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Clinical Utility of Telangiectasia of Hands in Scleroderma and Other Rheumatic Disorders

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SUMMARY **Telangiectasia of the hands were observed in 76% of patients with scleroderma (n = 53) as compared with 12% of patients with other rheumatic disorders (n = 100) and in 13% of healthy subjects (n = 30).**
In scleroderma, telangiectasia of the hands were commonly multiple (mean number ± SD = 22.9 ± 30.1) with 7.3% being >1 mm in size. They were found in greater numbers in those patients with the limited subtype. The distribution of telangiectasia was observed on all but 4 of 158 sectors of the hand with significant higher numbers on the ventral surface of the digits. Significant associations of telangiectasia of the hands were also observed with numbers of telangiectasia on face and lips (p = 0.001), disease duration (p = 0.002), surface area of digital calcinosis (p = 0.03) and with the presence of the centromere antibody (p = 0.005). Possible associations were observed with prior gastrointestinal bleeds (particularly with telangiectasia >1 mm) and with isolated pulmonary hypertension. No significant correlation was found between number of telangiectasia and with nailfold capillary size or density.
In conclusion, multiple telangiectasia of the hands were most commonly observed in patients with the centromere positive, limited subtype of scleroderma of long duration. Their pathogenesis is unknown.

Telangiectasia are simply defined as macroscopically visible dilated capillaries or venules occurring in the skin. They can be classified as essential telangiectasia occurring in such disorders as Hereditary Haemorrhagic Telangiectasia and Ataxia Telangiectasia or acquired telangiectasia following UV or irradiation exposure to the skin, post topical steroids or in selective rheumatic disorders such as scleroderma or related conditions.[1] Previous studies have emphasized the occurrence of telangiectasia of the hands and face, particularly in the CREST variant of the limited form of scleroderma[2] but there is limited information concerning other clinical associations or telangiectasia occurring in other rheumatic disorders.

In the current study our aim has been to determine the number, size, site and clinical and serological association of telangiectasia of the hands in patients with scleroderma and other rheumatic disorders (and in healthy controls). A further aim was to correlate in scleroderma the number of telangiectasia of the hands with nailfold capillary density and diameter as assessed by quantitative video image analysis technique. This clinical study of telangiectasia of the hands in scleroderma and other rheumatic disorders follows our previous studies investigating the pathogenesis of telangiectasia in scleroderma.[3]

PATIENTS AND METHODS

Patients and controls

Fifty three patients with scleroderma were selected from the South Australian Scleroderma

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Register.[4] Thirty eight of these had limited scleroderma (M:F = 4:34, mean age 64 years [range 36-86]) and 12 had diffuse scleroderma (M:F = 4:8, mean age 50 years [range 37-78]) and the remaining three had overlap scleroderma (M:F = 0:3, mean age 51 years [range 33-74]).

One hundred patients with other rheumatic disorders were obtained from the rheumatology consulting clinics at Flinders Medical Centre (M:F = 22:78, mean age 58 years [range 18-88]). Thirty of these patients had rheumatoid arthritis, 15 osteoarthritis, 13 SLE/Sjögren's and the remainder a variety of inflammatory and soft tissue disorders.

Thirty healthy subjects (M:F = 8:22, mean age 44 years [range 30-55]) were obtained from medical and laboratory personnel at our hospital.

The skin of the hands and face of all patients and controls was examined under standard lighting conditions at room temperature and the number, size (<1 mm and >1 mm) and distribution of all telangiectasia accurately mapped. Other clinical and serological information was obtained from the case notes.

Surface area of hand calcinosis

The surface area of calcinosis in the hands was determined in 15 patients from hand radiographs as previously described.[5] These values were then correlated with the number of hand telangiectasia.

Nailfold capillaroscopy and video image analysis.

Nailfold capillary mean density and diameter was quantitated by video image analysis in 12 scleroderma patients as previously described[6]. These values were then correlated with number of hand telangiectasia.

Statistical analysis.

All data represented as mean ± standard deviation or 95% confidence intervals. Statistical analysis was performed using the Chi-square or the Wilcoxon Rank Sum test. Differences were considered significant when p values were less than 0.05. Correlation co-efficients between two variables were assessed using linear regression.

RESULTS

Telangiectasia of the hands were observed in 76% of patients with scleroderma compared with 12% of patients with other rheumatic disorders and in 13% of the healthy subjects. In scleroderma the telangiectasia tended to be multiple with 7.3% being >1 mm in size whilst in the other 2 categories the telangiectasia were few in number and in all instances <1 mm in size. In patients with other rheumatic disorders, telangiectasia were noted in 3 SLE patients, 2 patients with dermatomyositis and in single patients with rheumatoid arthritis, polymyalgia rheumatica, arthralgia,

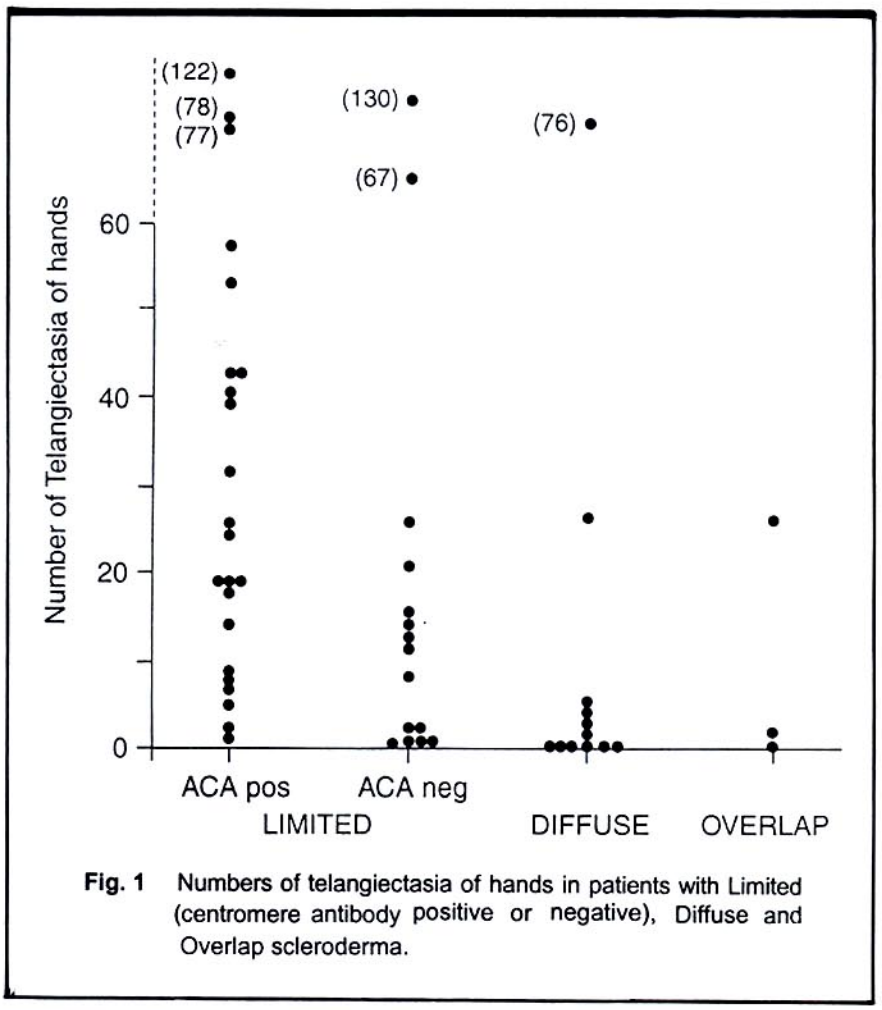

Fig. 1 Numbers of telangiectasia of hands in patients with Limited (centromere antibody positive or negative), Diffuse and Overlap scleroderma.

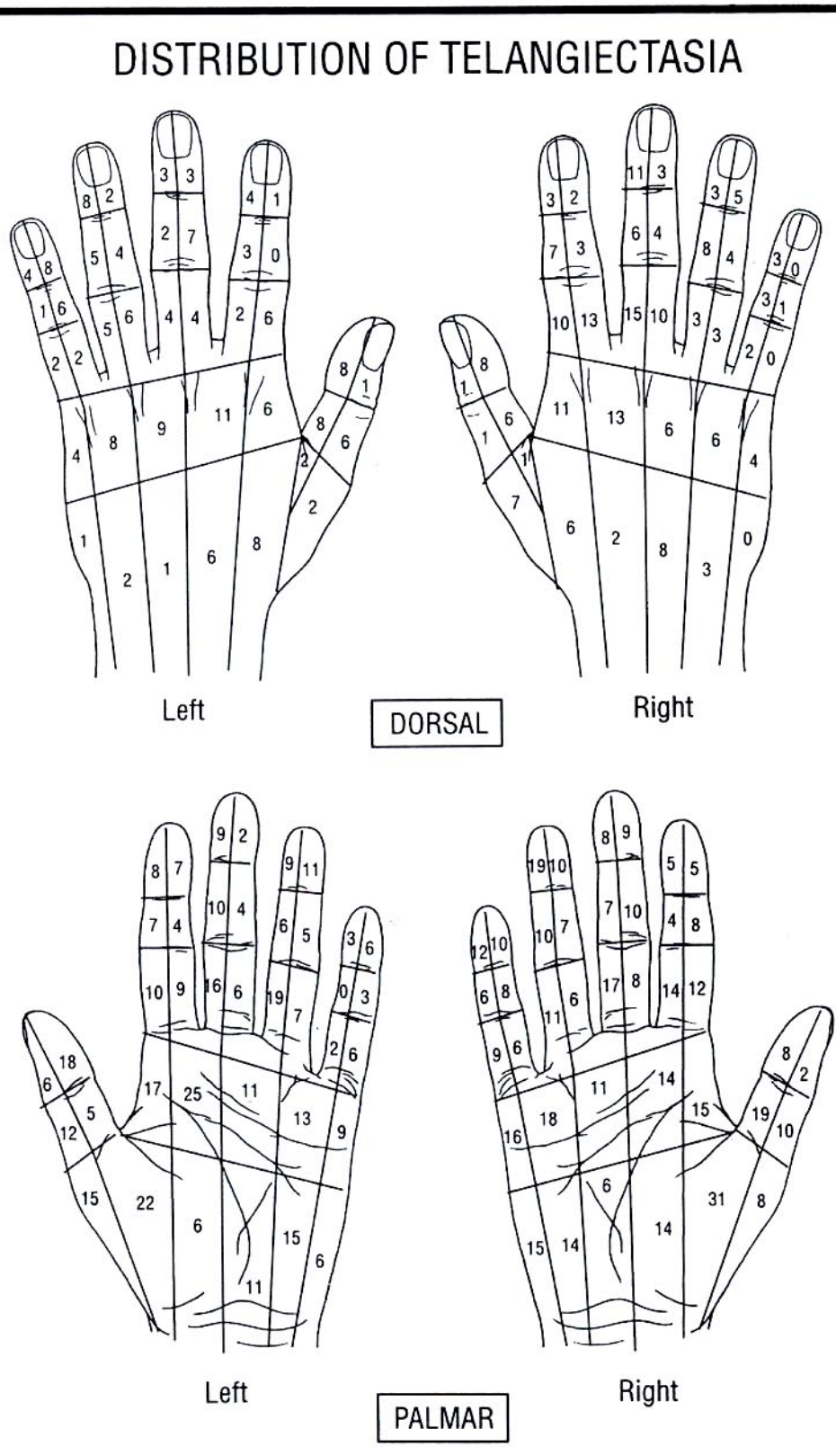

Fig. 2 Numbers of telangiectasia occurring in 158 sectors of the hands in 53 patients with scleroderma.

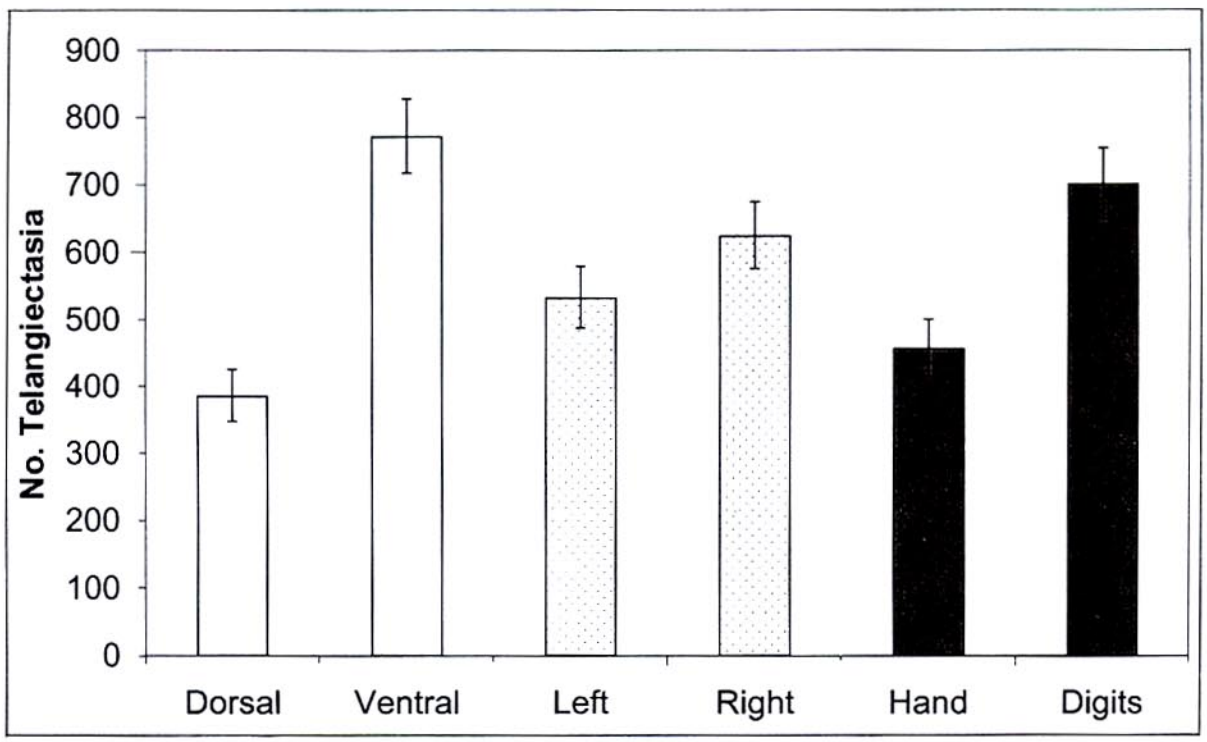

Fig. 3 Mean number (± 95% CI) of telangiectasia occurring on ventral or palmar surfaces, left and right hands and on digits or remainder of hand in 53 patients with scleroderma.

unclassified polyarthritis, B27 related arthritis and fibromyalgia. The number of hand telangiectasia in these patients varied from 1-6. Three healthy subjects demonstrated single telangiectasia whilst a fourth had 10 small telangiectasia.

The number of telangiectasia of the hands in the scleroderma subtypes is shown in Fig. 1. Patients with limited scleroderma had significantly more telangiectasia than those with diffuse scleroderma (p = 0.004) and centromere positive limited scleroderma significantly more than centromere negative limited scleroderma (p = 0.03).

In the scleroderma patients the telangiectasia were spatially distributed across all but 4 of 158 sectors of the hands with significant localization to the ventral surface of the digits (Figs. 2 and 3). Numbers of telangiectasia of hands correlated with numbers on face and lips (p = 0.001), disease duration (p = 0.002) and with surface area of calcinosis of the hands (p = 0.03). Large telangiectasia (>1 mm) correlated strongly with absolute numbers of telangiectasia (p < 0.001). The presence of telangiectasia was also associated with presence of the centromere antibody (p = 0.005).

In limited scleroderma, numbers of telangiectasia in patients with prior substantial gastrointestinal bleeds (not due to peptic ulceration) or with isolated pulmonary hypertension were compared with the remaining patients (Figs. 4 and 5). There was a trend for patients with these complications to have multiple telangiectasia and, in those patients with prior bleeds, of a size >1 mm. No correlation was observed between total number of telangiectasia and with nailfold capillary diameter (p = 0.97) or density (p = 0.66) as measured by image analysis techniques, in 12 scleroderma patients studied.

DISCUSSION

From our study we can conclude that the detection of multiple telangiectasia of the hands (> 5 in number) in patients attending a rheumatology clinic is highly suggestive of a diagnosis of scleroderma. Furthermore, multiple telangiectasia are found particularly, but

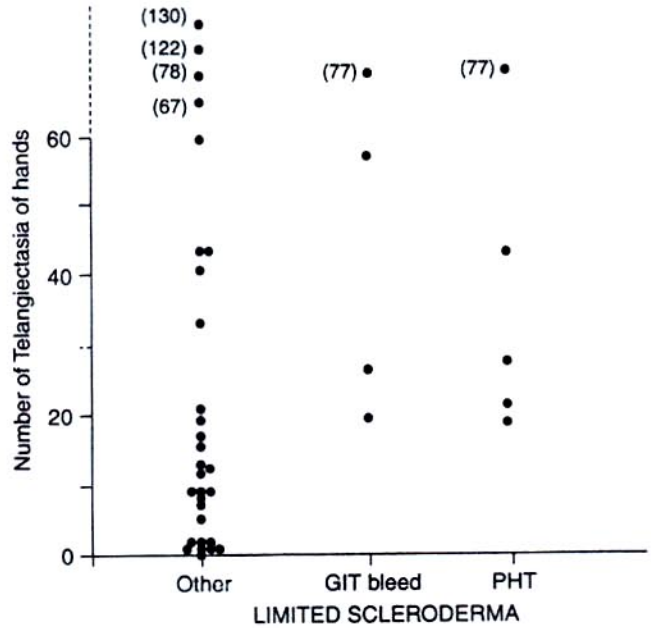

Fig. 4 Number of telangiectasia of hands occurring in 38 patients with limited scleroderma subdivided according to presence of prior gastrointestinal (GIT) bleed or isolated pulmonary hypotension (PHT). One patient had both these complications.

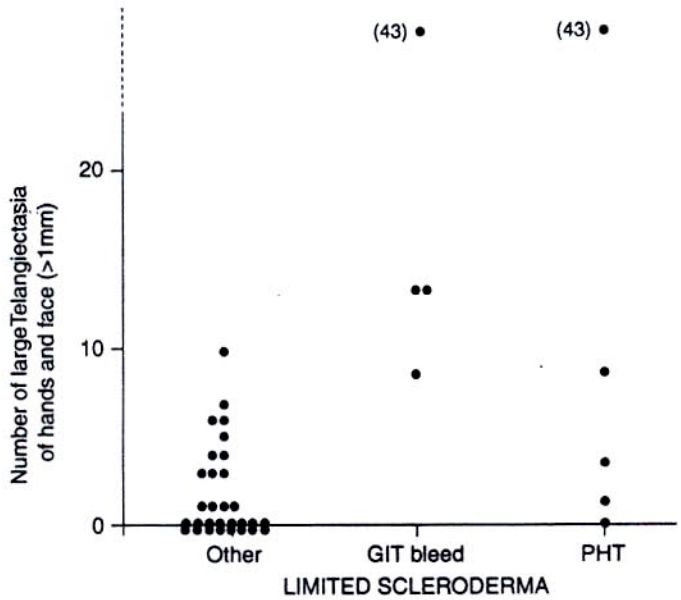

Fig. 5 Number of telangiectasia of hands and face greater than 1mm in size in 38 patients with limited scleroderma subdivided according to presence of prior gastrointestinal (GIT) bleed or isolated pulmonary hypertension (PHT). One patient had both these complications.

not exclusively, in patients with the limited subtype of scleroderma and are significantly associated with disease of long duration, with the presence of the centromere antibody and with digital calcinosis. The presence of large telangiectasia (>1 mm in size) suggests that the patient may be at an increased risk from GIT bleeding. Finally, telangiectasia of the hands are not specific for scleroderma as small numbers are seen in other rheumatic disorders or indeed, in healthy subjects.

In scleroderma the spatial distribution of telangiectasia is of some interest. Across the scleroderma cohort, telangiectasia were observed on all sectors of the skin of the hands with some preference for the ventral surface of the digits. This is in distinct contrast to the localization of calcific deposits which are found predominantly on the distal flexor surfaces of the thumb, index and fifth digits, particularly on the right side.[5] These apparent random distribution of hand telangiectasia may suggest stochastic vascular processes are in operation to explain this distribution. The significant correlation of hand telangiectasia with disease duration indicates that it is a time dependent process and furthermore, the significant correlation with number of telangiectasia on the face indicates that these vascular telangiectatic pathogenic processes are occurring in other vascular beds. Why, the beds are spatially restricted, however, *eg*.. hands, face, lips and gastrointestinal mucosa is not understood. Occasionally, one observes telangiectasia in scleroderma at other sites, *eg*. olecranon or rarely in the digits of the feet, but this is uncommon suggesting that the dermal vessels in these other sites are different to those where telangiectasia repeatedly form. The discordance between Raynaud's occurring in both the hands and feet and telangiectasia in hands only, suggests that Raynaud's phenomenon is unlikely to be causally linked. Furthermore, the occurrence of telangiectasia in other rheumatic disorders or in health indicates that the pathogenic process is not scleroderma specific. Despite the strong association with the centromere antibody, we have observed the absence of telangiectasia in those centromere positive subjects who lack any evidence of Raynaud's phenomenon or sclerodactyly. This observation indicates that the centromere antibody is not causally related to the development of telangiectasia.

We were predicting a positive correlation between nailfold capillary dilation and the number of hand telangiectasia but this was not found. To our knowledge, no such correlation has been reported elsewhere in the literature. However, our patient numbers were small, allowing for the possibility a type II error and larger studies are awaited with interest. In general, we have been impressed with the close relationship between nailfold capillary abnormalities and the sclerodermatous processes and it is an attractive hypothesis that telangiectasia in scleroderma arise because of a disruption of endothelial homeostasis in the microcirculation.

From our study we can conclude that multiple telangiectasia of the hands are most commonly observed in patients with the centromere positive limited subtype of scleroderma of long duration. A knowledge of the absolute number of telangiectasia of the hands has limited additional clinical utility, perhaps with the exception that the finding of large telangiectasia may alert the clinician to possible GIT mucosal bleeds.

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