DEPIGMENTING ERYTHEMA MULTIFORME
(A Clinical and Histopathological Study)

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Summary

Two patients with a peculiar variant of erythema multiforme are described. The classical iris lesions appearing seasonally in response to as yet unidentifiable agents resolved leaving behind persistent depigmentation. Although, the EM lesions aborted on corticosteroid therapy, the latter seemed to have no effect on the depigmented spots. The histopathological features were characteristic of EM. The depigmentation perhaps is the result of a permanent damage to the melanocytes. This variant of EM may be tentatively designated as depigmenting erythema multiforme-DEM.

Erythema multiforme-EM is a distinct clinicopathological reaction which can be precipitated by a variety of agents. The clinical manifestations are variable and different morphological forms of lesions are known to display a distinct tissue reaction[1]. The most characteristic lesion that draws attention is identifiable as "target" or "iris" lesion composed of a clear red area at the periphery that surrounds a pale pink zone and a central livid area[2]. The lesions usually fade within a few weeks leaving behind faintly hyperpigmented or brownish stains which may persist longer. In rare instances, however, the lesions have resulted in depigmentation[3,4] which disappear following treatment with oral prednisolone[4]. The present report pertains to 2 cases of erythema multiforme manifesting with classical iris lesions resulting on healing in persistent spots of depigmentation.

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Case Report

Case 1: A 24 years old lady complained of recurrent urticarial papular and target lesions on the extremities and trunk of 9 years' duration. She started in March 1969 with sudden wheals and target lesions on the back of the trunk and the dorsal aspects of the hands and the feet. The lesions were mildly itchy, persisted for a month and disappeared after some treatment leaving faintly hypopigmented areas. Over the next 4 years she had similar lesions at the onset of summer (March-April) every year. In April 1973 she was first seen by the author when she was put on 20 mg of prednisolone daily for 2 weeks. The lesions aborted but depigmented areas persisted. She had since been experiencing successive crops of new lesions during the same season and when last seen in March 1978, admitted that for the last 2 years she had started having lesions even at the time of onset of winter (Oct-Nov). The lesions showed a tendency to disappear on their

own but aborted faster whenever she took prednisolone. Each crop of the lesions resulted in new depigmented spots which persisted inspite of treatment. The patient was otherwise healthy and there was no history of prior drug intake, mucosal lesions or constitutional symptoms.

Examination showed multiple erythematous papular, target and urticarial lesions on the extremities and the trunk. Irregular areas of speckled depigmentation with interspersed normal skin were present at the sites of old healed lesions (Fig. 1). The face and the mucosal surfaces were free of lesions. Systemic examination did not reveal any abnormality. Routine investigations on the blood and urine were normal. A skin biopsy was obtained from a fresh target lesion on the trunk.

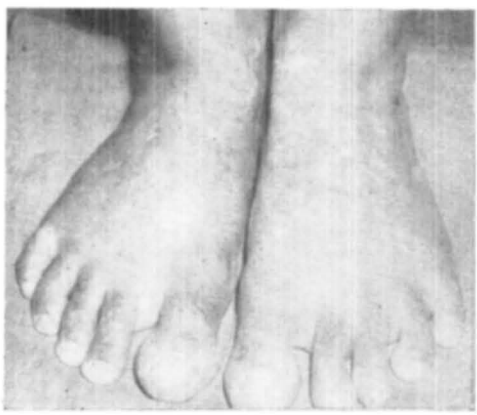

Fig. 1 Healed lesions dorsa of the feet (case 1)

Case 2 : A 20 years old man presented in May 1978 with recurrent crops of itchy erythematous papular and iris lesions on the extremities and the face of 2 months' duration. The lesions first appeared as red targets on the forehead and the extensor aspects of the elbows. In a week's time the central area became depigmented with redness persisting peripherally. New lesions appeared on the face and the exposed parts of the chest and disappeared leaving behind areas of depigmentation with peripheral hyperpigmentation. There was no history of prior drug intake, mucosal lesions or constitutional symptoms. He was put on 20 mg of prednisolone daily for 4 weeks and during the follow up period of next 6 months he has not had any fresh lesions but depigmented areas have persisted.

Examination revealed a young man showing erythematous papular and iris lesions on the extensor of the elbows, chest and the face. Depigmented spots surrounded by hyperpigmentation were seen at the sites of old healed lesions (Fig. 2). The mucous membranes were free and the systemic examination did not reveal any abnormality. A biopsy was taken from a fresh iris lesion on the elbow.

Fig. 2 Healed lesions on face with central depigmentation and peripheral hyperpigmentation (case 2)

Histopathological Features

Biopsies of early iris lesions from both cases revealed identical histological features. The epidermis showed moderate spongiosis intracellular edema and focal areas of basal cell degeneration. There was a dense perivascular inflammatory infiltrate composed of lymphocytes and histiocytes in the

corium. Variable amount of melanin pigment was seen in the upper dermis corresponding to the epidermal areas of basal cell necrosis. The latter was most marked in the central part of the sections whereas the dermal inflammation was most conspicuous in the peripheral part of the lesion where the basal cell layer appeared more or less normal (Fig. 3).

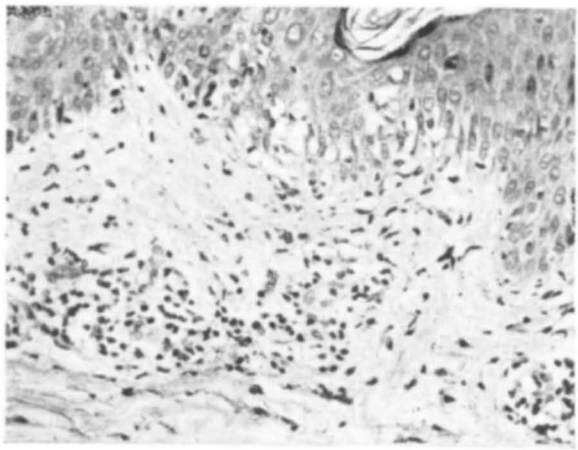

Fig. 3 Showing focal basal cell neerosis and pigment incontinence (H & E × 110)

Comment

The causative agennts attributable to EM are of extremely diverse nature. In some patients the mere avoidance or eradication of the cause leads to cure following a self limiting course. But in many the causative factor defies identification or the attempt at eradication proves unsuccessful and the disease runs a rather prolonged course in successive crops of lesions with relapses and remissions. The seasonal attacks in some cases are well known without any clue to the cause. A percentage of recurrent cases have been associated with and attributed to the accompanying herpes simplex[5],[6]. In still others mycoplasma infection is held responsible[7] but a majority remains idiopathic.

In the patients presented herein the diagnosis was based on clinical characteristics in the form of iris lesions distributed over favourable sites and histopathological tissue reaction consistent with EM. The cause however could not be defined. Historically, there was a relation to seasonal appearance of the lesions. In case 2 there was also some suggestion as if the lesions were localized to photosensitive areas of the body. A number of cases have been observed to display such distribution of lesions but one is not clear whether these are the usual sites of predeliction in EM without any relation to sunlight. What appears more interesting in these cases is the observation that the lesions on healing leave behind depigmentation. There is sufficient clinical evidence as if the central portion of iris lesion becomes depigmented on healing and the peripheral red zone either returns to normal colour or becomes slightly hyperpigmented. Histologically the basal cell necrosis most marked in the central part appears to be attended by pigment incontinence. The resultant depigmentation rather than a central darkening is however, most intriguing. It is possible that in a variety of EM in addition to the focal areas of basal cell necrosis the bystander melanocytes also undergo a degenerati,e change resulting in permanent depigmentation. The histochemical and electron microscopic investigation of the depigmented lesions may prove fruitful. These patients have however not been as lucky as those described by Handa et al[4] where complete repigmentation could be achieved with oral prednisolone.

Whether the expression of EM in these patients is related to a viral infection or a systemic disease is not clear at this stage. Laboratory investigations for lupus erythematosus in case 1 have been negative on one occasion. The

presentation is not that of vitiligo preceded by some inflammatory component. The clinical characteristics and evolution of the lesions and no family history of vitiligo in both cases makes it highly unlikely to be a form of vitiligo. The exact nature of the disease remains unsolved and at most it may be regarded as a depigmenting variant of EM.

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Announcement...

International Dermatopathology Symposium

The 2nd International Dermatopathology Symposium, entitled "Dilemmas and directions in differential diagnosis" will be held at Grosvenor House Hotel, Park Lane, London 12-15 July 1981. The symposium is co-organized by Prof. E. Wilson Jones and Dr. Martin Black of the Institute of Dermatology, University of London, and Dr. A. Bernard Ackerman of the Section of Dermatopathology, New York University School of Medicine. The emphasis will be on histologic clues to diagnosis by conventional microscopy, clinical, electron microscopic and immunopathological aspects of inflammations, neoplasms, malformations and deposits of abnormal materials in the skin.

All enquiries should be addressed to : Marcus Summersfield, London Symposium 1981, Conference Co-ordinates, Regent House, 60 King Street, Twickenham, Middlesex TW1 3SH, with the exception of specific enquiries regarding the medical aspects of the programme which should be addressed to: Prof. E. Wilson Jones, Institute of Dermatology, St. John's Hospital for Diseases of the Skin, Lisle Street, Leicester Square, London WC2H 7BJ.

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