

Schnitzler's Syndrome (Urticaria and Macroglobulinemia): Evolution to Waldenström's Disease Is Not Uncommon

Sir,

The association of chronic urticaria with monoclonal IgM was described by Schnitzler et al. in 1974 (1). In the first case described, and in those subsequently reported, a benign evolution without evolution toward a lymphoproliferative disease was claimed. We here report the follow-up of a previously described case (2) with a 7-year history of monoclonal gammopathy and a 10-year history of urticaria with evolution to Waldenström's disease.

CASE REPORT

A 69-year-old man had a 5-year history of chronic urticaria and a 2-year history of monoclonal IgM dysproteinemia. Clinical examination showed annular erythematous and maculopapular non-pruritic lesions over the trunk. There was no lymph node enlargement and no hepatomegaly or splenomegaly. Laboratory data have earlier been reported (2). Because of the high IgM spike and the presence of double dysproteinemia, a close follow-up was instituted. Monoclonal IgM increased gradually to 58 g/l within 5 years.

A further bone marrow biopsy in January 1995 revealed overt infiltration by lymphoplasmacytoid cells (72%) without osteosclerosis. Blood cell count was normal except for mild anemia (hemoglobin 112 g/l). Chlorambucil (2 mg daily) was started and gradually increased to 6 mg daily. Six months later the patient complained of painful paresthesia of the lower limbs. Electric nerve conduction was decreased and no cryoglobulin was revealed despite repeated examination. Though no anti-myelin-associated glycoprotein activity was found in the serum of the patient, a series of 15 plasma exchanges was instituted, without improvement of neurological symptoms and without improvement of electric nerve conduction. Monoclonal IgM was decreased (20.2 g/l) but urticaria still persisted.

DISCUSSION

Our patient initially had all the signs usually encountered in Schnitzler's syndrome: chronic urticaria, macroglobulinemia and osteocondensation, without any evidence of lymphoproliferative disease at the beginning of his disease. Evolution to Waldenström's disease occurred 5 years after the diagnosis of Schnitzler's syndrome was made, and 7 years after the discovery of monoclonal gammopathy.

The benign and prolonged evolution of dysproteinemia in Schnitzler's syndrome has always been emphasized, since the first patient described had a 23-year follow-up (3). Since its original description 22 years ago, 7 out of 33 patients suffering from Schnitzler's syndrome have developed lymphoproliferative disease: Waldenström's disease (5 cases, including the present case) (3–5), lymphoplasmacytoid lymphoma (6) and IgM myeloma (7), one case each.

Schnitzler's syndrome seems therefore to require a close follow-up. In addition the absence of signs of lymphoprolifer-

ative disease for initial diagnosis of Schnitzler's syndrome may be inappropriate.

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