THE CLINICAL COURSE OF DIMORPHOUS MACULAR LEPROSY IN THE BELGIAN CONGO

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The two polar types of leprosy—the lepromatous and the tuberculoid—have been more or less precisely described, but there remains a not inconsiderable group of cases in which the cutaneous lesions are "atypical lepromatous" or "atypical tuberculoid," or share some of the features of both groups. The clinical appearances and evolution of this "dimorphous" form of the disease may vary from country to country, just as its relative frequency may vary, and its epidemiologic importance.

The skin lesions of dimorphous macular leprosy in the tropical rain-forest of the Belgian Congo seem to possess characteristic features which serve to distinguish them from kinds of macules that have been long defined, i.e., the indeterminate, the maculoanesthetic, and the lepromatous. Khanolkar and Cochrane (4) suggested that the commonest macule was an indeterminate one embodying some maculoanesthetic features, and that these macules should be called "dimorphous." Ryrie (6) had earlier recognized these macular lesions in Nigeria.

Relevant factors in the development of this form of leprosy in central Africa may be the combination of constant, fairly high temperatures and high relative humidity (with little seasonal variation), leading to great activity of the skin and its appendages. Another factor may be the tendency of deeply-pigmented skin to over-react to stimuli causing pigmentary changes and subcutaneous fibrosis.

This paper concerns 62 cases of dimorphous macular leprosy at present in the Yalisombo Leprosarium in Oriental Province of the Belgian Congo. These 62 cases constitute over 10 per cent of the total number of patients (mainly "open" cases) isolated there for special reasons. In the domiciliary leprosy service, well-marked typical dimorphous macular cases constitute 3.2 per cent of 5,349 cases of leprosy under treatment.

To complete the picture, it may be stated that lepromatous cases account for 20.6 per cent of all cases under treatment, tuberculoid cases for 72.7 per cent and indeterminate cases for 3.5 per cent. Many cases of tuberculoid leprosy are no longer under treatment, and many others have undergone spontaneous retrogression without treatment. The very high endemicity of the disease is indicated by the fact that 11.9 per cent of the total population was under treatment for leprosy in 1957.

Systematic observation of these 62 dimorphous macular cases over the years suggests that the group possesses certain characteristics in common, especially as regards the macular manifestations. Some of these are exemplified by four case photographs which illustrate this paper (Figs. 1-4).

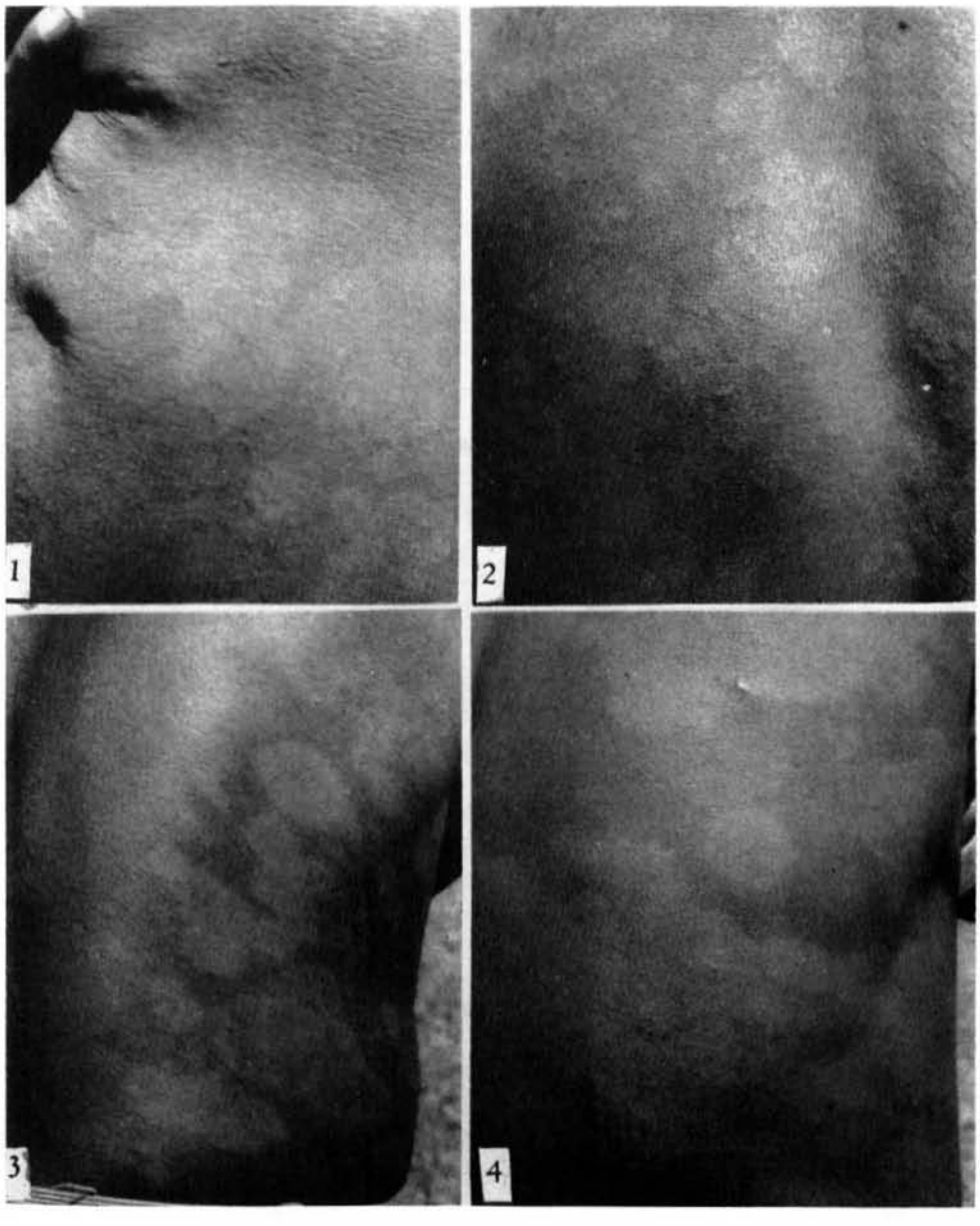
Regular examination at quarterly intervals of material obtained from six sites of the body by the scraped-incision method as suggested by Wade (7) and Zanetti (8), and the calculation of the bacteriologic index (3, 4), confirm the contention of Davison (2) and of MacDonald (5) and others that many cases which on clinical grounds should be initially and constantly noninfectious do indeed constitute for long periods a perhaps little-recognized source for the dissemination of *M. leprae*.

In the diagnosis of dimorphous macular leprosy, the modifications in the clinical appearances of other, dissimilar lesions induced by treatment must constantly be borne in mind. Thus, a tuberculoid lesion may quite rapidly lose its papular raised edge, and the differentiation between normal skin and the lesion may be one mainly, if not entirely, of pigmentation; and, on the other hand, a lepromatous macule may become more defined as it heals and may eventually come to resemble a dimorphous macular lesion, or an atypical tuberculoid one. It is possible, too, that these lepromatous macules developed originally from dimorphous lesions, and that as the lepromatous element regressed under therapy, the residual dimorphous element became more obvious.

In the last resort, it is the concomitance of two or more "incompatibles" that suggests the diagnosis of dimorphous macular leprosy, especially and most frequently the presence of well-defined, regularly hypopigmented macules that are highly positive bacteriologically. In those cases the transition from normally-pigmented skin to the uniformly hypopigmented skin of the macule is so abrupt that, on clinical grounds, the lesion could not be termed lepromatous.

While there may be some variation in the clinical course and manifestations of dimorphous macular leprosy in the Belgian Congo, where in any one district the degree of cutaneous pigmentation may show a wide range from one individual to another, there is discernible a typical natural history to which the majority of cases conform.

The first sign is the sudden appearance, without premonitory symptoms, of a large, well-defined, irregularly circular hypopigmented macule on the lumbar region, the scapular region, or elsewhere, as the following frequency distribution indicates: lumbar region, 17; scapular region, 8; arm, 7; abdomen, 6; thighs, 6; forearm, 4; buttocks, 4; chest, 3; forehead, 3; hand, 2; face, 1; foot, 1. Total 62. This distribution is similar to that of the early lesions of tuberculoid leprosy in the district concerned. If there is more than one macule initially, the lesions are usually confined to one area of skin.



DESCRIPTION OF PLATE

Cases of dimorphous macular leprosy described, showing primary and secondary macules, with colonial lesions. Regarding bacteriologic findings:

FIG. 1. Bacilli in the lesions of this case were scanty, and no globi were present.

FIGS. 2 and 3. These cases were highly positive bacteriologically, with globi in every field of the oil-immersion objective.

FIG. 4. This case was less highly positive, but many globi were found in smears from the nasal mucosa.

The border of the macule is quite clear-cut from the beginning, and this distinction is maintained, or becomes even better marked with time. The lesion is completely flat, and there are no papillary elevations anywhere—on or near the edge, or towards the center—and none make their appearance subsequently. There is usually no change in the initial size of the macule, although sometimes there is a tendency towards general centrifugal extension during the months following its appearance by the engulfing of colonial, or satellite, macules which have meanwhile developed. These colonial macules are a very typical accompaniment of the dimorphous macules, and occur at some time or other near many of the primary macules and also in relation to some of the secondary macules (see below).

The macule is uniformly hypopigmented, of a dull yellowish-brown color on a dark brown skin. The pigmentary changes usually persist under treatment, and there may even be further loss of pigment.

The colonial macules may be single or multiple. These are typically small (0.5-1.0 cm. in diameter), and are separated by a narrow band of healthy skin from the primary macule. Their borders are usually not so well-marked as those of the primary macule; in fact, when seen isolated, these colonial macules might well be mistaken for lepromatous lesions.

After a variable period, reckoned in months rather than in years, there appears a more or less widespread eruption of secondary lesions in which the individual elements are small and of similar appearance. These are of three kinds:

1. Resembling the parent macule (although smaller) in its hypopigmentation, its well-defined edge, and its flat, featureless surface;
2. Resembling the colonial macules near the original macule, i.e., small, with vague, fluffy edges, and diffuse hypopigmentation, more marked centrally than peripherally;
3. Resembling small minor tuberculoid lesions in its slight elevation, slightly raised borders, dry surface, and irregular hypopigmentation.

Further crops of lesions may make their appearance thereafter. Those lesions arising at any one time resemble each other, but may differ in definition and in pigmentation from the preexisting lesions or from those that may develop subsequently. Thus, at the time of the clinical examination, in addition to the well-defined primary macule, which may give the impression of a tuberculoid lesion healing spontaneously or under treatment, there may be macules that are almost typically tuberculoid and others that are almost typically lepromatous.

An infrequent diagnostic aid concerns the elevation of the pseudotuberculoid lesions: the highest point on their surface is not at the edge, or just within an advancing hypopigmented edge, but rather towards the center.

Colonial macules may also arise in connection with these secondary

lesions, often to be engulfed in turn by the centrifugal spread of the secondary macules.

The pigmentary variations are interesting to observe. Thus, the earlier lesions on the anterior aspect of the trunk are often well-defined, quite flat and uniformly hypopigmented, whereas the later lesions on the scapular and lumbar regions are less well-defined, very slightly raised, and reddish-coppery in color. Not only may there be distinct differences in pigmentation and in definition between lesions on the anterior and those on the posterior aspects of the trunk, but in individual cases there may be differences not less well-marked between, for example, the buttocks and the scapular region, or between the scapular and the lumbar regions. In some few cases, macules may disappear spontaneously or as the result of treatment on one aspect of the trunk and persist on another.

Neurologic examinations of the macules.—The neurologic findings in the primary macule resemble those in tuberculoid leprosy, i.e., progressive loss of tactile sense, reduction in thermal appreciation, reduction in sweating, and loss of hair.

In the case of secondary macules, the findings resemble those in lepromatous leprosy, i.e., slight loss of tactile sense demonstrable with a wisp of cotton wool, no loss of thermal sense to ordinary methods of examination, hairs and sweating unaffected.

General neurologic complications.—Such complications, including trophic effects without marked paralysis or anesthesia, were seen in 35 out of the 50 bacteriologically positive cases, and in 8 out of the 12 bacteriologically negative cases (70% and 67%, respectively). These proportions, however, may be higher than those obtaining in patients under domiciliary treatment, since cases with trophic lesions tended to gravitate to the leprosarium for rehabilitation.

These findings serve to emphasize the importance of the clinical observation that acute edematous reactions in nerve trunks, even following standard doses of sulfones, are of frequent occurrence and of serious prognosis.

Bacteriologic examination.—At the earliest stage of the condition, material obtained by the scraped-incision method from the most active edge of the primary macules, and from all other parts of the skin (e.g., the earlobe, forehead, cheek, apparently normal skin, and nasal mucosa) is, by standard methods of examination, free from acid-fast bacilli.

At some point of time in the majority of cases (50 out of the 62 on which this description is based), the primary lesion becomes highly positive by standard methods, and *M. leprae,* singly or in globi, may be found in its clinically active border. In those cases, and often at approximately the same time, the nasal mucosa, the earlobes, and perhaps other parts of the skin, also become bacteriologically positive.

Thus, the primary macule, which may by this time resemble a healing tuberculoid lesion in the definition of its edge, may be highly positive, and also the nasal mucosa.

The colonial macules, as they appear, may be highly positive, whether they are near the primary macule or the secondary macules. The successive crops of hyperemic, infiltrated, slightly hypopigmented lesions, slightly dome-shaped, and widely distributed over the skin, are usually very highly positive with numerous globi in every field of the oil-immersion objective. At this time, too, smears from the nasal mucosa usually, and the earlobe frequently, contain numerous globi.

These findings invalidate the previous classification of these cases, on clinical grounds, as "reactional tuberculoid." Clinically active tuberculoid cases, with raised, infiltrated, hyperemic, succulent borders, may—in this district—pass through a phase, of variable length and possibly recurrent, in which numerous bacilli and even globi may be found by standard methods of examination (*"poussées bacillifères"* of French authors), but the nasal mucosa is invariably negative in those cases.

It is interesting to observe that in dimorphous macular cases material from the nasal mucosa gives positive bacteriologic results more frequently than material from the most active portion of the macule, or from the earlobe. Thus, in 31 out of the 50 bacteriologically positive cases in my series, globi were present in the nasal mucosa, and all the remaining 19 cases had bacilli but no globi. Material obtained from the macule itself contained leprosy bacilli in all cases but 2 (i.e., in 48); in 12 cases the numbers of them were very small, however, and in only 19 cases were globi present. Material from the earlobes showed globi in 18 cases, and bacilli in a further 25 cases, in 11 of which they were scanty.

In 42 of the 50 cases the nasal mucosa was the last site to become bacteriologically negative to standard methods of examination.

The 12 cases in which *M. leprae* were rarely or never found constituted a homogeneous group with puzzling features. They could not be distinguished on clinical grounds from the larger group of 50 cases which were highly positive bacteriologically. The aspect of the individual elements in this small group of cases was generally lepromatous, and the distribution of the macular eruption was invariably of lepromatous habit. In only 3 cases of the 12, however, were scanty bacilli ever found on repeated examination, and no globi were ever seen in any of the sites examined.

Histopathology.—Biopsy specimens taken from representative areas of the macular lesions at different stages, revealed a great variety of histologic features, ranging from scattered groups of shrunken "foamy cells" to slight cellular infiltration of the corium, diffuse and nonspecific.

Quite frequently there was a focally distributed tuberculoid infiltrate, with epithelioid and rare giant cells, but the subepidermal zone was respected in a manner typical of lepromatous lesions. Nerves were sometimes invaded with lymphocytic cells, and in other cases acid-fast bacilli or granules were to be seen between the nerve fibers. In some sections, only one nerve among several would be invaded with round cells.

Pari passu with the clinical changes, the histologic appearance sometimes progressed from "atypical tuberculoid" to "resolving lepromatous" in successive biopsies made at intervals of several months.

SUMMARY

The clinical features of the development of dimorphous macular leprosy as seen in a series of 62 cases in the Belgian Congo are reviewed, together with some of the bacteriologic data of those cases.

Routine examination is advocated, of material obtained by the scraped-incision method, of all cases of leprosy, especially of those showing atypical features.

The epidemiologic and neurologic importance of dimorphous macular leprosy in central Africa is briefly emphasized.

RESUMEN

Repásanse las características clínicas de la aparición de la lepra macular dimorfa, tal como fueron observadas en una serie de 62 casos en el Congo Belga, junto con algunos de los datos bacteriológicos de dichos casos.

Abógase por el examen sistemático del material obtenido con la técnica de la incisión-raspadura, en todos los casos de lepra, y sobre todo en los que revelen características atípicas.

Recálcase la importancia epidemiológica y neurológica que reviste la lepra macular dimorfa en el Africa Central.

ACKNOWLEDGMENTS

My grateful thanks are due to Dr. R. G. Cochrane, F.R.C.P., for help both in the clinical examination of the cases studied in this paper, and in its preparation.

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