DERMATOLOGICAL DISORDERS RESEMBLING LEPROSY

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ABSTRACT

Three dermatological conditions — epidermolysis bullosa dystrophica (EBD), granuloma multiforme (GM) and mycosis fungoides (MF) were diagnosed elsewhere as leprosy either clinically or histologically. Although the morphology of the lesions were suspicious of leprosy there were few striking clinical findings which were unfavourable. Leprosy is still an important disease that should not be missed. However, the recognition of these skin disorders is highlighted so that unnecessary and prolonged treatment for leprosy can be avoided in endemic countries.

Key Words: Leprosy, Epidermolysis bullosa dystrophica, Pseudosyndactaly, Granuloma multiforme, Mycosis Fungoides.

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INTRODUCTION

Leprosy, once regarded as a worst scourge inflicting untold miseries to mankind is fast becoming a fascinating disease for the clinicians, immunologists and experimental pathologists. Hither-to segregated and disposed of into the asylums of no hope and bleak future — leprosoria, the sufferers of this often dreaded disease are now able to regain the lost or injured human pride and dignity, credit to the scientific innovations which have either reduced or prevented the complications and offered them salvation. In our endeavour not to miss a case of leprosy, we should not relinquish the pursuit for a definite diagnosis also, as the consequence of a hasty decision will be a life — long stigma. Such a problem is encountered in areas where this disease is common. There are several skin disorders that should be considered in the differential diagnosis of leprosy (1, 2). The author faced three such cases, two of which GM and EBD were on dapsone at presentation and the other, MF — a T-cell lymphoma had contradictory clinical findings and histopathological interpretation. GM, exclusively found in Africa is reported here for its rarity in this part of the world. Granuloma annulare (GA) and GM are within a spectrum of palisade granulomatous disorders with overlapping characteristics and their distinction from leprosy is discussed.

CASE REPORTS

Case 1

An 11-year old Chinese boy from a rural area, on dapsone therapy, was referred in June 1980 with deformities of the hands and feet for confirmation of leprosy. He was of small stature but of normal intelligence. The most striking clinical feature was psuedosyndactaly — fusion of web spaces and fingers and toes bound within an envelope of scar tissue (Figure 1). Joints of the fingers were

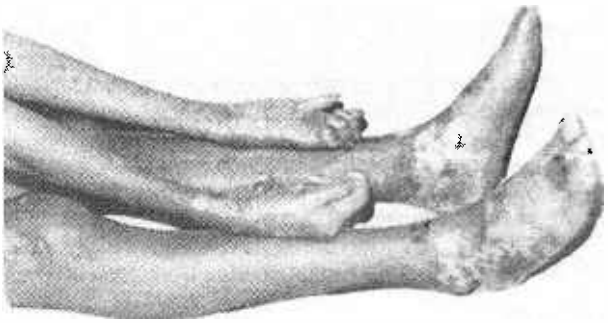

Figure 1: EBD with pseudosyndactaly and atrophic scars.

flexed and thumbs were adducted. Nail dystrophy, wasting of the intrinsic muscles of the hands and feet and thin 'papery' scars over the extremities, elbows and knee were also noted. There was no alopacia, bullae, dysphagia or impairment of sensation. He was born, after an uneventful pregnancy, at term, with bullae, on othermise normal skin which appeared in early infancy. Repeated cycles of blistering triggered by pressure, trauma and hot weather and subsequent scarring resulted in gradual encasement of the extremities by an epidermal 'cocoon'. His parents were unaffected and of non-consanguinous marriage. A male and a female siblings out of five had died during early childhood of similar disease. The clinical presentation was most convincing of EBD, probably a recessive type.

Within two weeks of cessation of treatment, few tense blisters appeared on the body and back of elbows. Skin biopsy showed a large subepidermal bulla with scanty acute inflammatory cells. Electron microscopic studies to find out the site of cleavage could not be done. He was given prednisolone and then referred for reconstructive surgery. Leprophobia, deeply ingrained in the parents was allayed.

Case 2

A 46-year old Chinese male was seen in April 1983 with polymorphic rashes over the body of 2 years dura-

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tion. He was seen at another centre and was started on dapsone when a skin biopsy was reported as boderline tuberculoid (BT) leprosy. He presented with large circinate lesions with raised erythematous border and normal-looking centre over the extensor aspects of the forearm (Figure 2). Small, annular lesions were seen in the flexor regions (Figure 3) and elevated plaque-like lesions were found over the abdomen (Figure 4). The trunk was scattered with multiple small and oval-shaped papules which were pruritic. Touch, pain and temperature sensations over the affected parts were intact and the corresponding superficial nerves were not enlarged. There was no wasting or weakness of muscles. Slit skin smear for acid fast bacilli (AFB) was negative.

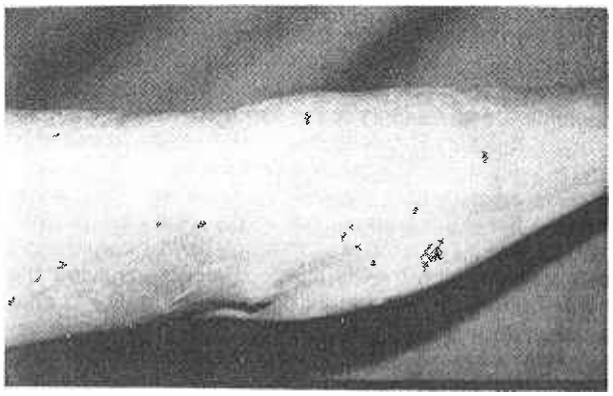

Figure 2: GM: Circinate lesions with raised border over the forearm.

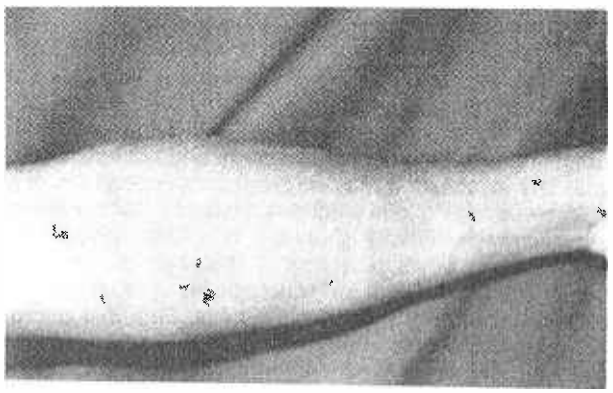

Figure 3: Annular lesions with normal centre over the flexor aspect.

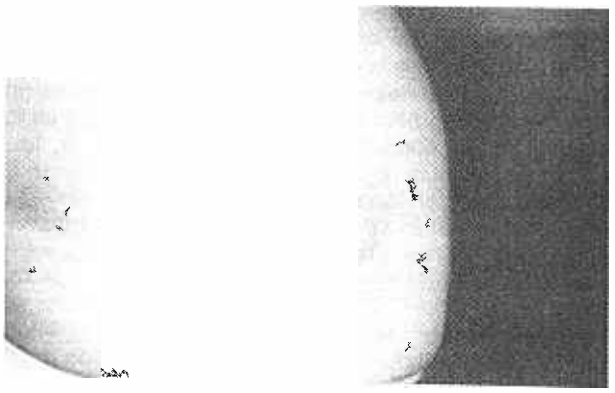

Figure 4: Plaques over the side of the abdomen in GM.

Clinically he was not a case of leprosy but considered to have a variant of GA. A skin biopsy was done which showed multiple non-caseating granulomas in the dermis consisting of epithelioid cells, Langhans' giant cells and lymphocytes (Figure 5,6). The appendages were surrounded by mild infiltrate. On special staining, there was focal excess of mucin, little phagocytosis of elastic fragments by giant cells and macrophages in the granuloma but there was no collagen degeneration. It resembled GM. Patient was reassured and the fear of leprosy was dispelled. He was treated with topical and intralesional steroid. The lesions regressed considerably but did not disappear completely during the 5-year follow-up period.

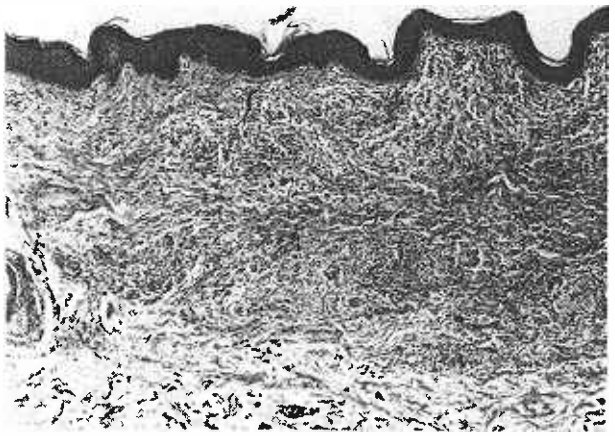

Figure 5: GM: Prominent multiple granuloma scattered away from the nerve bundle (H & E x 100).

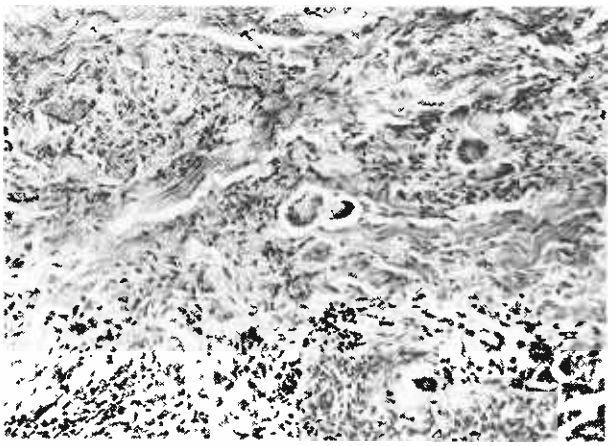

Figure 6: Multinucleated giant cells with phagocytosis of elastic fibres with chronic inflammatory cells (x 400).

Case 3

A 50-year old Malay male who was advised to seek treatment for leprosy was seen in April 1987 with multiple nodules of different sizes (Figure 7) which appeared in rapid succession over a period of 2 months. There was no history of preceding rash, erythema or associated constitutional symptoms. The nodules were concentrated more on the trunk with a large ulcerated one on the forehead. They were pink, firm, bright, mobile and a few were 'juicy' (Figure 7). The ear-lobes and eye-brows were normal. Axillary lymph nodes were enlarged, the superficial nerves were non-tender and there was no area of loss sensation. Spleen and liver were not palpable. The clinical features were in favour of lymphoma.

On investigation, haemoglobulin 12g/dl, ESR 52mm, packed cell volume 37%, urine for Bence Jones protein negative, smear for AFB negative; full blood picture and peripheral smear were normal with no blast cells. Skin biopsy from a representative lesion showed a massive infiltrate of lymphocytes in the dermis separated from the normal epidermis by a clear grenz zone. The infiltrate was in patchy distribution around the appendages which were completely destroyed. There was absence of histiocytes, granuloma or abnormal cells. In view of the massive infiltrate and cell type, it was reported that lymphocytoma

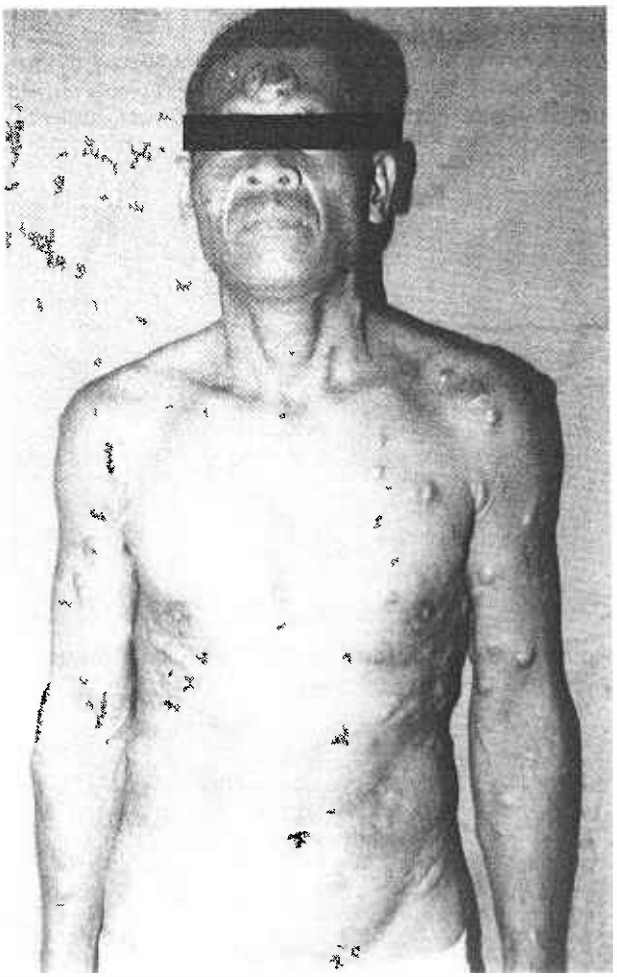

Figure 7: MF: Multiple nodules; ulcerated one on the forehead.

cutis could be considered. The clinical course of the disease was certainly not of benign nature but most likely that of MF, tumeur d'emblee variant. To rule out B cell lymphoma lymph node biopsy, bone marrow trephine and immunoglobulin studies were considered, but the patient, against our advice, did not comply and later did not survive.

DISCUSSION

The three patients described here presented with deformities of the limbs, annular lesions and nodules of the skin respectively which are also seen in different types of leprosy. Leprosy is classified into lepromatous type (LL) on one end of the spectrum with lack of cell-mediated immunity, tuberculoid (TT) on the other end with good immumo-responsiveness and an intermediate or borderline one (BB) which is further split into borderline lepromatous (BL) and borderline tuberculoid (BT) (3). The clinical features of these various types are distinct (4), but a certain degree of overlap may be encountered, thus avoiding diagnostic error which can be brought about when an isolated feature is taken as evidence and other findings are not corroborated. The three cardinal signs of leprosy are:

a. Impairment or loss of sensation.
b. Enlargement or tenderness of the superficial nerve corresponding to the skin involvement.
c. Demonstration of AFB in slit skin smear.

In tuberculoid leprosy (a) and (b) are seen early and unilateral and (c) practically not possible; in LL (c) is an early finding and (a) and (b) occur in advanced stage in bilaterally symmetrical distribution.

In case 1, the family history, pseudosyndactaly and presence of sensation are supportive of EBD which belongs to a heterogenous group of genetically determined blistering disorders in children-EB involving the skin and mucosa (5). Although bilateral deformities can occur in advanced LL, fusion of fingers and toes within scar tissue is not a feature of leprosy. With better health-care and effective chemotherapy against leprosy available at present in most developing countries, such debilitating deformities should be the events of the past.

GM was first described by Leiker et al in 1964 in Mkar area of Northern Nigeria while reviewing several hundreds of patients who did not respond to dapsone since their skin lesions simulated tuberculoid leprosy (6). Later it was discovered in Eastern Nigeria, Kenya, Zaire (7) and Indonesia (2). GM is a clinico-pathologically distinct form of GA in these parts of Africa. GA, GM and necrobiosis lipoidica are all variants of the same process: a granulomatous reaction in the dermis with damage to collagen and elastic fibres caused by unknown injury (8). In case 2, there were distinct features: no impairment of sensation, no nerve involvement and the lesions were pruritic. Histologically BT leprosy is characterized by epitheloid cell granuloma in the dermis, commonly involving the nerve bundle resulting in its destruction (9). GA is characterised by a histiocytic granuloma in palisade arrangement around a central focus of mucin scattered in the dermis around degenerating collagen bundles (8). Nerves are not involved. But in GM elastic tissue is reduced and the giant cells contain elastic fragments (7, 8). These features were noted in this patient who is the first case of GM in a Malaysian Chinese.

The clinical course of case 3 was rapid and fatal. Although few noduled were 'succulent' the lesions were large and firm. Nodules in LL are highly bacilliferous. MF — a misnomer, is a cutaneous T cell lymphoma with fungating growths in advanced stage and not associated with any mycoses. It may evolve from a premycotic erythematous rash to form plaques and tumours with visceral involment (11). Tumour d'emble'e is a variant of MF in which lesions develop de novo and carries a poor prognosis. The hall mark of MF histologically is the presence of accumulation of mononuclear cells, Pautriers microabscess in the epidermis. But, in certain phase, especially tumour stage predominant non-epidermo-trophoic picture with dense dermal infiltrate of mono or pleomorphic cells (12) may be seen beneath a grenz zone which was the feature in this patient. Lymphocytoma cutis or pseudolymphoma of Spiegler-Fendt is a benign lymphoproliferative disease characterised by papules and nodules predominantly occurring on the face. It may mimick leprosy but the heavy lympho-histiocytic infiltrate in follicular arrangement in the dermis should enable us to distinguish this condition from MF and BT or BL (9, 13).

In conclusion, it is emphasized that leprosy is a treatable condition like any other infectious diseases with few variations in the management (14) and early detection, prompt treatment and better follow up will ensure a good cure rate and thereby break the transmission cycle. A high index of suspicion should be aroused in endemic areas, but before the final decision is made it is often rewarding to remember the advice, 'if there is one diagnosis that should not be established unless there is absolute certainty, it is that of leprosy' (15).

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