

## [Abstract 22]

### **AN ADDITIONAL EXAMPLE OF THE DIVERSITY OF THE MANIFESTATIONS ASSOCIATED WITH MACROGLOBULINEMIA: THE SCHNITZLER'S SYNDROME.**

**JP Femand, Hospital Saint Louis, Paris, FRANCE.**

The clinical manifestations associated with Waldenström's macroglobulinemia may be related to tumour infiltration and to the amount of the monoclonal component. Various symptoms may also be consecutive to specific properties of the IgM, such as auto-antibody activity, propensity to cryoprecipitate or to form amyloid fibrils, which may translate into symptoms either intra-vascular (cryo-globulinemia, cold agglutination disease) or consecutive to IgM deposition in various tissues such as the peripheral nerve or the kidney.

Other mechanisms may be involved. Thus, absorption to malignant cells of C1q esterase inhibitor and von Willebrand factor were documented in a few patients with WM and acquired angioedema and with MW and von Willebrand syndrome, respectively. The production by clonal cells or their environment of biological mediators such as cytokines may also be implicated leading, for example, to hyper-eosinophilia or disease-related biological inflammatory symptoms.

Although this is speculative, this mechanism may be implicated in the so called Schnitzler's syndrome which associates a monoclonal IgM gammopathy and urticarial skin lesions. In addition, the syndrome is frequently featured by intermittent fever, asthenia, arthralgia and bone pain, with imaging evidence of osteosclerosis. Biological findings usually show elevated white blood cell count and elevation of erythrocyte sedimentation rate, which reflects both an inflammatory biological syndrome and the presence of the monoclonal IgM. Skin histological lesions may be featured by urticarial vasculitis. The physio-pathological link between urticaria, systemic symptoms, hyperostosis and the monoclonal B-cell proliferation is unknown. Accordingly, present treatment is only symptomatic and unsatisfactory. We observed a dramatic and sustained improvement of urticarial and systemic manifestations of Schnitzler's syndrome in a patient who was treated by pefloxacin for an urinary infection. This prompted us to study the efficacy of the antibiotic in 10 other consecutive patients. We will report this experience and speculate on its physio-pathological implications.