

Livedoid vasculopathy associated with cryoglobulinemia revealing multiple myeloma: a case report

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Introduction

Livedoid vasculopathy is a rare chronic painful dermatosis of the lower limbs evolving in flare-ups related to a process of micro thrombosis of the vessels of the dermis. The diagnosis is late, often confused with vasculitis and its treatment remains controversial and often ineffective. We report a case of livedoid vasculopathy associated with cryoglobulinemia and successfully treated with rivaroxaban.

Case report

A 70-year-old female patient with a pathological history of venous insufficiency and a recurrent ulcer of the right lower limb treated with antiplatelet medication for 1 year, was hospitalized for ulcerative-necrotic, erosive, and annular lesions that were initially vesiculobullous and purpuric involving symmetrically the lower limbs. An arterial echo-Doppler and an angiography scanner were performed and returned negative. A biological assessment was unremarkable except for the presence of type 1 cryoglobulin, IgG Kappa isotype. Anatomopathological examination showed that the superficial and deep dermis vessels were congestive, showing inflammatory damage to their walls with numerous fibrinoid thrombi obstructing their lumen, with endothelial proliferation. Standard cytogenetic and FISH studies were in accordance with a multiple myeloma type (MGUS): monoclonal gammopathy of undetermined significance. The therapeutic management consisted of

corticosteroid therapy and Rivaroxaban at 20 mg daily. A good evolution was noted with regression and healing of the lesions and the disappearance of the pain within one month.

Discussion

Our observation is particular by its clinical presentation, the association with cryoglobulinemia, multiple myeloma, and the favorable evolution under rivaroxaban treatment. The typical clinical manifestation of LV is a triad composed of a livedo racemosa of the skin, and painful episodic ulcerations of the distal face of the legs followed by white-porcelain scars called "white atrophy" (1). Our patient presented with a very extensive necrotic feature of the lesions reflecting the severity of the involvement and the association with coagulopathy and venous insufficiency. LV is seen in patients with and without identifiable coagulation abnormalities or comorbidities such as connective tissue diseases and hematologic malignancy. The association with multiple myeloma is rarely reported. Treatment of LV remains challenging and is currently uncodified. However, various treatments have been studied such as anticoagulants, anabolic steroids, intravenous immunoglobulins, and antiplatelet agents, and have shown good clinical results (2). Many studies reported the beneficial effects of treating LV with rivaroxaban. This suggests the existence of an occult coagulation anomaly which presaged the occurrence of livedoid vasculopathy. Our case supports the potential utility of rivaroxaban in the treatment of livedoid vasculopathy with few adverse effects.