8 Skin Manifestations of Antiphospholipid Syndrome

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Several skin manifestations have been described in patients with antiphospholipid syndrome (APS) (Table 8.1) [1–3]. The most frequent skin lesions are livedo reticularis and skin ulcers.

Vascular occlusion is generally the first and most frequent manifestation observed in patients with antiphospholipid antibodies (aPL), accounting for 41% of the cases. Forty percent of these patients present with other multisystem thrombotic phenomena during the course of the disease, underscoring the significance of skin lesions as a diagnostic marker and predictor of systemic involvement.

In spite of the association of skin lesions with different isotypes of immunoglobulins, the presence of IgA anticardiolipin antibodies (aCL) has been reported as an independent predictive factor for skin lesions (skin ulcers, chilblain lupus, and vasculitis) [4].

Livedo Reticularis

Livedo reticularis is the most common skin manifestation in patients with APS, characterized by a dark purple reticular pattern usually involving the upper and lower limbs [3, 5].

The skin normally receives its blood supply through a vascular system arranged in the form of cones with their base towards the skin surface. Each cone is supplied by an arteriole. The pattern of livedo reticularis corresponds to areas of anastomo-

Table 8.1. The skin and antiphospholipid syndrome.

- Livedo reticularis
- Sneddon's syndrome
- Skin ulcers
- Necrotizing vasculitis
- Livedoid vasculitis
- Cutaneous gangrene
- Super cial thrombophlebitis
- Pseudovasculitic lesions: Nodules, papules, pustules, palmar–plantar erythema
- Subungual bleeding
- Anetoderma

Table 8.2. Livedo reticularis and associated diseases.

- Antiphospholipid syndrome
- Systemic lupus erythematosus (with or without aPL)
- Systemic vasculitis (polyarteritis nodosa, cryoglobulinemia)
- Pseudovasculitic syndromes (cholesterol embolization)
- Overlapping syndromes
- Scleroderma
- Infectious diseases (syphilis, tuberculosis)

sis between the two cones where diminished blood flow is associated with the dilatation of venules and capillaries. Therefore, the alteration in arterial blood flow caused by the livedo may result from:

- Blood inflow obstruction
- Blood hyperviscosity
- Blood outflow obstruction

Livedo may be observed in normal subjects, especially women, after exposure to cold, displaying a symmetrical and regular mottled pattern. However, the relationship with a large number of pathological conditions (Table 8.2) is very important. A detailed examination of the features of the reticular pattern, including location, extension, symmetry, and regularity, and the presence of associated skin lesions will contribute to the differential diagnosis [3, 6–8].

The pattern of involvement associated with APS is generally disseminated, with incomplete circular segments, non-infiltrated, persistent, or irregular with wide ramifications (livedo racemosa). Some patients present a fine, regular, and complete network (Fig. 8.1).

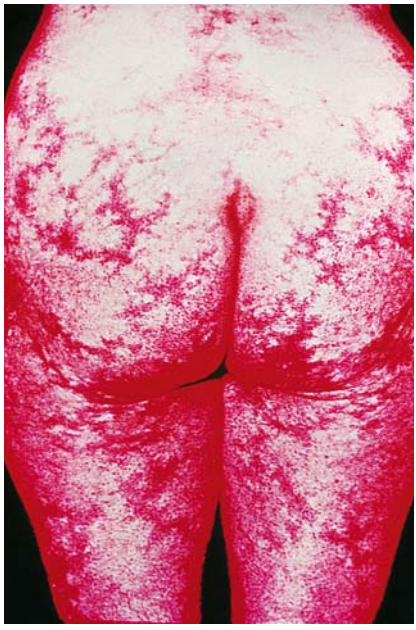

Figure 8.1. Livedo reticularis of the gluteal region and both thighs of a patient with SLE and APS.