SKIN MANIFESTATIONS OF INTERNAL DISEASE

# VASCULITIS AND IGA MONOCLONAL GAMMOPATHY OF CUTANEOUS SIGNIFICANCE

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Background: Monoclonal gammopathy of cutaneous significance is a subgroup of monoclonal gammopathy with skin disease, without myeloma or lymphoproliferative disorder. We report 3 patients with vasculitis and IgA monoclonal gammopathy of cutaneous significance.

Observation: Three men, 38, 37 and 50 years old, had a vascular purpura for more than 10 years. One patient had inflammatory arthralgia while none of them had renal, digestive or neurologic involvement. In all three cases, a serum monoclonal IgAκ could be detected. Skin biopsy showed leucocytoclasic vasculitis with notable neutrophilic infiltrate contrasting with limited fibrinoid necrosis of small vessels' wall, and small vessel focal thrombosis in one case. Skin vascular deposits of IgA and C3 using direct immunofluorescence were found in all cases. Monoclonal IgAκ vascular deposits was positive in two cases. Colchicine treatment (1mg/day, 3 months) was inefficient in 2 cases. The patient with the most severe skin symptoms did not achieve remission using 8 different lines of treatment (colchicine, systemic steroids, dapsone, methotrexate, azathioprine, intravenous immunoglobulins, cyclophosphamide, plasma exchanges). Salvage treatment using rituximab induced complete remission, systemic steroid weaning and complete disappearance of the IgA monoclonal gammopathy. Rituximab maintenance infusions were given every year allowing complete disease remission with 2.5 years of follow up.

Conclusions: Nineteen patients with IgA monoclonal gammopathy of renal significance (glomerular nephropathy with possible renal failure) have been recently published. Other organs could be involved: skin (n=5), digestive tract (n=1) and lung (n=1). Monotypic IgA

deposit was found in 14/19 cases. Twelve cases of IgA vasculitis treated with rituximab have been reported with a good efficacy. Relapsing cutaneous IgA vasculitis may evoke a vasculitis associated with IgA monoclonal gammopathy of cutaneous significance. Rituximab efficiency in one patient should be confirmed in future studies.