Case Report

A 43-year-old man developed several discrete, painful, erythematous small macules and bullae with dusky violaceous centers that evolved in skin erosions and ulcerations on both legs (Figures 1a, 1b). The patient referred pain and swelling of his knees and ankles and he reported an episode of rectal bleeding. He was a smoker, had not lost significant weight, and was afebrile; otherwise his physical examination was normal and his vital signs were stable. Ematochemical assay demonstrated an increase of mean corpuscular hemoglobin (MCH), red cell distribution width (RDW), fibrinogen (PT-Fg), and gamma-glutamyl transpeptidase (GGT). Erythrocyte sedimentation rate (ESR) and c-reactive protein (CRP) were normal. Antinuclear antibodies and cryoglobulins were negative while myeloperoxidase antibody (MPO-ANCA) was positive. Enzyme-linked immunosorbent assay revealed that MPO-ANCA was 246 EU and antiproteinase-3 antibody (PR3-ANCA) was negative. Gastroenterologist evaluation showed no signs of inflammatory bowel diseases and the rectal bleeding was attributed to hemorrhoids.

A skin biopsy taken from the lesions suggested a leukocytoclastic vasculitis and direct immunofluorescence (IF) was negative. Renal function and echography were normal. A CT scan excluded a pulmonary involvement or alveolar hemorrhages. There were no signs of uveitis or visual disorder. The final diagnosis was microscopic polyangiitis with a rare presentation that was limited to the skin. Local medications were started with hydrogel three times a week and systemic therapy with prednisone 45-mg/day gradually reduced over 3 months. Local wound care with hydrocolloid and collagen
Figures 1a, 1b. Vasculitic ulcers in MPA.

Figure 2. Histological pattern of leukocytoclastic vasculitis.

Figures 3a, 3b. Clinical appearance following therapy.
dressings 3 times/week was started and completely resolved the wounds within 4 months (Figures 2a, 2b).

**Discussion**

Microscopic polyangiitis (MPA) is a vasculitis of small to medium size vessels according to Chapel Hill Consensus Conference (CHCC) classification. It belongs to the family of anti-neutrophil cytoplasmic auto-antibody (ANCA)-associated vasculitis and is strongly associated with anti-myeloperoxidase (MPO)-ANCA.

Skin involvement is usually linked to internal manifestations and resolves with systemic treatment. The main symptoms are characterized by renal involvement (78.8%), weight loss (72.9%), skin manifestations (62.4%), fever (55.3%), mononeuritis multiplex (57.6%), arthralgias (50.6%), myalgias (48.2%), hypertension (34.1%), lung involvement (24.7%), alveolar hemorrhages (11.8%), and cardiac failure (17.6%).

Although skin involvement is frequent, a precise description has been limited. Patients usually present purpura and petechiae, livedo, and erythema especially on the hands and fingers.

Only one case of cutaneous involvement without visceral diseases has been described in the literature.

Therapy is often divided into two phases: an initial “remission-induction” phase is used to control active disease and a “maintenance” phase, which uses less intensive therapy and is used to maintain disease remission while lowering the risk of adverse medication related events. The first-line treatment is represented by cyclophosphamide plus corticosteroids or methotrexate plus corticosteroids. Treatments such as plasma exchange, intravenous immunoglobulin, anti-tumor necrosis factor (TNF)-α therapy, T-cell depletion therapy with antithymocyte globulin, and B-cell depletion therapy with rituximab have been used. After the induction of clinical remission with an agent such as cyclophosphamide, patients are generally converted to azathioprine or methotrexate; additional agents that have been used in selected patients include mycophenolate mofetil (MMF), leflunomide, and cyclosporine.

The diagnosis of pyoderma gangrenosum would make sense in this case but was excluded due to the absence of systemic disease and the clinical course.

**Conclusion**

To our knowledge this is a rare case of microscopic polyangiitis characterized by a long time cutaneous involvement in absence of any internal organ lesions and it demonstrates that clinical manifestations may be limited to the skin. It is possible that a MPA variant limited to the skin may exist and it does not evolve or rarely relates to systemic disease. Further studies are necessary to characterize this possible variant. We hope this report contributes to the ongoing development of knowledge about the clinical-pathological spectrum of MPA and provides further indications about the wide range of its possible clinical manifestations.

**References**