Vasculitis Simulating Eczematous Dermatitis Due to C2 Deficiency

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In a patient with total C2 deficiency and repeated respiratory infections a papulovesicular rash showed to be due to leukocytoclastic vasculitis. The patient's brother also had a total deficiency of C2 but no clinical symptoms. HLA-investigation in both him and the proband showed the genotype commonly reported in cases with homozygous C2 deficiency. Their heterozygous mother and sister had slightly decreased C2 levels. *Key words: Complement deficiency; C2; Vasculitis.* (Received December 9, 1986.)

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Selective, genetically determined deficiency of the second component of the complement system (C2) has previously been reported both in connection with different disease manifestations and in healthy individuals (1, 2). There is a well-known linkage between inherited C2 deficiency and inflammatory collagen disorders such as rheumatoid arthritis and systemic lupus erythematosus (3).

Cases with vasculitis sharing clinical or pathogenetic features with SLE due to C2 deficiency have also been presented including patients with anaphylactoid purpura and fulminant necrotizing vasculitis with cutaneous ulcers (2, 4, 5, 6, 7, 8).

In this report we present a case of total C2 deficiency with mild cutaneous vasculitis producing papulovesicular lesions without ulceration and purpura.

CASE REPORT

The patient is a woman born in 1958 with a left sided ptosis, which has been surgically corrected. As a child she suffered from repeated upper respiratory infections and pneumonia. She also had a nephropathy between 3 and 6 years of age with proteinuria and erythrocytes and granular casts in the urine. Since 1982 she has had recurrent symptoms of rhino-conjunctivitis. Routine prick testing has not demonstrated any allergy. From 1984 she has again had repeated upper respiratory infections.

In January 1984, itching skin lesions appeared on the elbows, around the infragluteal folds and on the ears. With successive spreading of the small papulovesicles the skin disease eventually engaged the arms, hands and lower part of the trunk. The patient has had no abdominal or urinary symptoms since childhood. In the beginning of 1985 a total absence of the second component of the complement system was disclosed. Treatment with topical steroids groups III and IV was tried with uncertain effect of short duration. After 4 months of peroral treatment with oxychloroquine (Plaquenil®) 400 mg daily there was a remarkable improvement.

Laboratory investigations

The erythrocyte sedimentation rate has varied between 2 and 22 mm/h. The haemoglobin value as well as the platelet count has been normal. White cell counts have shown slightly increased values. Serum electrophoresis has revealed an increased haptoglobin up to 4.4 g/l, but there has been no elevation of immunoglobulins. Serum IgE showed a value of 40 kU/l. There has been no occult intestinal bleeding and no albuminuria and urine sediment has been normal. Antibodies to ANA, DNA, ENA or anti-RO antibodies have not been found. The patient had no circulating antibodies to gliadin or reticulin.

Histological investigations

Three punch biopsies from the skin were taken on separate occasions. All of them showed the same picture with slightly hyperkeratotic epidermis without spongiosis. Around blood vessels in the upper

Table I. *C2 level and HLA types in proband and her close relatives*

	C2 in % of a normal standard	HLA type
Proband	0	A 25; B 18; Cw 6?; DR 2, x
Mother	66	A 2, 25; B 15, 18; Cw 3, w6?; DR 2, 4
Sister	63	A 2, 25; B 15, 18; Cw 3, w6?; DR 2, 4
Brother	0	A 25; B 18; Cw 6?; DR 2, x

dermis there were cellular infiltrates consisting mainly of neutrophils with few mononuclear cells and eosinophils. There were scattered nuclear fragments in the infiltrates. Direct immunofluorescense investigation of a skin biopsy showed deposition of C3 and fibrinogen in the basement membrane zone and around small dermal vessels, but there was no deposition of IgA.

Complement and HLA-investigation in the proband and her close relatives (Table I)

The patient's father died several years ago in an accident. In the proband no total haemolytic complement activity was observed. Her C3 was 200%, C4 310%, C1Q 110% and C1S 162% of a normal standard. C2 was not detectible. The patient had the HLA-type A 25; B 18; Cw 6?; DR 2, x. Her mother and sister were found to be heterozygous for the A 25; B 18; DR 2 haplotype and had both slightly decreased C2-levels. None of the proband's close relatives showed reduced levels of C3 or C4. Her brother however showed a total deficiency of C2, though at the time of the investigation he remained in full health. He as well as the proband was homozygous for HLA A 25; B 18 and probably DR 2 antigens. Their haplotype corresponds to the one commonly reported in cases with inherited C2 deficiency (2).

DISCUSSION

The proband's repeated respiratory infections during childhood and since 1984 are probably to some degree dependent on her C2 deficiency. C2 deficiency associated with recurrent infections has previously been reported (9, 10).

It is interesting to know that the proband's skin manifestations exacerbated in connection with upper respiratory infections. Rendall et al. has described a patient with C2 deficiency and leukocytoclastic vasculitis with the same correlation between skin symptoms and episodes of infections (6). One similar case with chronic vasculitis was described by Friend et al. (11).

The exacerbations and remissions of pruritic papulovesicular skin lesions initially gave the impression of an eczematous dermatitis. The type and localization aroused suspicion of an atopic disease. However, the patient did not fulfil the criterias for the diagnosis of atopic dermatitis (13).

Histological investigation showed leukocytoclastic vasculitis. It is not common that leukocytoclastic vasculitis produces only papulovesicles, but it occurs in some individuals (12).

One patient's leukocytoclastic vasculitis was exclusively cutaneous without clinical or laboratory signs of engagement of any other organ. Neither clinically nor serologically our patient had lupus erythematosus of LE-like disease.

In earlier described cases of vasculitis combined with C2 deficiency the clinical picture has been more dramatic with palpable purpura, ulcerations of the skin or systemic vasculitis (4, 5, 6, 7, 8). Our patient shows that C2 deficiency also may present as a mild vasculitis of the skin.

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Relationship of Dietary Gluten Intake to Dapsone Dose in Dermatitis herpetiformis

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Mobacken H, Andersson H, Dahlberg E, Kastrup W. Relationship of dietary gluten intake to dapsone dose in dermatitis herpetiformis. Acta Derm Venereol (Stockh) 1987; 67: 267–270.

The gluten intake was quantitated utilizing a dietary history method in 43 patients with dermatitis herpetiformis on non-restricted diet. The mean daily gluten intake was 15 g. The individual intake of gluten was related to the maintenance dose of dapsone. It was significantly higher in patients on 100–150 mg dapsone daily than in those taking 0–25 mg daily. There was a significant correlation between amount of gluten in the diet and the dapsone dose ($p<0.01$, $r_S=0.43$). Villous atrophy was not related to the dapsone dose. It is suggested that the gluten-sensitive enteropathy changes the intestinal permeability and thus contributes to the development of blisters. *Key word: Dapsone treatment.* (Received August 15, 1986.)

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Virtually all patients with dermatitis herpetiformis (DH) have a coeliac-like enteropathy (1). The skin lesions are also gluten-dependent and clear following a gluten-free diet (2).