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Dr J M Beare: This case illustrates the spectrum of these conditions, ranging from fulminating dermatomyositis at one end to chronic benign morphœa at the other.

Dr H R Vickers: Some Members may remember a patient referred by Dr H T Calvert and shown at the Annual Meeting of the British Association of Dermatology in Oxford in 1966. He had very extensive morphœa and eventually died from peritonitis. Post-mortem examination showed no evidence of systemic sclerosis in any of his internal organs.

Dr H T Calvert: I do not consider that the patient referred to by Dr Vickers is similar to the one now under discussion. He was an example of generalized malignant morphœa where the patient goes steadily downhill and dies before the stage of natural remission can set in, without showing any systemic involvement by scleroderma. If he were re-investigated today we would not find laboratory abnormalities as revealed here. Today's patient is probably an example of mixed connective tissue disease (Sharp *et al.* 1972, *American Journal of Medicine* **52**, 148; *British Medical Journal*, 1972, iv, 315) – opposed to the triad of scleroderma, dermatomyositis and lupus erythematosus.

Professor F F Hellier: Ordinary morphœa usually disappears without showing permanent changes though some forms, such as the 'coup de sabre' type, may leave atrophy. This patient has myositis in addition to the skin changes which might lead to residual atrophy even if the superficial lesions clear up.

Dr K D Crow: I agree with Dr Calvert that there is a lethal form of generalized morphœa in which, at autopsy, no systemic lesions are discovered. I have had one such case.

The following cases were also presented:

Pseudoxanthoma Elasticum
Dr J D Boxley (for Dr P F D Naylor)
Nævus of Ota
Dr J D Boxley
(for Dr P F D Naylor and Mr P K B Davis)
Acute Febrile Neutrophilic Dermatosis
Dr W Morison (for Dr E L Rhodes)

Dr Andrew Warin (*St Thomas' Hospital, London SE1*) presented a short paper entitled **Skin Lesions in Gonococcæmia.**

Meeting 21 December 1972

Cases

Dapsone-resistant Leprosy

P S Friedmann MRCP (for D I Williams FRCP)
(*King's College Hospital, London SE5*)

K B, woman aged 48

History: From Guyana but living in UK for 15 years. Developed painless papules which had gradually enlarged on face, arms and legs over the last 4–5 months. On direct questioning she denied having any previous illness or taking any form of medication. Only on her third visit, when confronted with the diagnosis, did she admit to having had Hansen's disease at age 13 and having taken dapsone (sent from Guyana) ever since.

On examination: Firm, slightly translucent nodule on right cheek and smaller papules on side of nose, left eyebrow, both forearms and calves. No thickened nerves and no neurological deficit. Histopathology: multiple sharply defined non-caseating epithelioid granulomas in dermis around small neurovascular bundles. In Ziehl-Neelsen stain vast numbers of acid-fast bacilli were present in granulomas and throughout dermis.

Discussion

This patient admitted taking dapsone 50 mg twice weekly for at least 20 years and before that to anti-leprosy treatment under the supervision of the local hospital in Guyana. Despite this and in the absence of intercurrent illness or other factors which might alter immunological balance, nodules of leprosy towards the lepromatous end of the spectrum have appeared and it is logical to conclude that dapsone resistance has developed.

Dr Stanley Browne: The history of dapsone self-medication (perhaps irregular or intermittent) over many years and the recent subacute clinical exacerbation, together with the returning presence of a high proportion of morphologically normal *M. leprœ* in the bacilliferous skin lesions, are very suggestive of relapse of near-lepromatous leprosy occurring as the result of the emergence of drug-resistant organisms. The long residence in England and the wilfully misleading history should not obscure the possibility of leprosy. The papules are typical (though not pathognomonic) of clinical relapse: they are small, fleshy and of no special distribution.

If dapsone is being taken regularly and in adequate dosage, and is being absorbed (plasma levels and urine

examination would confirm), a *prima facie* assumption of drug resistance is justified. Confirmation is obtained by recourse to the elegant mouse footpad inoculation technique, the mice being given different dietary drug concentrations – 0.1, 0.001 and 0.0001%. Resistance develops in a stepwise fashion, and cross-resistance (e.g. between sulphones and sulphonamides, and between thiambutosine and thiacetazone) may occur. Fortunately, all patients hitherto found with dapsone-resistant bacilli have responded to either clofazimine or Rifampicin.

Juvenile Dermatomyositis

P S Friedmann MRCP (for R H Marten FRCP)
(*King's College Hospital, London SE5*)

A W, boy aged 14½

History: March 1972: Developed rash around eyes and raised patches on dorsa of hands and knuckles, extensor aspect of both elbows, fronts of knees.

On examination (June 1972): Heliotrope discoloration mainly below eyes with periorbital œdema. Purple papules over knuckles, elbows and knees. Musculoskeletal examination: nothing abnormal detected.

Investigations: Creatine phosphokinase (CPK) 230 iu/l. (normal <130 iu/l.). EMG compatible with myositis. Hb 13 g/100 ml. ESR 18 mm in 1 h (Westergren). ANF negative.

Progress and treatment: By October 1972 muscle weakness with flexion deformities at elbows and difficulty in rising from chair and in swallowing had developed, despite prednisolone 10 mg alternate days, increased after 2 weeks to 20 mg alternate days.

CPK 252 iu/l. Barium swallow: barium spilled into trachea after poor swallowing action. ECG: inverted T wave in leads V_{1-3}. Prednisolone was increased to 40 mg daily. Within 10 days dysphagia and ECG changes had improved. Two months later his strength was increasing and the skin rash fading. CPK had reverted to normal (Fig 1).

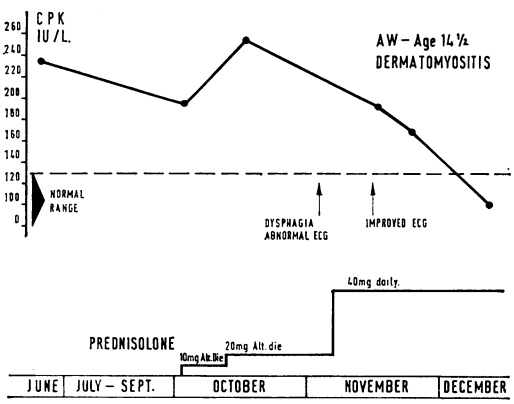

Fig 1 *CPK levels and clinical features responding to prednisolone*

Discussion

In this patient with a typical rash, myositis was detectable from CPK and EMG months before it became clinically apparent. This moderately severe case has improved surprisingly well on a modest dose of steroids, and the CPK has settled more than the clinical improvement might suggest.

Dr R H Marten: It should be added that at this stage prognosis must be guarded.

Ichthyosis Linearis Circumflexa and Netherton's Syndrome with Idiopathic Dwarfism

J E B Adamson MB MRCP
(for R H Marten FRCP)
(*King's College Hospital, London SE5*)

L C, girl aged 10

History: Seen at the age of 8 days with a 6-day history of an erythematous rash which had started on the face and spread rapidly to involve the whole body surface. On examination at that time she was noted to be bright red with exfoliating areas on the limbs, particularly around the flexures. She attended intermittently over the next few years because of an eczematous rash and was also under the care of the pædiatric department for investigation of her small stature. She had made normal progress until 10 months old. Since then her weight and height had been below the 3rd percentile. She is now growing at her own percentile. No definite cause for this has been found. In autumn 1972 she reattended for scaling on the scalp and a rash on her arms and legs.

On examination: Noticeably short and thin for her age. Arcuate and erythematous lesions on legs and arms with marginal scaling. The lesions showed a distinctive double-edged scale caused by superficial peeling at both edges of the serpiginous erythematous band. Her scalp was scaly. Scalp hairs showed pili torti while eyebrow hairs showed trichorrhexis invaginata.

Investigations: Urinary amino acids normal. X-ray of wrist: bone age 6 years 10 months (chronological age 10 years 5 months).

Skin biopsy: 'The epidermis is hyperkeratotic and acanthotic with prominent rete processes. Foci of parakeratosis are present. There is a very minor degree of suprabasal epidermal spongiosis.