

Prevalence of cutaneous findings in systemic sclerosis patients - Experience of a teaching hospital

Prevalência de achados cutâneos em portadores de esclerose sistêmica - Experiência de um hospital universitário*

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Abstract: *BACKGROUND - Systemic sclerosis or scleroderma is a rare collagen disease presenting several cutaneous manifestations.*

OBJECTIVE - To study the prevalence of cutaneous manifestations in systemic sclerosis and its subtypes (limited form, diffuse form and overlap syndrome).

METHODS - We studied 32 patients with scleroderma (20 with the limited form; 8 with the diffuse form and 4 with overlap syndrome) considering skin sclerosis, Raynaud's phenomenon, digital scars, telangiectasia, leucomelanoderma (pigmentary changes), microstomy, calcinosis and pruritus.

RESULTS - We found skin sclerosis and Raynaud's phenomenon in 100% of patients, digital scars in 65.6%, telangiectasia in 43.7%, pigmentary changes in 43.7%, microstomy in 31.25%, pruritus in 28.1% and calcinosis in 12.5%. No statistically significant differences were observed in the limited and diffuse forms of scleroderma ($p=1.0$ for digital scars; $p=0.69$ for teleangiectasis; $p=0.22$ for microstomy; $p=1.0$ for calcinosis and pruritus). The overlap syndrome was not different from the isolated forms (limited and diffuse) of systemic sclerosis.

CONCLUSION - The most common skin manifestations in scleroderma are skin sclerosis and Raynaud's phenomenon and calcinosis is the rarest. The other cutaneous manifestations presented similar frequency in all subtypes of scleroderma.

Keywords: *Mixed connective tissue disease; Scleroderma, diffuse; Scleroderma, limited; Scleroderma, systemic*

Resumo: **FUNDAMENTOS** - A esclerose sistêmica é colagenose pouco comum e muito rica em manifestações cutâneas.

OBJETIVO - Estudar a prevalência das manifestações cutâneas na esclerose sistêmica em geral e nos seus diferentes subtipos (formas limitada, generalizada e mista).

MÉTODOS - Analisaram-se 32 pacientes de esclerose sistêmica (20 com forma limitada, oito com generalizada e quatro com forma mista) quanto à esclerose de pele, fenômeno de Raynaud, cicatrizes estelares, telangiectasias, leucomelanoderma, microstomia, calcinose e prurido.

RESULTADOS - Encontraram-se esclerose de pele e fenômeno de Raynaud em 100% dos pacientes; cicatrizes estelares em 65,6%; telangiectasias em 43,7%; leucomelanoderma em 43,7%; microstomia em 31,25%; prurido em 28,1% e calcinose em 12,5%. Não se observaram diferenças entre as formas localizada e difusa da doença, sendo $p=1$ para cicatrizes estelares; $p=0,69$ para telangiectasias; $p=0,22$ para microstomia, $p=1$ para calcinose e prurido. A forma mista de doença não diferiu das formas isoladas (limitada e difusa) quanto aos mesmos achados.

CONCLUSÕES - As manifestações mais comuns na esclerose sistêmica são a esclerose de pele e o fenômeno de Raynaud, e a mais rara é a calcinose. As três formas apresentam frequências semelhantes de Raynaud, cicatrizes estelares, microstomia, telangiectasia, calcinose e prurido.

Palavras-chave: Doença mista do tecido conjuntivo; Escleroderma difusa; Escleroderma limitada; Escleroderma sistêmico

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INTRODUCTION

Systemic scleroderma is a collagen disease that prevails in women and has a large number of cutaneous findings that should be well recognized by clinicians that manage this condition.¹ It may present as an exclusively cutaneous (limited form) or systemic (SS) disease, which may be divided into limited form, when the skin thickens only in the extremities (face and "gloves and boots" type distribution), and generalized or diffuse form, when it affects the whole body.^{1,2} Exclusively visceral involvement (scleroderma *sine* scleroderma) is extremely rare.^{3,4} Sometimes SS may present with other connective tissue conditions, and is then called overlapping or mixed syndrome.¹

In the systemic forms of the disease, skin sclerosis, which initially affects hands, may begin with an edematous aspect. In the well-established disease, it tends to have induration and, after some time, it may or may not develop into an atrophic form.¹

These infiltrative changes are associated with pigmentary changes, pruritus, telangiectasia, calcinosis and ulcerative and scarring phenomena secondary to ischemia due to Raynaud's syndrome. Microstomy is a finding secondary to cutaneous face infiltration and causes perioral folds, contributing to the typical aspect of SS patients.^{1,2}

The present study was performed with the objective of studying the prevalence of skin manifestations in the local population with SS, and of comparing findings between the limited, diffuse and overlapping forms.

PATIENTS AND METHODS

We studied all patients with SS (limited and diffuse forms, and overlapping syndrome with a scleroderma component) seen at the Rheumatology Outpatient Clinic of Hospital Universitário Evangélico de Curitiba during the year of 2003. The sample comprised 32 patients, 20 of which with the limited form, eight with the diffuse form and four with overlapping syndrome.

After signing the informed consent form, patients answered a questionnaire and were submitted to a physical examination regarding the most common cutaneous manifestations of the disease. The manifestations registered were: presence of Raynaud's phenomenon (Figure 1), history of digital pulp ulcerations and current or past itching crises. The physical examination aimed to show digital scars, telangiectasia (Figure 2), pigmentary changes (Figure 3), calcinosis (Figure 4) and microstomy (Figure 5). All patients whose oral opening was less than three middle fingers of the right hand were considered as having microstomy. Calcinosis was radiologically con-



FIGURE 1: Raynaud's phenomenon. The narrowing of the digital pulps and hypotrophy is suggestive of long-term Raynaud's

firmed for all subjects.

Moreover, all patients were submitted to modified Rodnan skin score (MRSS), which consists of the sum total of scoring 17 anatomical sites, graded as 0 = normal skin; 1 = mild thickening (skin is thickened, but can still be pinched); 2 = moderate thickening (skin is thickened and cannot be pinched, but is not completely adhered to deep planes, allows light sliding); 3 = intense thickening (very thickened skin, cannot be pinched, adhered to deep planes, does not allow sliding).⁵

Only the individuals with complaints of intense itching followed by fast progression of skin thickening, which is attributed to mastocyte degranulation, were considered as patients with pruritus.⁶⁻⁸

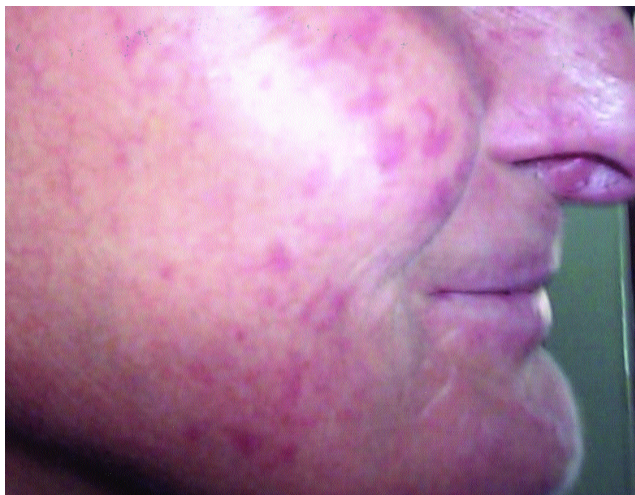


FIGURE 2: Multiple telangiectasia in face in the limited form of systemic sclerosis

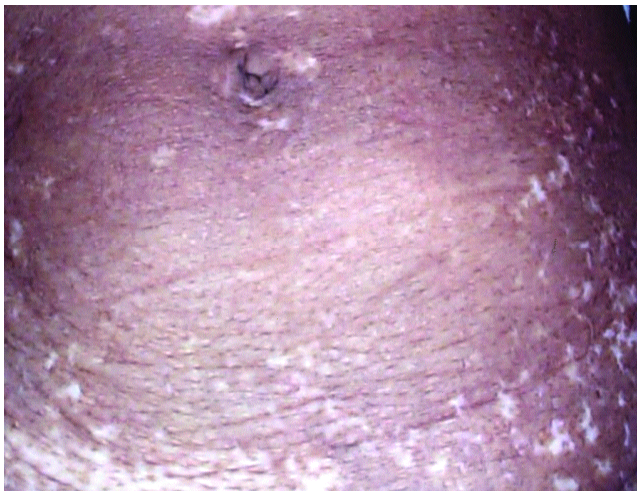


FIGURE 3: Pigmentary changes in a patient with the diffused form of the disease

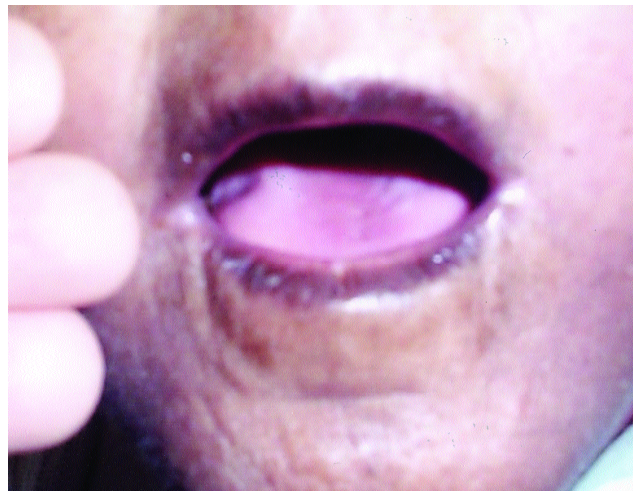


FIGURE 5: Microstomy

Data were submitted to statistical analysis using the Prism (Graphpad, version 2.0) software; descriptive studies and the Fisher test were used for the comparative analysis between samples with discrete data, and the Kruskal-Wallis test was applied to continuous data. The level of significance adopted was 5%.

RESULTS

Out of 32 patients studied, there were 31 women, and one man. Age ranged from 22 to 67 years, with a mean of 47.11 ± 11.99 years. The demographic features of the three subgroups studied are in table 1.

All patients had Raynaud's phenomenon and its onset ranged from zero to 13 years prior to diag-

nosis of SS. In the limited form, the mean time for developing Raynaud's before diagnosis was 2.1 ± 3.44 years; in the diffuse form, 0.62 ± 1.40 years, and in the overlap syndrome, 3.2 ± 2.50 years ($p=0.152$).

Involvement of hands was present in all patients as induration, except for one patient who had the edematous form.

As to other cutaneous findings, their prevalence in the scleroderma population as a whole was: digital scars in 21 patients (65.6%), telangiectasia in 14 (43.7%), pigmentary changes in 14 (43.7%), microstomy in 10 (31.25%), pruritus in nine (28.1%) and calcinosis in four (12.5%). These data are presented in graph 1.

The results of these cutaneous findings according to the form of systemic sclerosis, in addition to frequency comparison in the different groups, are summarized in tables 2 and 3.

The modified Rodnan skin score in the three forms represented a mean of 16.10 ± 6.56 in the limited form; 24.0 ± 5.97 in the diffuse form and 18.0 ± 3.16 in the overlapping syndrome, $p=0.030$. The comparison of this variable in the different SS subsets showed a $p=0.02$ between the diffuse and limited forms; $p=0.279$ between the diffuse and overlapping forms and $p=0.59$ between the overlapping and limited forms.

DISCUSSION

SS is a relatively rare disease, but stands out for the variety of cutaneous findings that assist in making diagnosis.

In the population studied, patients with diffuse SS had a shorter period of disease than those

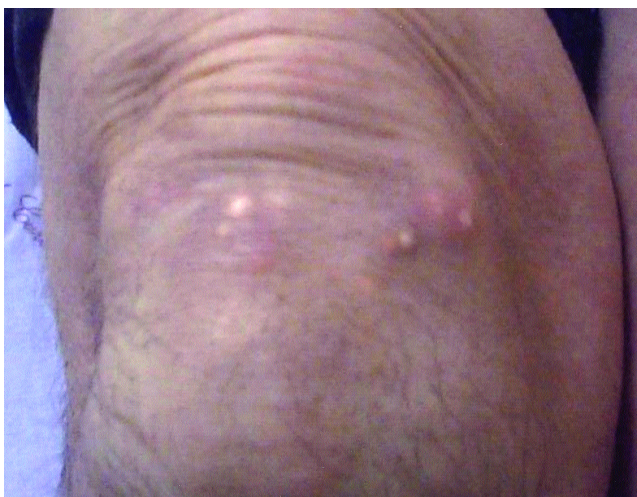


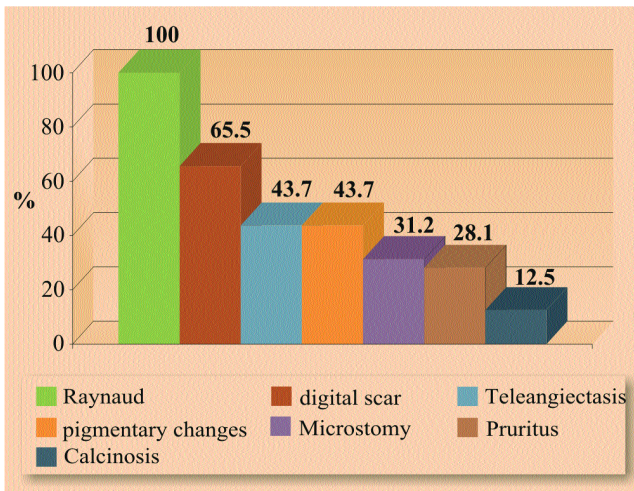
FIGURE 4: Calcinosis in the knee of a patient with the limited form of systemic sclerosis

TABLE 1: Demographic features of the population studied

	Limited form (n=20)	Diffuse form (n=08)	Overlapping form (n=04)	p
Mean age of patients (years)	47.05±12.73	47.38±9.08	46.57±6.85	p=0.953
Disease Period (years)	12.45±8.75	4.12±1.95	14.25±13.96	p=0.013

n= sample

GRAPH 1: Prevalence of skin changes in the scleroderma population studied



with other forms, a fact that can be easily understood, given that in this form the higher aggressiveness of the disease makes patients seek doctors sooner.

There was skin sclerosis in all patients and it was more severe in the diffuse form than in the limited one, which is translated as a higher modified

Rodnan skin score. As a matter of fact, this component relates to poorer prognosis regarding patient survival.⁹

Raynaud’s phenomenon was present in all patients; this is a very important finding since it shows that its absence makes the diagnosis of SS less likely. The high frequency of Raynaud’s phenomenon in SS is acknowledged in the literature, with a prevalence ranging between 90 and 99%,^{1,10} moreover, it is the initial complaint of 70% of patients.¹¹ According to Seibold, its absence is more frequently observed in male patients,¹² a fact that cannot be appreciated in this sample composed of only one man.

The interval between the onset of Raynaud’s phenomenon and diagnosis of SS was shorter in patients with the diffuse form, although the data did not show statistical significance.

A study performed with 63 patients showed a prevalence of 46% of digital scars.¹⁰ In the sample studied, the high prevalence of digital scars (65.6%) strengthens the importance and severity of Raynaud’s phenomenon in these patients, given these scars result from micro infarctions in finger pulps.¹¹

All other cutaneous manifestations studied, such as pigmentary changes, telangiectasia, micros-

TABLE 2: Cutaneous findings in the limited and diffuse forms of systemic sclerosis

	Limited systemic sclerosis (n=20)	Diffuse systemic sclerosis (n=08)	p
Digital scars	n= 13 (65%)	n= 05 (62.5%)	1 (NS)
Telangiectasia	n= 08 (40%)	n= 04 (50%)	0.69 (NS)
Calcinosis	n= 03 (21.5%)	n= 01 (12.5%)	1 (NS)
Pigmentary changes	n= 11 (55%)	n= 02 (25%)	0,22 (NS)
Pruritus	n= 06 (30%)	n= 02 (25%)	1 (NS)
Microstomy	n= 08 (40%)	n= 02 (25%)	0.66 (NS)

n= sample

NS= not significant

TABLE 3: Cutaneous findings in isolated forms of systemic sclerosis (diffuse and limited)

	Isolated form (n=28)	Overlapping syndrome (n=04)	p
Digital scars	n= 18 (64.2%)	n= 03 (75%)	0.60 (NS)
Telangiectasia	n= 12 (42.8%)	n= 2 (50%)	1 (NS)
Calcinosis	n= 04 (14.2%)	n= 0	1 (NS)
Pigmentary changes	n= 13 (46.4%)	n= 2 (50%)	1 (NS)
Pruritus	n= 08 (28.5%)	n= 01 (25%)	1 (NS)
Microstomy	n= 10 (35.7%)	n= 0	0.28 (NS)

n= sample

NS= not significant

tomy, pruritus and calcinosis, were similarly observed in the different types of disease in the population studied, and the first two followed Raynaud´s phenomenon and digital scars in prevalence. Akenson et al. had already observed similarity in the distribution of calcinosis, Raynaud´s phenomenon and telangiectasia in the diffuse and limited forms of SS.¹³

Tager et al. showed a lower prevalence of telangiectasia (33%) and higher prevalence of calcinosis (19%) in their study, than those found in the present study.¹⁰ This may be explained by the ethnic and/or environmental differences of the populations analyzed, given that the authors only studied Black

individuals in Southern Africa with a high rate of exposure to silica.

CONCLUSION

In conclusion, it could be emphasized that the most common cutaneous manifestations of SS in the present population, in addition to skin sclerosis itself, was Raynaud´s phenomenon and digital scars. These findings should lead to inclusion of SS in the differential etiological diagnosis. Identifying associated cutaneous findings may help making presumptive diagnoses, even if they do not help distinguishing among the three subtypes. □

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