oxygen. Application of a stabilized hydrogen peroxide cream induced delayed tanning on the pressure sites, while the placebo-treated contralateral sites remained unpigmented.

The results of this study show that the hydrogen peroxide-containing cream has biological effects on intact normal skin, and that the effects of pressure on delayed pigmentation by UVA will be normalized by an extra percutaneous H_2O_2 supply. This supports the assumption that oxygen is responsible for the induction of delayed pigmentation by UVA (1, 2). This is in contrast to pigmentation caused by UVB and PUVA, which seems to be independent of the presence of oxygen in the tissue at the time of exposure (5).

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Subtotal C4 Deficiency and SLE-like Disease

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Irestedt M, Svensson Å, Månsson T. Subtotal C4 deficiency and SLE-like disease. Acta Derm Venereol (Stockh) 1986; 66: 67–70.

Extremely low C4 values were found in a 65-year-old man with relapsing arthritis and skin lesions of many years duration of the scalp, face, hands and feet together with painful ulcerations of the toes and fingers. The discovery was made during an exacerbation, but the deficiency of C4 persisted in repeated controls after remission. The clinical findings in connection with these low C4 values are in congruence with the diagnosis of inherited deficiency of C4. *Key words: Complement; Genetic deficiency; IgM.* (Received June 18, 1985.)

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Hereditary deficiency of all components of the classical pathway of complement activation may occur (1). It has been shown that deficiency of C1, C2 and C4 is related to immune complex diseases similar to SLE (1). Both complete and partial genetic C4 deficiency are rare conditions but have been described in connection with immune complex diseases (3, 4). The present report is of a further case of subtotal C4 deficiency with a SLE-like disease.

CASE HISTORY

A man born in 1919 suffered during his teens from arthralgias. Since about 1970 he has recurrent episodes of angina pectoris and in 1977 a myocardial infarction was diagnosed. For at least 10 years he has had skin lesions. In the scalp there has been scarring alopecia and purple lesions with telangiecta-

sias accompanied by porcelain white scarring have appeared on the distal dorsal parts of the fingers and around the nails. Exacerbations of tender and sometimes ulcerated infiltrations localized to the fingertips, palms, heels, toes and knees have occurred. Arthralgias of the fingers and toes and pains in the wrists, the knees and the ankles have appeared sometimes with objective signs of arthritis. Treatment with oxychloroquine (Plaquenil®) has resulted in marked relief of the symptoms.

Family data

The patient is number 4 of 7 siblings. An older brother has polyarthritis with high titers of ANA and the youngest sister has scleroderma and persistent partial C4 deficiency.

Laboratory investigations

During a period of 10 years the erythrocyte sedimentation rate has varied between 10 and 30 mm/h. The hemoglobin value has been normal. On one occasion his white blood cell count showed a slight leucopenia. The platelet count has on two separate occasions shown subnormal values of 87 respectively 95×10^9/l. Repeated bone marrow examinations have revealed normal results.

In 1978 it was discovered that the patient had a monoclonal abnormal gammaglobulin of IgM type. The concentration has increased from 8 g/l to 19 g/l. Urine electrophoresis has been normal.

The serological investigation disclosed antibodies to RNP in a titer of 1/800, but rheumatoid factor, ANA, antibodies to DNA and anti-Ro antibodies have not been found.

Complement investigation during an exacerbation in the spring of 1984 showed an extremely low value of C4: 3% of the normal standard (reference value 53–207%) C2 was 62% (reference value 75–163%). C3, C1Q, C1S and C1R as well as C1 esterase inhibitor showed normal values. There was no dysfunction of the C1 esterase inhibitor. The M-komponent showed no interaction with C4. Repeated complement investigations during 14 months have shown persistent low C4 values, 1–2% of the normal standard, even during and after clinical remission.

Microscopic examination of a punch biopsy from a dorsal lesion of the left ring-finger showed a hyperkeratotic epidermis with hydropic degeneration of the basal cells and in the papillary dermis clusters of lymphocytic cells. Direct immunofluorescence examination of a punch biopsy from normal skin from the lower back revealed a granular lupus band reaction along the dermo-epidermal junction with IgG, C3 and fibrinogen.

X-ray investigation of the skull, columna, pelvis, right and left femur as well as right and left humerus showed no destructions.

DISCUSSION

It is well known, that SLE-patients with exacerbations nearly always have decreased levels of the early complement factors above all C3 and C4 and there is some correlation between reduced levels and the severity of the disease. Our patient has normal values of C3 but extremely low C4 values persisting in repeated controls in spite of clinical remission. This speaks against a consumption of complement factors. The patient has no C1 esterase inhibitor deficiency or dysfunction, which may be rarer causes of low C4 (5). His M-komponent of IgM-type could not be shown having autoantibody properties against C4, which has been reported in another case (6). These data reveal that the patient has a hereditary C4 deficiency. The finding of partial C4 deficiency in his sister with scleroderma speaks in favour of this.

The patient's C2 value was slightly decreased, which has been observed in other C4 deficient patients and is believed to be due to the close linkage of the structural genes for the two proteins (7).

The production of C4 is controlled by two distinct but very closely linked loci localized to the short arm of chromosome 6 (8). There may be carriers for one, two, three or four "silent" genes. If all four loci are "silent" the clinical result is a total deficiency of C4 (7, 8, 9, 10, 11).

In heterozygous individuals the level of C4 varies between very low and subnormal (3, 4, 7). It has been suggested that the C4 value reflects the number of expressed C4 genes (9). According to this it must be presumed that our patient has three "silent" genes.

Patients with total absence of C4 are in most cases affected by a SLE like disease, but with no or low titer of ANA (1), even if some patients have had rather high titers of ANA (11, 12). Many individuals with partial deficiency of C4 escape immune complex disease but some develop a picture of systemic and some discoid lupus erythematosus. ANA may be found but not in all cases (3, 4).

Our patient has a disease affecting several organs: skin manifestations, trombocytopenia on two separate occasions and polyarthritis. His skin lesions of the hands and feet are of the type seen in both discoid and systemic lupus erythematosus. His disease cannot be classified as a systemic lupus erythematosus according to the ARA criteria (13). A strong evidence of the diagnosis of SLE, however, is that he has a positive lupus band test by direct immunofluorescence investigation of uninvolved skin of the lower back (14). The only positive autoantibody finding in our patient is a low titer of antibodies to RNP.

Elevation of IgM has been described in earlier cases of C4 deficiency (7, 11, 15). In one patient with homozygous deficiency of C4 a high polyclonal IgM-value was found (10). Our patient differs from these earlier cases in having an increasingly high IgM-fraction of monoclonal type. The investigation has not shown any signs of myeloma. The relevance of the high monoclonal IgM-value and its connexion to the C4 deficiency remains obscure.

ACKNOWLEDGEMENTS

We are grateful to the Immunological Department of the Bacteriological Central Laboratory of Lund for performing the complement investigations and to Björn Dahlbäck, Department of Clinical Chemistry, Malmö General Hospital, for the analysis of the IgM-complement interaction.

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IgE-mediated Anaphylaxis to Mustard

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Widström L, Johansson SGO. IgE-mediated anaphylaxis to mustard. Acta Derm Venereol (Stockh) 1986; 66: 70–71.

A young woman had recurrent urticaria and angioneurotic edema following ingestion of mustard and mayonnaise. IgE-mediated allergy to mustard seeds and seeds of botanically related plants was confirmed by RAST. *Key words: Urticaria; Crusiferae; RAST.* (Received June 20, 1985.)

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In 1980 Panconesi (1) described a case of anaphylactic shock following ingestion of pizza. Allergy to mustard was proven by intradermal skin tests and RAST (2) to extracts of mustard seeds. Contamination of the pizza with mustard was suspected.

CASE REPORT

A 25-year-old woman came to us in March 1984, because of recurrent urticaria. As a child she had had severe atopic dermatitis and reacted with rhinitis and swelling of the throat when eating fish or egg. Now her dermatitis has almost cleared and she can eat both fish and eggs without trouble. She still gets rhinitis, when exposed to cats or dogs.

From August 1981, to February 1984, she had had six episodes of acute severe generalized urticaria and angioneurotic edema of the face and the neck. On all occasions except the last one she had ingested either mustard or mayonnaise just before the appearance of the symptoms. The last attack of urticaria occurred after eating a hot dog with ketchup purchased from a hot dog stand in the street.

Allergy to mustard was suspected. Besides eggs, oil and vinegar mustard is usually an ingredient of mayonnaise. At the last attack the patient had not consciously eaten mustard or mayonnaise but it is very likely that the hot dog was contaminated with mustard from the utensils used in the hot dog stand. In Sweden, at least, hot dogs are usually eaten with large amounts of mustard and there must therefore be a great chance of contamination.

Mustard is made from the seeds of two plants of the Crusiferae family, namely *Brassica nigra*, black mustard, and *Brassica alba*, white mustard. Closely related to mustard are several vegetables such as cabbage, (*B. olerata capitata*), rutabaga (*B. napobrassica*) and rape (*B. napus*).

As Panconesi's patient developed an anaphylactic reaction when skin tested, we decided to analyse for possible IgE antibodies to mustard by RAST. A blood sample for immunological tests was drawn in March 1984, using the Phadebas IgE PRIST (Pharmacia Diagnostics, Uppsala), the total IgE concentration was found to be 350 kU/l.

Extracts of ground seeds of *B. nigra*, *B. alba*, *B. napus* and eatable parts of various