/scars	12	60%
6	Raynaud's phenomenon	12	60%
7	Microstomia	12	60%
8	Hands & Feet swelling	7	35%
9	Flexion contracture of digits	5	25%
10	Amputation of digits	3	15%
11	Digital gangrene	2	10%
12	Mat like telangictasia	2	10%
13	Nail changes	7	35%
14	Pruritus	5	25%
15	Diffuse alopecia	8	40%
16	Calcinosis cutis	1	5%
17	Dyspnoea	11	55%
18	Dysphagia	6	30%
19	GERD symptoms	9	45%

Tab no .2 Laboratory profile of Systemic sclerosis

S.No	Lab parameter	No of patients	%
1	ESR elevation	16	80%
2	Proteinuria	2	10%
3	CXR-PA abnormalities	8	40%
4	HRCT chest	13	65%
5	Abnormal PFT	12	60%
6	UGI Endoscopy	16	80%
7	ANA	17	85%

Tab no.3 ANA Profile of Systemic sclerosis

S.no	Antibodies	No. of patients	%
1	ANA		
	Total positivity	17	85%
	Speckled pattern	10	58.80%
	Homogenous pattern	4	23.50%
	Nucleolar pattern	2	11.76%
	Speckled + Nucleolar	1	5.88%
2	Anti SCL 70	7	35%

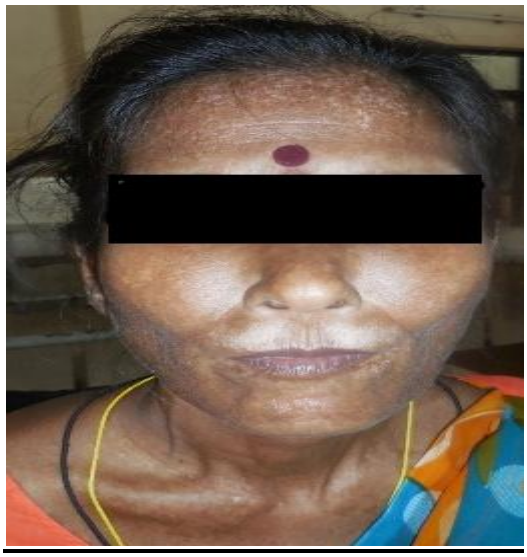

Fig no.1 mask like face and radial furrowing around mouth

Fig no. 2 salt pepper pigmentation

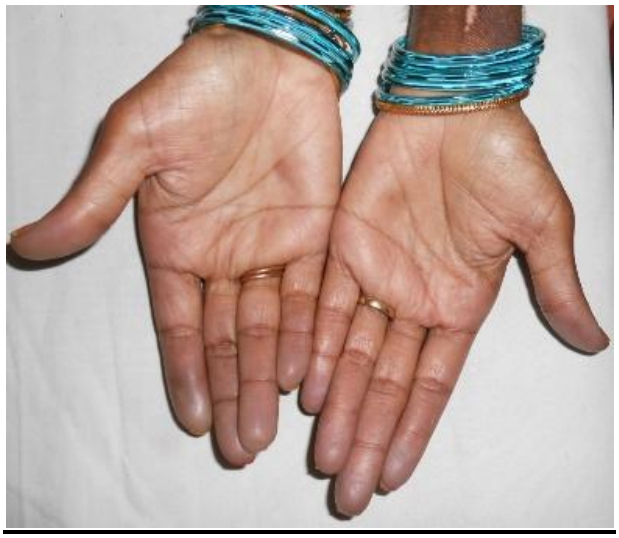

Fig no.3 Raynaud's phenomenon

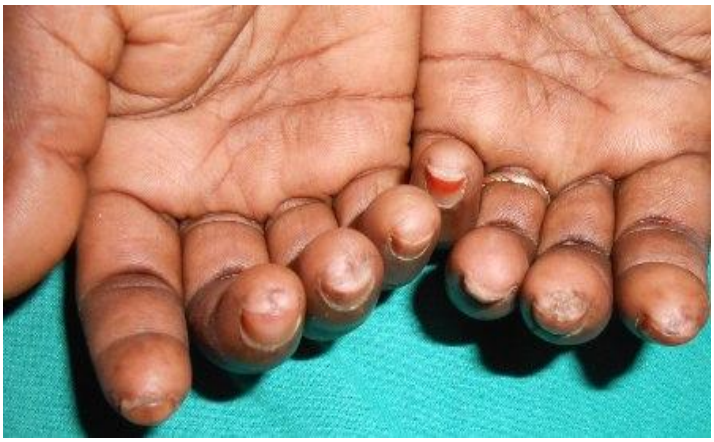

Fig no.4 digital pitted scars

V. Discussion

In the present study all the patients of systemic sclerosis were females(100%). In a study by Sharma VK et al[7], there was female preponderance , the male to female ratio being 1 : 5.2.Ghosh et al[8] in his study found male to female ratio of 1 : 8.2. In a recent study from Saraswak[9] and in another study on Afro-Caribbean population [10] 96% patients were females. The mean age in the present study was 36.15 ±12.71 years, which was comparable to studies done by Sharma VK et al and Ghosh et al who reported mean age of 37.25yrs and 29.6 yrs respectively.[7,8]

The skin is almost universally involved in SSc. In the present study cutaneous sclerosis was found in 90% of patients. More or less similar figures have been reported by Indian studies (98.5%, 100%)[7,11], studies from Iraq (96.5%)[12], and saraswak (100%).[9] In African American study skin sclerosis was found in 62% patients.[13] Raynaud's phenomenon was present in 60% of patients in the present study. More or less similar frequency of Raynaud's phenomenon was noted in a north Indian (92.9%),[7] African American study (96%),[13] Afro- Caribbean population (93%),[10] and a study from Iraq (100%)[12]. However a much lower frequency has been reported in a south Indian study (28%).[11] These discrepancies were probably due to the ethnic variation of the study population and climatic differences. Finger tip ulcerations and scarring was observed in 60% of patients in the present study. This was comparable to African American study (82%).[13] The range of frequencies of finger tip ulcerations was 52%-70% in previous studies.[7,8,9,10,12] The most common pigmentary disturbance in present study was mottled pigmentation (85%) followed by diffuse hyperpigmentation (55%). This is in contrast to study by Sharma VK et al[7] **,** who reported diffuse hyperpigmentation (88.1%) most commonly followed by mottled pigmentation (51.2%).Microstomia was observed in 60% of patients in the present study which was higher than that reported by Sharma VK et al[7] (55.5%) and lower than that reported by Ghosh et al[8] (82.6%).Sclerodactyly was reported in 60% of patients in the present study. Ghosh et al[8] found a higher incidence (82.6%) in their study. Calcinosis cutis was observed in one patient (5%) in our study. Ghosh et al[8] observed calcinosis cutis in one patient (2.2%) in their study. Digital gangrene was seen in 10 % of

patients of the present study. Ghosh et al and Sharma VK et al reported an incidence 4.3% and 6.7% of digital gangrene in their studies respectively.[8,7] A lower frequency of Telangiectasia (10%) was seen in the present study in contrast to other studies by Sharma VK et al (36.8%)[7],Ghosh et al (23.1%)[8], Afro- Caribbean study (48%).[10] Nail changes were seen in 35% of the patients in the present study, which was lower than that reported by Ghosh et al (28.3%).[8] Pruritus was observed in 25% of patients in the present study. Ghosh et al observed generalized pruritus in 8.7% of patients in their study.[8]

In the present study pulmonary involvement was seen in 13 out of 20 (65%) patients on HRCT, 8 out of 20 (40%) on CXR PA, in contrast Hassan et al found pulmonary involvement in 80% (20/25) of cases on HRCT as against 16% (4/25) on chest X ray.[14] GERD symptoms(45%) and dysphagia(30%) were the commonest symptoms observed in our study. UGI Endoscopy abnormalities were detected in 16 (80%) with esophagus being the commonest site affected, this was comparable to the study by M.B Adarsh et al.[15]

Among laboratory abnormalities ESR was elevated in 80% patients, this was comparable to the study by Sharma VK et al (87.8%).[7] Renal dysfunction in the form of proteinuria was seen in 2 (10%).Sharma VK et al observed proteinuria in 6% of their patients. Renal involvement was reported in 23-40% in western countries in contrast to our study,which are similar to that of Iraq and Thailand.[12,16] ANA test was positive in 11 out of 12 patients (91.66%) in the present study which was higher than that reported by Ghosh et al (78.2%)[8] and was comparable to that reported by Sharma VK et al (89.1%).[7] Anti- Scl 70 was positive in 7 out of the 20 (35%) patients.

VI. Conclusion

Skin sclerosis and pigmentary abnormalities were more frequent presenting features of Systemic sclerosis in our study. HRCT , PFT changes and Upper GI endoscopy findings were seen in majority of patients irrespective of symptoms making them mandatory in all patients of SSc, for early intervention and better outcome.

References

[1] Black CM, Denton CP. Scleroderma and related disorders in adults .In: Maddison PJ, Isenberg DA, Woo P, Glass DN, editors. Oxford Textbook of Rheumatology. 2nd ed. (Oxford Medical Publication; 1998).p.1217-47.

[2] Wells AU, Steen V, Valentini G. Pulmonary complications: One of the most challenging complications of systemic sclerosis.Rheumatology (Oxford 2009);48 Suppl 3: iii40-4.

[3] Ferri C, Valentini G, Cozzi F, Sebastiani M, Michelassi C,La Montagna G, et al. Systemic sclerosis: Demographic,clinical, and serologic features and survival in 1,012 Italianpatients. Medicine (Baltimore) 2002;8:139-53.

[4] Steen V. Predictors of end stage lung disease in systemic sclerosis. Ann Rheum Dis 2003;62:97-9

[5] Sjogren RW. Gastrointestinal features of scleroderma. Curr Opin Rheumatol 1996; 8(6): 569–75.

[6] Sallam H, McNearney TA, Chen JD. Systematic review: pathophysiology and management of gastrointestinal dysmotility in systemic sclerosis (scleroderma). Aliment Pharmacol Ther 2006; 23(6): 691–712.

[7] Sharma VK, Trilokraj T, Khaitan BK, Krishna SM. Profile of systemic sclerosis in a tertiary care center in North India, Indian J Dermatol Venereol Leprol 2006;72:416-20.

[8] Ghosh SK, Bandyopadhyay D, Saha I, Barua JK. Mucocutaneous and demographic features of systemic sclerosis: A profile of 46 patients from Eastern India.Indian J Dermatol 2012;57:201-5.

[9] Teh CL, Kuan YC, Wong JS. Systemic sclerosis in Sarawak: A profile of patients treated in the Sarawak General Hospital. Rheumatol Int 2009;29: 1243-5.

[10] Flower C, Nwankwo C. Systemic sclerosis in an Afro- Caribbean population : A review of demographic and clinical features. West Indian Med J 2008;57:118.

[11] Krishnamurthy V, Porkodi R, Ramakrishnan S, Rajendran CP, Madhavan R, Achuthan K, et al. Progressive systemic sclerosis in south India. J Assoc Physicians India 1991;39:254-7.

[12] Al – Adhadh RN, Al- Sayed TA. Clinical features of systemic sclerosis. Saudi Med J 2001;22:333-6.

[13] Reveille JD, Fischbach M, McNearney T, Friedman AW, Aguilar MB, Lisse J et al . Systemic sclerosis in 3 US ethnic groups: A comparison of clinical, sociodemographic, serologic and immunogenetic determinants. Semin Arthritis Rheum 2001;30:332-46.

[14] Hassan I, Nisa N, Hamid M. Pulmonary involvement in systemic sclerosis: An imaging study from Kashmir. Indian J Dermatol 2015;60:102.

[15] M.B Adarsh,Shefali Khanna Sharma, Varun Dhir, Aman Sharma Rakesh Kochhar, Saroj K Sinha, Anish Bhattacharya, Satyavati Rana, Surjit Singh. Gastrointestinal manifestations of Systemic sclerosis –clinical and investigative study of 50 patients. Indian J of Rheumatology 2014: 9:supplement 1:S52.

[16] Ruangjutipopan S, Kasitanon N, Louthrenoo W, Sukitawut W, Wichainun R. Causes of death and poor survival prognostic factors in Thai patients with systemic sclerosis. J Med Assoc Thai 2002;85:1204-9.