

CHRONIC PIGMENTED PURPURIC DERMATOSES

(A Case Report)

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Summary

A case of chronic pigmented purpuric dermatoses is described in detail. The case presented with bilateral symmetrical lesions on both the legs of four years duration.

The specific eponyms applied to chronic pigmented purpuric dermatoses include:—

- (1) Progressive pigmentary dermatoses (Schamberg's disease)
- (2) Purpura annularis telangiectodes (Majocchi's disease)
- (3) Pigmented purpuric lichenoid dermatitis (Gougerot and Blum disease)
- (4) Eczematid like purpura (Doucas and Kapetanakis disease)

Angioma serpiginosum (Hutchinson's disease) is sometimes erroneously included in this group.

They are closely related to one another and probably are variants of each other. Often they cannot be differentiated on clinical or histological grounds. To date the etiology of this group of disorders is unknown.

Clinically the primary lesion consists of purpuric puncta appearing in groups and extending slowly so that patches of various sizes are formed. Gradually telangiectatic puncta may appear as the result of capillary dilatation and pigmentation as the result of haemosiderin deposits. In some cases telangiectasia predominates (Majocchi's disease) and in others pigmentation (Schamberg's disease). Not infrequently clinical signs of inflammation are present, such as, erythema, papule and scaling (Gougerot and Blum disease) or papules, scaling and lichenification (Eczematid like purpura). Often the disorder is limited to the lower extremities, but it may be extensive. Gravity and increased venous pressure are important localizing factors in many cases. The intrinsic haematological factors show no significant aberration nor are there accompanying constitutional disturbances as a rule. The Rumpel-Leede tourniquet test for capillary fragility is positive in over one half of the cases.^{1,2,3}

Histologically they are all similar. Common denominator being increased dermal capillary permeability and fragility leading on to extravasation of red cells. Haemosiderin deposits are present in macrophages. The basic process is

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a lymphocytic type of vasculitis usually limited to upper dermis. The infiltrate may invade epidermis and provoke mild spongiosis of the stratum malpighii and patchy parakeratosis. In older lesions the capillaries often show dilatation of their lumen and proliferation of their endothelium.^{2,4}

Differential diagnosis must include stasis dermatitis, anaphylactoid purpura carbromal sensitivity, purpuric clothing dermatitis and hyperglobulinaemic purpura.

Case Report

A 29 years old male reported in October 1970 to the Skin and V. D. Clinic of V. J. Hospital, Amritsar, with the complaint of pigmented areas on both legs of 4 years duration (Fig. page No. 192).

Four years ago the disease started as reddish streaks with mild itching in the middle of left shin and gradually changed its colour to dull brown and got enlarged. More lesions appeared on the same and other leg later. At the time of the hospital visit the disease had been somewhat stationary for 1½ years. There was no history of trauma or taking of carbromal like drugs.

There was nothing contributory in the past, family or personal histories.

Examination showed a well built and well nourished individual. Pulse was 84/mt. and regular, B.P. 130/85mmHg, Temperature 98°F. No significant lymphadenopathy was present.

Examination of abdomen, chest, heart and nervous system revealed no abnormality.

Hess capillary test was negative both in upper and lower extremities.

Local examination showed that legs were involved all round. They were below the knees and extended upto dorsae of feet. The lesions were in the form of brownish black irregular patches 1-5 cm. in diameter. The pigmentation gradually merged with the surrounding skin, giving the lesions 'Smoked paper' appearance. The patches were non-tender and no exudation was present. At places small adherent scales were seen. The skin was neither thickened nor atrophied. There was no loss of hair or telangiectasia. There were no excoriation marks.

Investigations : Hb, WBC. T.&D. Rbc.count, Platelet count, Bleeding time, clotting time, fasting blood sugar, Blood urea, S.Proteins--Total, Albumin & Globulin, VDRL & Kahn test, Routine Urine and stools all revealed normal values. Blood for L.E. Cells were negative.

Skin Biopsy : Revealed patchy areas showing proliferation of young capillaries and an occasional one showing proliferation of endothelial cells. There was infiltrate mostly of round cells. At places in addition haemorrhages were present. A diagnosis of chronic pigmented purpuric dermatoses was made.

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