

Dapsone for the treatment of chronic IgA vasculitis (Henoch-Schonlein)

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Dear Editor,

IgA vasculitis (IgAV; Henoch-Schonlein) in adults is a rare disease belonging to the category of immune complex small-vessel vasculitides.^{1,2} Patients present with purpura, nephritis, arthritis and/or abdominal pain. A chronic form of IgAV occurs predominantly in patients with isolated cutaneous involvement.³ The aetiology of this chronic form of IgAV is unknown. In 40% of patients a trigger is identified such as infections, drugs, toxins and/or malignancies.^{3,5} Furthermore, IgAV can be associated with other diseases such as liver disease, inflammatory bowel disease, and/or ankylosing spondylitis. Treatment of IgAV with internal organ involvement consists mainly of corticosteroids. Unfortunately, this is ineffective for the chronic cutaneous form of IgAV.^{3,6}

A 21-year-old woman presented to the outpatient department of internal medicine because of chronic skin lesions of the legs. The lesions had already been present for two years. This was a symmetrical, non-pruritic rash which dominated on the extremities and which was influenced by orthostasis. Her medical history showed recurrent urinary tract infections. Apart from oral contraceptives she was not on any medications. On physical examination no abnormalities were found besides a non-palpable skin rash of the legs (*figure 1*). A skin biopsy was taken which showed a leukocytoclastic vasculitis with depositions of IgA and C₃. A diagnosis of chronic adult IgAV was made. Further laboratory investigations showed a C-reactive protein level of 1 mg/l, erythrocyte sedimentation rate of 7 mm/hour, glomerular filtration rate >90 ml/min, serum IgA of 2.13 g/l (normal level 0.7-4.0 g/l), absence of cryoglobulins, ANA 1:80, anti-dsDNA negative, ENA negative, ANCA negative, normal complement (C₃ 1.18 g/l, C₄ 0.15 g/l), HIV negative, hepatitis B negative, hepatitis C negative, EBV and CMV IgM negative. Urine examination disclosed no erythrocytes or proteinuria. An ultrasound of the heart did not show any signs suggestive of endocarditis. Treatment with dapsone 100 mg once daily was started.

Figure 1. Henoch-Schonlein vasculitis



Within 24 hours the skin lesions diminished and totally disappeared within a week. During follow-up (12 months), the patient remained asymptomatic with dapsone 50 mg as maintenance therapy.

In conclusion, dapsone was found to be very effective in treating chronic leukocytoclastic vasculitis in our patient with IgAV. The use of dapsone should be advocated in such patients with chronic adult IgAV. The exact mechanism how dapsone works, is poorly understood. Unfortunately, the skin lesions frequently relapse after stopping dapsone and renal involvement does not respond to this treatment.⁷⁻¹⁰

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