Severe Leg Ulcers in a Multiple Myeloma Patient with Cryoglobulinemic Vasculitis

Multiple myeloma (MM) is a hematological malignancy characterized by the aberrant expansion of plasma cells. Patients with MM may develop cryoglobulins which are immunoglobulins that precipitate at cold temperatures (1). Cryoglobulinemia (CG) may present with a hyperviscosity syndrome and/or small vessel vasculitis. There are three types of CG. Type 1 accounts for 10–15% of CG and is associated with hematologic malignancies (1). Types 2 and 3 are associated with chronic infections and autoimmune disorders. Herein, we present a 62-year-old man with an eight-year history of MM-IgG-kappa who developed severe and intractable ulcers in lower extremities. Physical examination showed a large necrotic ulcer in the right leg with surrounding purpuric lesions and livedo reticularis (Figure 1A). In the left leg, two ulcers with the same characteristics were present over the medial malleolus and the dorsal aspect of the foot (Figure 1B). Laboratory tests showed normochromic normocytic anemia (Hgb = 7.2 g/dl), thrombocytosis (Plt = 537 x10³/mm³), and markedly elevated serum IgG levels (5784mg/dl). Serum cryoglobulins were detected. Rheumatoid factor was negative. C3 complement levels were decreased whereas C4 complement levels were normal. Skin biopsy of a healed ulcer revealed eosinophilic deposits within the vascular lumen. Biopsy of an active ulcer showed leukocytoclastic vasculitis. These findings were consistent with cryoglobulinemic vasculitis. The patient was treated with local care (debridement of necrotic tissue and appropriate dressings), bortezomib and dexamethasone followed by plasmapheresis for seven days. Two months later the ulcers decreased in depth, without necrotic tissue or surrounding livedo reticularis (Figures 2A and 2B). Only few cases of CG associated with MM have been reported. Overall, cutaneous manifestations are present in most patients varying from purpuric lesions to severe necrotic lesions and gangrene requiring amputation (2-5). As in our patient, nearly half are responsive to plasmapheresis. Treatment for severe MM-related type 1 CG requires prompt and effective MM treatment and plasmapheresis to achieve a rapid control of CG-related symptoms and to avoid disease relapse.

References


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Figure 1. (A) Right leg: Large necrotic ulcer with contiguous purpuric lesions and livedo reticularis. (B) Left foot: Necrotic ulcer over the medial malleolus surrounded by purpuric lesions and livedo reticularis.

Figure 2. (A) Right leg and (B) left leg: Healthy granulating wound with epithelialization 2 months after plasmapheresis. Note the complete resolution of livedo reticularis and purpuric lesions.