Institute of Experimental Morphology and Anthropology with Museum
Bulgarian Anatomical Society

Acta morphologica et anthropologica, 12
Sofia • 2007

Livedo Reticularis: A Clinico-Pathological Study

Mary Gantcheva

Department of Dermatology, Medical Faculty, Sofia

We studied a series of 10 patients with livedo reticilaris. Four of them were diagnosed as livedo vasculitis or so called segmental hyalinized vasculitis, two with idiopathic livedo reticularis and four of them were with Syndroma Sneddon. Histological examination of livedoid network disclosed in all cases the presense of thrombotic occlusion of venules and capillaries in papillary and reticular dermis, with occasional fibrinoid necrosis of the vascular wall, sparse perivascular lymphocytic infiltrate and variable extravasation of erythrocytes. In biopsy specimens obtained from areas adjacent to, but not involved by ulceration arteriolar thrombosis was also seen. Neither neutrophils nor nuclear dust were ever presented.

Our findings show that the capillary and venular thrombosis is the common histological hallmark of livedo reticularis, suggesting that vascular occlusion, but not leucocytoclastic vasculitis, is the primary event of the clinical condition: livedo reticularis.

Key words: Livedo reticularis, livedo vasculitis, Syndroma Sneddon, morphology, histopathology.

Introduction

Livedo reticularis (LR) is a persistent violaceous reticulated skin pattern usually found on the extremities and rarely trunk. Its coloration is due to the stagnation of blood in dilated superficial capillaries and venules, and implies pathologic changes in the deeper larger vessels. Normal skin areas are clearly demarcated from affected ones [1]. This type of lesion may occur on cold exposure in healty people or in a variety of pathologic states characterized either by blood hyperviscosity or by pathology in the arterioles, capillaries, or venules of the skin [2]. LR occurs in connective-tissue diseases such as systemic lupus erythematosus, thrombotic disorders such as thrombotic thrombocytopenic purpura, idiopathic thrombocytopenia. It is a main clinical sign of atheromatous vascular disease and occlusive vasculopathies, such as Livedo vasculitis (LV) [3, 4]. It could be found also in cerebrovascular thrombosis as Syndroma Sneddon [5] and antiphospholipid syndrome [1, 6].

Materials and Methods

We have reviewed the hystopathologic findings in 10 patients with skin manifestation of LR. Four patients carried a diagnosis of LV (3 females and 1 male, mean age 31 years). Clinically they were characterized by focal purpuric painful lesions of the lower legs (three

of the patients had necrotic ulcerations around the ankles), livedo like areas of hemosiderinic hyperpigmentation, reticulate of atrophie blance (stellate porcelain-white atrophic scars) and signs of LR.

Four patients (4 females, mean age 64 years) had Syndroma Sneddonps with LR distributed on the extremities, cerebrovascular accidents and negative anticardiolipine antibodies.

Two of the patients (2 females, mean age 25 years) included in the study were diagnosed as idiopathic LR as they had only the skin lesion of cyanotic discoloration of the legs (cutis marmorata). Ten skin biopsy specimens, taken from all the cases were examined. They were fixed in 10% neutral-buffered formaldehyde solution, processed routinely, and stained with hematoxylin-eosin.

Results and Discussions

The histological findings from the patients with LV showed results of microthromboses in dermal vessels. Some blood vessels in upper derma were with swollen endotel, some were dilated, as the others were with partly or totally obstructed lumen because of microthromboses (Fig. 1). The histopathological findings from the lesions from patients with LV were homogeneous,showing thromboses mostly in medium and upper dermal vessels, endothelial swelling, segmental signs of hyalinization, extravasated erythrocytes and a variable grade of perivascular lymphocyte imfiltration (Fig. 2). Idiopathic LR showed morphological changes consisted of thickned wall and endothelial cell oedema that were caused stenosis of the vessels. Sparse round-cell inflammatory infiltrate in the dermis round the vessel was presented (Fig. 3).

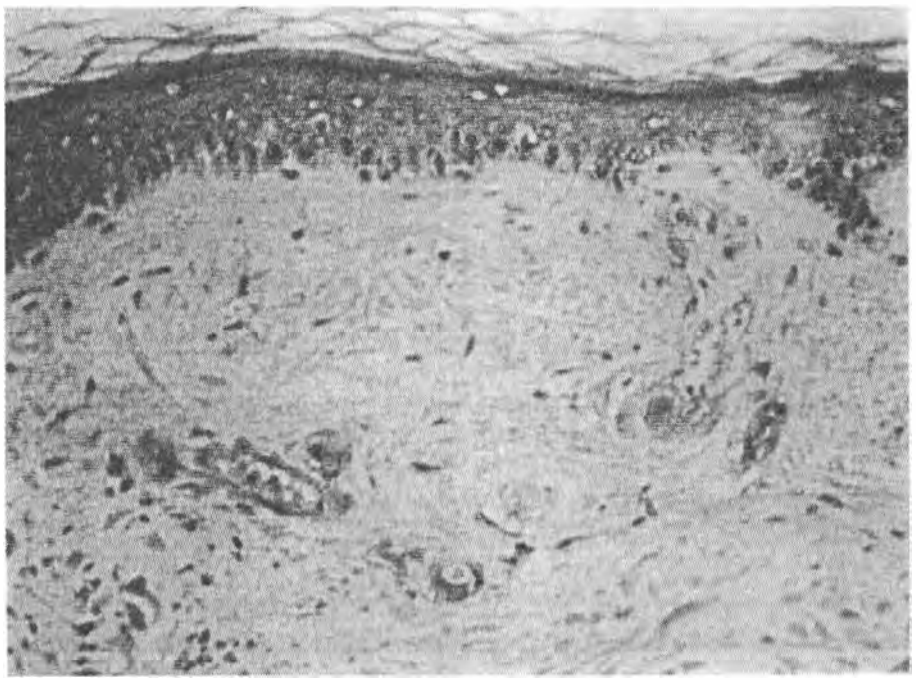

Fig. 1. Some blood vessels are with partly or totally obstructed lumen because of microthromboses. HE (×150)

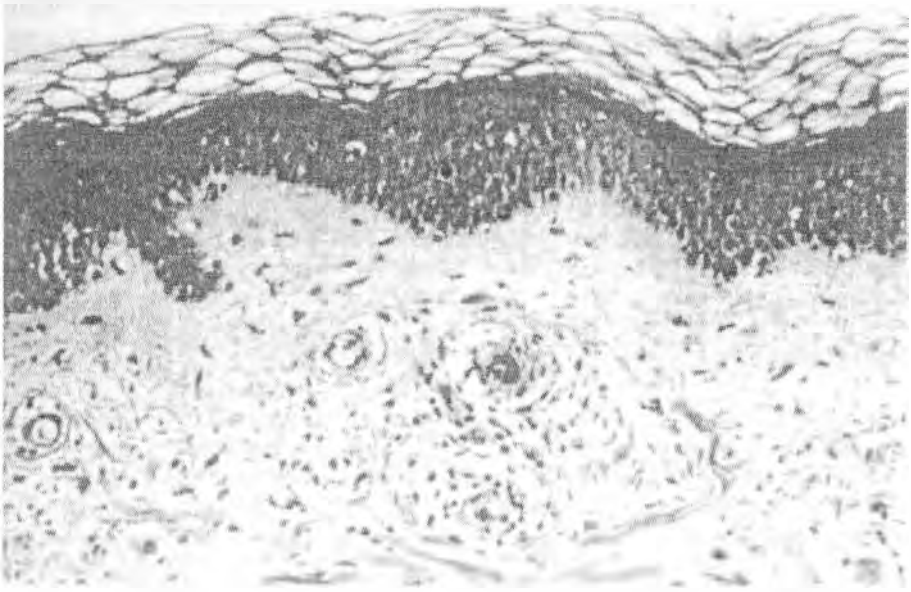

Fig. 2. Microvascular thromboses, slight perivascular lymphocytic infiltration in the upper dermis, without leucocytoclasis, segmental hyalinization of the dermal vessels. HE (× 80)

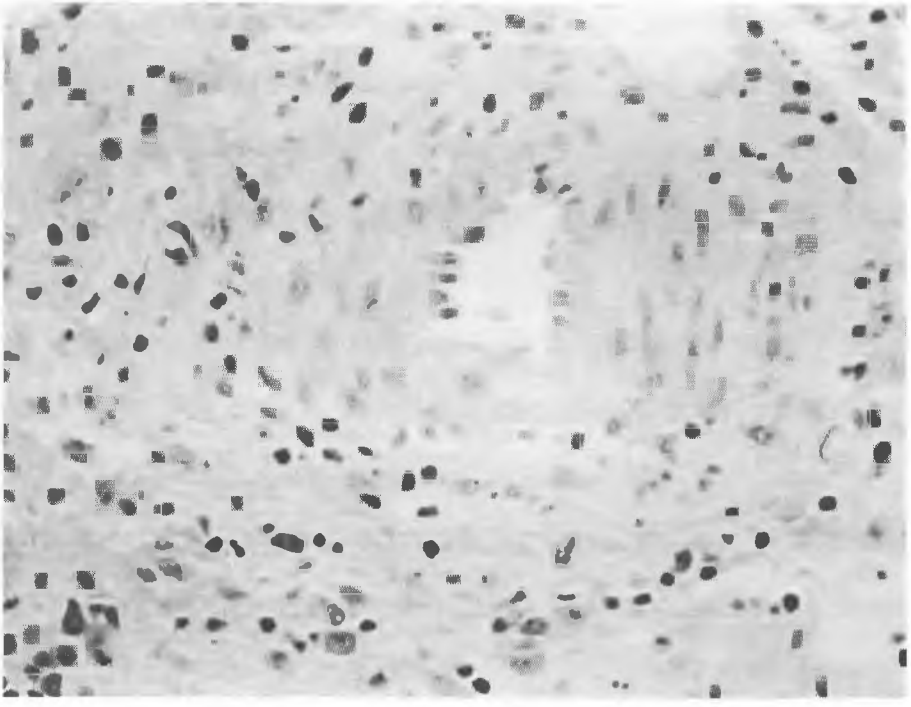

Fig. 3. Thickened wall and endothelial cell oedema that caused stenosis of the vessels, sparse roundcell inflammatory infiltrate. HE (× 250)

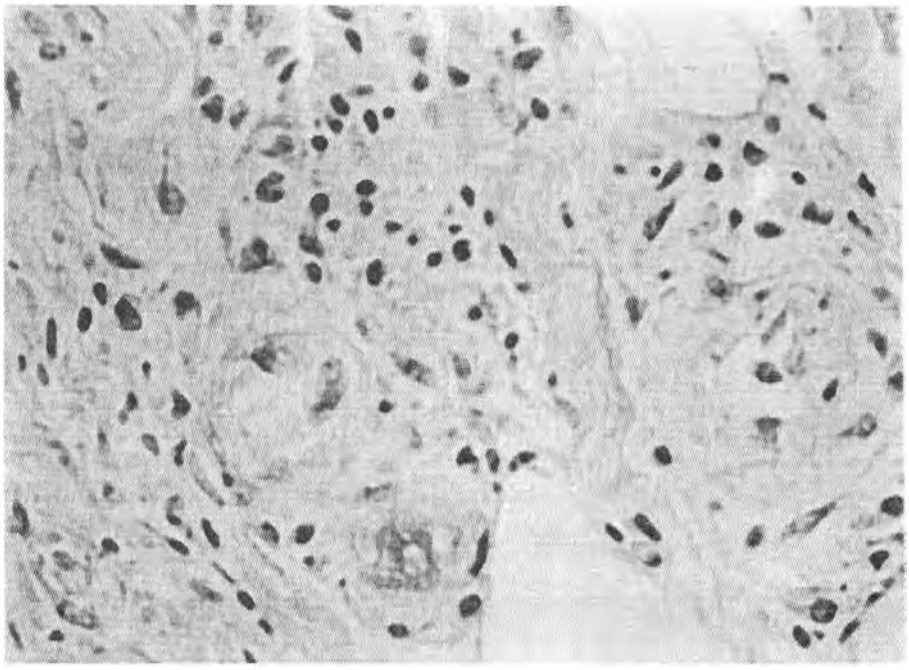

Fig. 4. Prominent endothelial cell oedema of capillares and post capillary venules. One of the capillaries is totally obturated with a plug by a thrombus. HE (× 250)

Thrombosis in the dermal vessels were found also in the biopsies from the lesions from patients with Syndroma Sneddon. It was seen that the capillary was with a totally plugged lumen as thrombosis existed. Prominent endothelial cell oedema of capillaries and post capillary venules was seen. One of the capillaries was totally obturated with a plug by a thrombus. Sparse round-cell infiltrate was seen round the vessels (Fig. 4).

Our findings show that capillary and venular thrombosis is the common histological hallmark of LR, suggesting that vascular occlusion, but not leucocytoclastic vasculitis, is the primary event of these clinical conditions. The mechanism of LR is unclear, but we think that the vasculopathy with occlusion is the main histopatological sign of LR. We didn't see the features of vasculitis. It could be demonstrated histologically when ulceration coexist with LR in systemic lupus erythematosus and neutrophils and nuclear dust could be seen [7].

Because of the histological absence of leucocytoclasia and the common finding of dermal vessel microthromboses, the hypothesis of a vasooclusive pathogenesis could be accepted.

References

1. G a n t c h e v a, M. Dermatological aspects in antiphosphoilipid syndrome. – Int. J. Dermatol., **36**, 1997, 173-180.
2. C o p e r m a n, P. Livedo reticularis: signs in the skin of disturbance of blood viscosity and of blood flow. – Br. J. Dermatol., **93**, 1975, 519-529.
3. P a p i, M., B. D i d o n a, O. De P i t a, M. G a n t c h e v a, L. C h i n n i. Purple (atrophie blance): clinical histological and immunological study of twelve patients. – J. Europ. Acad. Dermatol. Venerol., **9**, 1997, 129-134.
4. F r i t s c h, P., B. Z e l g e r. Livedo vasculitis. – Hautartz, **46**, 1995, 215-224.
5. G a n t c h e v a, M., N. T s a n k o v. Livedo reticularis and cerebrovascular accidents (Sneddon's syndrome) in the spectrum of antiphosphoilipid syndrome. – J. Europ. Acad. Dermatol. Venerol., **12**, 1999, 157-160.
6. G r o b. J., J. B u n e r a n d i. Cutaneous manifestations associated with the presence of the lupus anticoagulant. – J. Am. Acad. Dermatol., **15**, 1986, 211-219.
7. K u h n, A., P. L e h m a n n., T. R u z i c k a. Vasculitis and cutaneous lupus erythematosus. – In: Cutaneous lupus erythematosus. Berlin, Heidelberg, New York, Springer-Verlag, 2004, 156-160.