Use of intravenous immunoglobulin in livedo vasculitis

H. Amital, Y. Levy, Y. Shoenfeld

ABSTRACT
We describe a 36-year-old woman with livedo vasculitis (atrophie blanche) lasting for 15 years. After the failure of numerous therapeutic modalities the patient was treated with intravenous immunoglobulin. Significant resolution of the cutaneous lesions was seen with a concomitant alleviation of pain. To the best of our knowledge this is the first patient with livedo vasculitis described in the medical literature ever to be treated with IVIG and to achieve a successful outcome.

Introduction
Livedo vasculitis is a chronic cutaneous condition characterized by grouped and reticulated macules and papules which progress into excruciating ulcers, taking place primarily in the foot or in the distal portion of the foot. These ulcers turn subsequently into ivory-white plaques of sclerosis tinged with telangiectasias and encircled by hyperpigmentation.

Livedo vasculitis occurs most often in young to middle-aged women, although it has been described in males and during childhood as well (1). The etiology of the disorder is uncertain. It seems that regardless of the origin of the pathological process it ends with obliteration of the capillary lumen by endothelial proliferation, fibrin deposition and the formation of thrombi.

Livedo vasculitis usually presents as a solitary finding but several associations with rheumatic diseases such as systemic lupus erythematosus, rheumatoid arthritis, Raynaud’s phenomenon, scleroderma and the antiphospholipid syndrome have been reported (2-6). The association of these disorders with livedo reticularis, which has an established autoimmune origin (7), has led many physicians to treat patients with non-steroidal anti-inflammatory drugs (NSAIDs), corticosteroids, and cyclophosphamide (6). In this communication we describe the outcome of IVIG therapy in a patient with progressive disease resistant to treatment.

Case report
A 36-year-old woman was referred to our department due to a recent exacerbation of livedo vasculitis. The eruption had begun 15 years prior to this hospitalization and progressively developed into her current condition. Before the age of twelve years she had suffered a single attack of rheumatic fever without any known sequel. Previously she was treated with NSAIDs, warfarin, heparin, prednisone, azathioprine, cyclophosphamide and local plastic surgery without any real success.

On examination deep giant ulcers reaching a diameter of up to 7 cm were located on both of her feet and on the distal aspects of her legs. Additionally, blanching erythematous patches covered the plantar aspects of her feet and her ankles. Telangiectasias encircled the lesions as well as hyperpigmented sclerotic tissue. Avascular hypopigmented scars covered healed lesions (Fig. 1). No other findings were noted in her physical examination. Except for a mildly elevated sedimentation rate (60 mm/hr), no other abnormal laboratory findings were recorded. A biopsy from one of the lesions confirmed the diagnosis. The patient was treated monthly with a 2 gr/kg dose of IVIG divided equally throughout 5 days. Within 3 months we noticed healing of the ulceration, a significant relief of pain and alleviation of the erythematous eruption (Fig. 2).

Discussion
Livedo vasculitis (also termed atrophie blanche, segmental hyalinizing, vasculitis, livedo reticularis) is a chronic cutaneous disorder of young to middle-aged women that is characterized by persistent painful leg ulcerations. Primary lesions consist of purpuric macules and papules which undergo superficial ulceration, followed eventually by the development of irregular, atrophic, porcelain white scars with fine borders of ectatic vessels.

The etiology of this disorder is not clear. Several communications indicated that the disease is related to deposits of fibrin within the pericapillary cuffs (8). Others linked this finding to the markedly suppressed release of vascular tissue plasminogen activator found in these patients (9). Uncommon familial disorders of livedo vasculitis and a coexistence with chronic myelogenous leukemia have been observed as well (10, 11).
However, many implications have linked this disorder to autoimmunity: several reports have described the coexistence of livedo vasculitis with rheumatic autoimmune conditions like SLE, scleroderma and antiphospholipid syndrome (2-6). Furthermore, one of the beneficial modalities of treatment in this disorder is immunosuppressive therapy.

In this communication we describe a 36-year-old female with a progressive and agonizing disease lasting 15 years. Following numerous attempts to assist this patient with a large spectrum of medications and surgical intervention she was referred to our department for IVIG therapy. IVIG has become one of the cornerstones of modern therapy for autoimmune conditions (12-15) and is now referred to our department for IVIG treatment of psychosis secondary to SLE with high dose intravenous immunoglobulin. Clin Exp Immunol 1992; 10: 391-3.


20. MOBINI N, SARELA A, AHMED AR: Intravenous immunoglobulins in the therapy of autoimmune and systemic inflammatory disorders.

