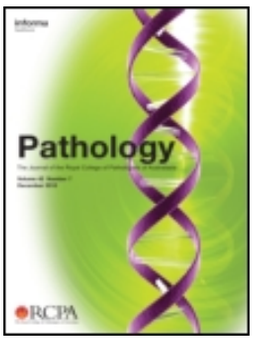

Pathology

ISSN: 0031-3025 (Print) 1465-3931 (Online) Journal homepage: http://www.tandfonline.com/loi/ipat20

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**To cite this article:** Jennifer G. Walker, John Stirling, Dimitra Beroukas, Kencana Dharmapatni, David R. Haynes, Malcolm D. Smith, Michael J. Ahern & Peter J. Roberts#Thomson (2005) Histopathological and ultrastructural features of dermal telangiectasias in systemic sclerosis, Pathology, 37:3, 220-225

**To link to this article:** http://dx.doi.org/10.1080/00313020500033262

Pathology (June 2005) **37(3)**, pp. 220–225

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ANATOMICAL PATHOLOGY

# Histopathological and ultrastructural features of dermal telangiectasias in systemic sclerosis

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Summary

*Aims:* To investigate the histological, ultrastructural and immunohistochemical features of the vascular lining of dermal telangiectasia, a characteristic clinical finding in scleroderma.

*Methods:* Standard histological, electron microscopic and immunohistological techniques were used to examine dermal telangiectasias in five patients with limited scleroderma, the most common scleroderma variant in Caucasian populations.

*Results:* The telangiectasias were dilated postcapillary venules located in the papillary and superficial reticular dermis. The vessel walls consisted of non-fenestrated endothelial cells surrounded by a variable number of pericytes and smooth muscle cells. There were no unique ultrastructural features. Thickened collagen fibres in the reticular or deep dermis were seen in all but one patient, although in variable and generally minimal quantities. Surrounding infiltrating inflammatory cells were scarce. No enhanced endothelial staining was obtained with antibodies directed against endoglin, endothelin, E-selectin and ICAM-1 suggesting a resting or inactivated state.

*Conclusion:* The immunohistological and ultrastructural features of the lining endothelium of established telangiectasias in long-standing, limited scleroderma appear benign. It would be of interest to examine telangiectasias in the early phase of their formation. Alternatively, other explanations need to be explored in understanding the aetiopathogenesis of telangiectasia in scleroderma.

*Key words:* Telangiectasia, systemic sclerosis, scleroderma, endothelium, endoglin, endothelin.

Received 20 October 2004, revised 1 March, accepted 2 March 2005

## INTRODUCTION

Telangiectasias are simply defined as visibly dilated capillaries or venules seen in the skin or mucosa. Multiple telangiectasias are particularly common in the systemic autoimmune disorder systemic sclerosis (scleroderma) where they are most commonly observed on the face and hands.[1] Their number and size tend to increase with disease duration and their occurrence is strongly correlated with the presence of the anti-centromere antibody which is a frequent finding in the more benign limited variant of scleroderma (also including the CREST subtype).[2] This limited variant is the most common form of scleroderma seen in Caucasian populations. Telangiectasias, however, are not specific for scleroderma as they are also found in other systemic rheumatic disorders, the familial disorder hereditary haemorrhagic telangiectasia (HHT), following actinic skin damage, or occasionally in health.[1]

We have recently reported in detail the clinical features of telangiectasias in scleroderma.[3] Here we report the histological and ultrastructural features of dermal telangiectasias obtained from patients with limited scleroderma. We were particularly interested in determining if there was altered expression of two endothelial proteins, endoglin (CD105) and endothelin (ET-1), both of these being implicated in the aetiopathogenesis of the vascular abnormalities observed in scleroderma.[4] Endoglin is an auxiliary protein to one of the TGF$\beta_1$ surface receptors on endothelial cells and mutations of this gene with reduced endoglin expression characterise the genetic defect in HHT.[5] We have previously shown enhanced endoglin staining of endothelium in the dermal vessels in the early active phase of scleroderma.[6] In contrast, endothelin is released from endothelial cells and is a potent vasoconstrictor.[7] Circulating levels are increased in scleroderma and there is one report of increased endothelin staining of dermal vessels in scleroderma.[8]

## METHODS

### *Patients and controls*

This was a cross-sectional study of the telangiectasias obtained by multiple 3-mm punch biopsies of the visibly dilated vessels of the otherwise uninvolved skin of the dorsum of the hand from five patients with limited scleroderma. Two to three punch biopsies of individual mat blanching telangiectasias were taken for each patient. These patients fulfilled the American Rheumatology Association criteria for scleroderma and were all listed on the South Australian Scleroderma Register.[9] All patients were female with a mean age of 74 years (range 69–79 years), mean disease duration (from first symptom of scleroderma) of 30 years (range 8–45 years) and four patients had a positive centromere antibody.

For comparative purposes, skin was also obtained from 10 patients with scleroderma during their initial diagnostic work-up, as previously described.[6] These patients consisted of six with diffuse cutaneous scleroderma (three male, three female; mean age 68 years) and four with

ISSN 0031-3025 printed/ISSN 1465-3931 
DOI: 10.1080/00313020500033262