

CORRESPONDENCE

Chronic vasculitis urticaria associated to a monoclonal gammopathy of IgM and IgA type, a Schnitzler Syndrome?

Urticaria is a skin disease affecting about 25% of the population and clinically appearing as crops of inflammatory wheals [1]. Schnitzler Syndrome (SS) is a rare condition, characterized by the simultaneous occurrence of chronic urticaria and IgM monoclonal gammopathy, associated with at least two of the following features: fever, arthralgia, bone pain, hepato-splenomegaly, lymphadenopathy, increased ESR, leukocytosis and increased bone density. It belongs to the group of dysproteinaemia-associated skin diseases with a favourable prognosis, even if patients have an increased risk of developing lymphoproliferative disorders. In the majority of cases, the monoclonal component is an IgM type, but several cases with IgG or IgA monoclonal gammopathy components have been reported [2].

We describe a 71-year-old woman, referred to our department for multiple urticarial skin patches located on the trunk and upper and lower limbs, present for fifteen years (*figures 1A,B*); the lesions were mildly pruriginous and did not disappear despite antihistaminic and corticosteroid therapy. In her past medical history, a monoclonal gammopathy of undetermined significance (MGUS), of IgM κ type associated with a IgA κ component, had been diagnosed about fifteen years before in concomitance with the appearance of the urticarial flares. Haemato-chemical tests, autoimmunity screening (ANA, ENA, Lupus anticoagulant, Rheumatoid Factor) and thrombophilic tests

(serum cryoglobulins and anticardiolipin antibodies) were in the normal ranges, except for elevated levels of ESR (94 mm/hr), serum C reactive protein (1.07 mg/dL), total IgA (1,500 mg/dL), total IgE (40.80 UI/mL) and the confirmation of a monoclonal component in the beta zone of electrophoresis (2.33 gr/dL), characterized by an IgM κ and an IgA κ pattern at immunofixation. A skin biopsy from one lesion showed swelling of endothelial cells, fibrinoid necrosis of vessel walls, neutrophilic infiltration with leukocytoclasia and extravasation of red blood cells, with no deposits of IgM or IgA at direct immunofluorescence assay. These features were compatible with the diagnosis of leukocytoclastic vasculitis-urticaria. The patient presented a chronic pain in the seventh right rib without any medical history of a previous fracture, so we performed a chest X-ray which showed an area of bone densification with a cortical hyperostosis, associated to periosteal apposition. A diagnosis of Schnitzler Syndrome was finally made, and the patient was treated with corticosteroids (deflazacort, 15 mg daily) during the flares of cutaneous manifestations, with good results.

The pathophysiology of SS is unclear; some authors focused attention on the IgM deposits localized on the dermoepidermal junction as well as on the capillary walls, with the subsequent complement activation, leading to tissue damage and probably responsible for the urticarial skin lesions [3]. Nevertheless, this feature is only detected in 25% of patients affected by SS [4]. Other studies consider that IL-1 is probably involved in the pathophysiology of the disease, so that the recombinant IL-1 receptor antagonist (Anakinra) has been found to improve the clinical manifestations. First-line therapy for SS is based on NSAIDs, anti-histamines and steroids [5], usually with unsatisfactory results, even if a combination therapy with hydroxychloroquine and low dose steroids has recently been used with an excellent response [6].

Our case highlights two rare features: the presence of monoclonal gammopathy composed of IgM and IgA types, with κ -light chain, only described in other four cases [2, 6], and an osteosclerotic lesion situated on the seventh right rib, considered as an atypical site compared with the more common ones (pelvis, femur and tibia).

In accordance with the different monoclonal components associated to chronic vasculitic urticaria and the systemic symptoms reported in literature, similar to those observed in SS, we suggest extending the definition of Schnitzler Syndrome to all these cases. ■

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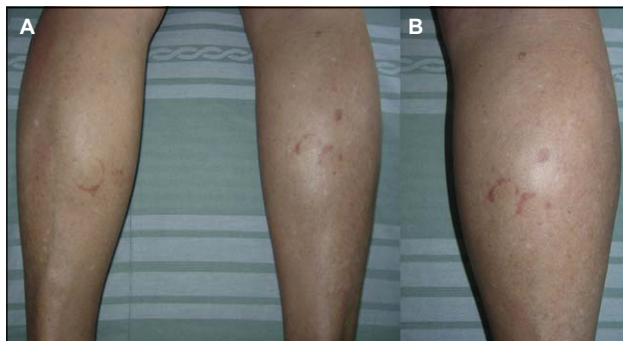


Figure 1. A) Erythematous circinate lesions on the posterior side of both legs. B) Detail of the right leg lesion.

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