Section of Dermatology

President S C Gold MD

Meeting 21 June 1973

Cases

Ulcerative Colitis, Myasthenia Gravis, Atypical Lichen Planus, Alopecia Areata, Vitiligo

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B L, man aged 38

History: Ulcerative colitis since 1957, confirmed by sigmoidoscopy and rectal biopsy. Barium enema showed involvement of the whole colon. Atypical lichen planus on the trunk since 1960, confirmed by skin histopathology. In the last four years the skin lesions have been atrophic. Myasthenia gravis developed in March 1969 with an excellent response to intravenous edrophonium. Three months later a severe exacerbation of myasthenia occurred which required artificial ventilation for one month, and for which he was given corticosteroids. During this time he suffered episodes of hypotension and coma; he developed a staphylococcal septicæmia with lung abscesses and the ulcerative colitis flared up. Since then he has had little trouble from myasthenia or ulcerative colitis and is controlled on prednisone 10 mg daily, codeine phosphate 30 mg three times daily, and chlordiazepoxide 5 mg once daily. He was presented to the Clinical Section of the Royal Society of Medicine in December 1970 and subsequently reported in the *Proceedings* (Miller 1971). A year later alopecia areata developed. It has extended rapidly and now only a few white hairs remain. In April 1973 LE cells were found in moderate numbers and his spleen became palpable. One month later vitiligo was noticed, probably unmasked by the recent sunshine.

On examination: No evidence of myasthenia gravis. Well circumscribed oval patches on the trunk with slight central atrophy and lichenoid margin (Fig 1). Capillaritis, atrophie blanche and necrotic ulcer on lower leg. Extensive alopecia areata, widespread vitiligo, and a slightly enlarged spleen.

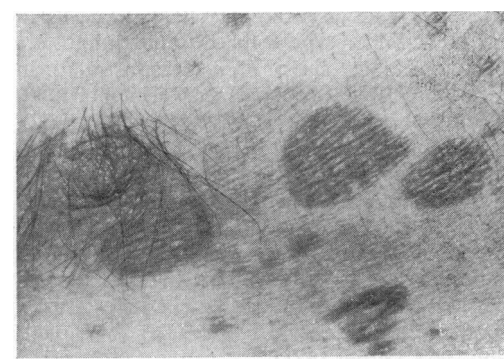

Fig 1 *Oval slightly atrophic areas on front of chest*

Investigations: Skin biopsy (1961) from abdomen (Fig 2): saw toothing, basal cell liquefaction, band-like lymphocytic infiltrate in upper dermis, prominent stratum granulosum; changes of lichen planus. Skin biopsy (1973) from abdomen: atrophic epidermis, slight fibrosis of papillary dermis with mild chronic inflammatory cell infiltrate; consistent with late stage of lichen planus. LE cells: 1969 negative, 1973 positive in moderate numbers. Autoantibody screen: smooth muscle 1969 and 1970 positive, 1973 negative; striated muscle 1969 positive, 1973 negative.

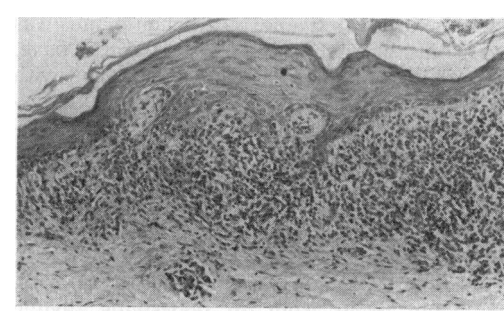

Fig 2 *Band-like lymphocytic infiltrate, basal cell liquefaction degeneration*

Thyroid colloid and microsomes, gastric parietal cells, mitochondria, and antinuclear factor persistently negative. Immunoglobulins: IgM 20 mg/100 ml (normal 47–170); IgA and IgG normal. X-rays: tomograms 1969 and thoracic inlet 1972 – no thymic tumour shown.

The following tests were negative or normal: blood film and ESR; plasma proteins and electrophoresis; lymphocyte function tests (phytohæmagglutinin, mixed lymphocyte reaction, candida antigen); Rose Waaler, latex RA; thyroid and adrenal function tests; muscle biopsy.

Comment

Many distinct conditions have arisen in this patient in the last sixteen years: 1957, ulcerative colitis; 1960, lichen planus; 1969, myasthenia gravis; 1971, alopecia areata; 1973, LE cells in moderate numbers, splenomegaly and vitiligo.

Autoantibodies have been demonstrated in ulcerative colitis (Broberger & Perlmann 1959) and myasthenia gravis (Beutner *et al.* 1962). These diseases are presumed by some to be manifestations of autoimmunity. Myasthenia gravis has been associated with other such diseases (Simpson 1964). Alopecia areata is likewise thought by some to be of the same nature, and has been associated with the organ-specific autoimmune diseases (Stankler & Bewsher 1972), ulcerative colitis and vitiligo (Muller & Winkelmann 1963, Lerner 1971).

The nature of the skin eruption is debatable. The diagnosis of lichen planus was made on the histopathology, as clinically it was atypical. If this is lichen planus, then its close temporal association with this group of conditions may be of etiological significance. Lichen planus has been remarkably free of reported associations with other conditions whether dermatological or internal. Copeman *et al.* (1970) described 4 cases in which it was impossible clinically or histopathologically to make a firm diagnosis between lichen planus or lupus erythematosus (LE). All 4 subsequently progressed to LE. In many respects this case is similar. The dense band-like lymphocytic infiltration with basal cell liquefaction on which the original diagnosis of lichen planus was made could have been due to LE. LE cells have, however, only appeared recently combined with splenomegaly. The association of LE with myasthenia gravis and ulcerative colitis is also recognized (Alarcón-Segovia *et al.* 1963, White & Marshall 1962).

The association of these separate disorders is not necessarily fortuitous and there may be some basic defect in the patient's immunological status, whereby multiple autoimmune conditions arise. This could result from an underlying thymic abnormality. The thymus, according to Burnet (1962), is important in preventing formation of autoantibodies; it is well recognized that 60–70% of cases of myasthenia gravis are associated with thymic abnormalities (White & Marshall 1962). It may be that at a future date a thymoma will develop. Conversely, the remarkable improvement in the myasthenia since the severe exacerbation four years ago could have been the result of a 'medical thymectomy', precipitated by the many different stresses suffered at that time.

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Thymoma, Acquired Hypogammaglobulinæmia, Lichen Planus, Alopecia Areata

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F H, man aged 67

History: Transient ptosis in 1959 – no cause found. Thymoma discovered in 1965. Thymectomy performed. Histology: well-differentiated lymphoeptheliomatous tumour with a good fibrous capsule. Onset of bilateral basal bronchiectasis in 1967, confirmed by chest X-ray and bronchogram, which has progressed with frequent pneumonic complications. Oral lichen planus appeared a year later associated with nail destruction and lesions on the trunk. The oral lesions became erosive in 1969 and have persisted with large areas of tongue frequently involved (Fig 1). Low levels of gamma globulins were first noticed in 1971 and the levels have continued to fall. Alopecia areata developed in 1972. Recently, *Trichophyton mentagrophytes* has been isolated from his groins and feet. The patient has reacted to transfusions of whole blood and reconstituted pooled dried plasma, but is able to tolerate fresh frozen plasma. Weight loss 25 kg in the last three years.

Treatment: 1971–72: systemic steroids for oral erosive lichen planus, often in large doses. Since February 1973: regular plasma exchange transfusions every 3–4 weeks using a continuous flow