PEDIATRIC RHEUMATOLOGY

MEETING ABSTRACT Open Access

P03-015 - Dapson treats chronic Pupura Schoenlein (PSH)

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Introduction

PSH is common and, with few exceptions, resolves spontaneously. In complicated cases treatment can be difficult.

Case Report

Here we present an 11 year old Iranian girl, of healthy parents suffered from chronic PSH with chronic exanthema, severe ulcerative dermatitis and arthralgia. Clinical observation: no fever, no arthritis, no signs of polyserositis or hepatosplenomegaly; Laboratory: normal values (full blood count, CRP, complement (C3, C4), no autoantibodies (ANA, ANCA, anti-ds-DNA), no cryoglobulins, zinc, IgG, IgM). Abnormal findings: highly elevated IgA in serum (maximum 2030 mg/dl), elevation of ESR and MRP8/ 14. Skin biopsy: leukocytoclastic infiltrates in subcutaneous fatty tissue. Immunohistochemistry: IgA deposits in tip of papillae and upper corium. No mutation in Marenostrin was found. Multimodal therapeutic approaches (Cortisone, Methotrexate, Azathioprine, Colchicine, i.v. Immunoglobulins) remained without success for 8 years. With the administration of Dapsone symptoms resolved within days and remain under control for > 8 months now. Met-Hb level is tolerable.

Discussion

Anti-inflammatory potency of dapsone is illustrated. Therapeutic efficacy of Dapsone has been reported in chronic PSH[1,2], but the mechanism remains to be fully elucidated. Hypothesis: Endothelial cells and hyperreactive B-cells (as illustrated by IgA elevation) secrete IL-8. IL-8 is elevated in patients with PSH [3]. IL-8 stimulates perivascular invasion by neutrophils. Dapsone can inhibit secretion of IL-8 [4], thereby impairing neutrophil function [5-7].

In conclusion Dapsone might be benefical for complicated cases of PSH. The mechanism of its anti-inflammatory potency remains to be elucidated.

Competing interests
None Declared.

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