# ARCIFORM PURPURA ANNULARIS TELANGIECTOIDES

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A case of purpura annularis telangiectoides (Majocchi's disease) with arciform lesions and marked atrophy is reported for its rarity. Lesions subsided with topical glucocorticoid application.

**Key words: Purpura annularis telangiectoides, Majocchi's disease, Atrophy**

## Introduction

Purpura annularis telangiectoides (Majocchi's disease) is a rare chronic idiopathic disease of capillaries usually seen in men between 30-50 years of age.[1] Its exact pathogenesis is not known; however drugs, labile hypertension, hypotension and polycythemia may have a role. Histopathologically it is characterized by chronic capillaritis with perivascular lymphohistiocytic infiltrate.[2] An unusual variant described by Touraine 3 shows lesions to be fewer, larger, irregular and arciform with marked atrophy. Lesions tend to be asymptomatic for months to years and eventually resolve spontaneously or with low potency topical glucocorticoid preparations.

## Case Report

A 38-year-old male presented with asymptomatic reddish patches over both shins of four years duration. Lesions were progressively increasing in size with gradual fading in the centre leaving relatively thinner skin. No other cutaneous lesions were present. General health of the patient was preserved without any history of bleeding tendencies or recurrent bruises.

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Examination revealed multiple erythematous plaque lesions 4 x 6cm size, over both shins. Lesions were arciform, annular, non-blanching with telangiectases at the periphery and giving rise to cayenne-pepper spots at place. Centre of the lesions showed marked atrophy with slight browhish hyperpigmentation. No evidence of varicose veins or stasis was found. A diagnosis of purpura annularis telangiectoides was considered. Complete blood count, and bleeding and clotting time were normal. A biopsy from the telangiectatic border of one of the plaques revealed mild lymphohistiocytic vasculitis limited to upper dermis with capillaritis along with dilatation of capillaries and endothelial cell proliferation. The lesions resolved completely leaving hyperpigmentation and mild atrophy with fluocinolone acetonide 0.025% cream applied twice daily for three weeks.

## Discussion

Majocchi's disease is a chronic form of pigmented purpuric dermatosis which usually starts symmetrically on legs and rarely may involve lower-trunk and arms with punctiform red patechial telangiectatic patches. Because of centrifugal growth, lesions coalesce forming annular or serpiginous patterns. The telangiectases eventually disappear and occasionally slight at-

rophy occurs in the center.[4] Although exact pathogenesis is not known it has been suggested that small arteries in dermis get constricted while the connecting capillaries of stratum papillae dilate in an ampulla-like fashion leading to diapedesis and bleeding. There is additional chronic capillaritis with perivascular lymphohistiocytic infiltrate.[2] A few cases have been reported but condition seems to be under reported because of its asymptomatic nature.

In the present patient disease was of cosmetic importance except marked atrophy of central part of patches which is rare in purpura annularis telangiectoides. With such atrophy it can simulate necrobiosis lipoidica diabeticorum. In purpura annularis telangiectoides marked atrophy can be due to focal elastolysis secondary to release of elastase from inflammatory cells which are probably present during active from of disease. This form with atrophy can be regarded as arciform variant of purpura annularis telangiectoides of Touraine.

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