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A Survey of Leprosy amongst the Lovale Tribe in the Upper Zambesi Basin, Northern Rhodesia*

BY

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PART II

Leprosy is a chronic disease due to the invasion of the skin, mucous membranes, peripheral nerves, the cells of the reticulo-endothelial system and, less commonly, certain other parts of the body (e.g., testis) with the Mycobacterium leprae. We do not know how the bacilli gain entry and become established in the tissues of the host, but all I have observed is in keeping with the view that most infections probably take place through skin abrasions and, just possibly, some, particularly in children, via intact skin.

In the patient with poor resistance, once infection is established, there is a general and symmetrical involvement of the skin, probably via the skin lymph channels and the bacilli are disseminated through the blood stream and reach the spleen, liver and bone-marrow and sometimes the testis. In these lepromatous cases the peripheral nerves usually, sooner or later, become symmetrically affected, but the central nervous system is exempt. Hard and grossly thickened nerves, so common in tuberculoid leprosy, are not usually seen in the lepromatous cases.

In the patient with high resistance the bacillary invasion tends to be limited to well-defined, asymmetric skin lesions and corresponding peripheral nerves.

In between these two well-defined polar types of low and high resistance there falls a considerable group of cases with varying resistance and varying clinical features. Under the Madrid, 1953, classification these include indeterminate and borderline or dimorphous cases.

46.3 per cent. of all cases of leprosy seen in the Balovale area show some involvement of peripheral nerve trunks.

It has not been uncommon in my experience to see cases of acute or subacute onset. In 1952, while visiting a village some 100 miles from my station, a woman requested treatment for a minor ailment and, while speaking to her, I noticed just a suspicion of roughness of the skin on her forehead. It was of very minor degree and very limited in extent. She did not com-

* Based on a thesis accepted for the degree of M.D., University of St. Andrew’s.
The literature of leprosy tends to place emphasis on the presence of anaesthesia in skin lesions, but skin anaesthesia to light touch (cotton wool) and thermal anaesthesia are often not found in the earliest lesions, especially of the lepromatous type. If present they usually indicate leprosy, but their absence is of no diagnostic value. In primitive people of low mentality it is difficult to detect these anaesthesias in their earliest stages.

CLINICAL SIGNS

Amongst these dark-skinned people the earliest sign of leprosy is commonly a hypopigmented skin macule which may appear anywhere on the body. (In leprosy the term “macule” has come to be used in a rather wider sense than its strict dermatological significance.)

Macules fall into three groups:—

I. Indeterminate

These hypopigmented macules are often single lesions, or perhaps there may be two or three, and typically seen in children. They have a clear-cut edge which is never raised, and the hypopigmentation is often only just sufficient to define the lesion. They are negative to bacteriological examination, give a negative lepromin test and show a simple inflammatory histology.

II. Tuberculoid (Major and Minor)

These are well-defined areas of hypopigmentation with raised margins.

The lesions are often multiple and present in different stages of development, asymmetrical, and sometimes as much as 12 inches or more in diameter. In a few cases the greater part of the body surface is covered—usually through the coalescence of a number of lesions. The larger lesions often show healing with flattening and return of pigment in the centre, while the spreading margins are raised and irregular and may show “colonial” or “pioneer” macules in advance of the spreading edge (Figs. 1 and 2). In African skins these lesions often appear almost white from superficial scaling; this is especially the case during reaction. When established they show analgesia, light tactile anaesthesia and thermal anaesthesia, and over 90 per cent. have a strongly positive lepromin test.

Patients with one or several tuberculoid lesions occasionally, in a very short time, develop generalised lesions of a similar type, often termed “lepraides.” These are allergic in nature and are comparable to the dermatophytids or secondary sensitivity eruptions seen in patients with local fungus infections. The term “lepride,” apparently first used by Unna (Arnold, 1953), is an accurate and suitable term for skin lesions of the tuberculoid type (Cochrane, 1947; Muir, 1940) (being comparable to the syphilide and tuberculide), but it has not been adopted in the international definitions of classification (Fig. 3).

III. Lepromatous

The characteristic feature of these macules is their vagueness. Usually multiple, symmetric and found anywhere on the body, they show slight hypopigmentation and ill-defined margins that merge gradually into healthy skin. They are easily missed unless carefully sought. The margins are never raised and the lesions do not, as a rule, show any marked impairment of sensation. They are almost invariably positive to bacteriological examination except in residual stages.

Lepromatous leprosy, other than the macular lepromata, is much more difficult to recognise in its earliest stages. An early diffuse leproma
will often escape all but a skilled observer, but smears, repeated if necessary, will confirm the diagnosis. In his 1939 African tour Muir recorded that “the diffuse infiltration lesions of the lepromatous type were not recognised” (Muir, 1940).

The classification of the Madrid Congress, 1953, now formally recognises a group of borderline or dimorphous cases. This group is defined as “malign, unstable, almost always strongly positive on bacteriological examination and lepromin reaction generally negative.” The skin lesions differ from lepromatous lesions in that they show “conspicuous asymmetry.” The skin lesions do not have clear-cut margins and are often of a succulent appearance. In the cases of this group seen here the lesions have appeared copper-coloured in the dark-skinned Africans. The nasal mucosa is often bacteriologically negative when all skin lesions are positive.

When the lepromatous state has become established with thickening of the skin, the formation of nodules and plaques, perhaps ulceration, characteristic features, symmetrical loss of eyebrows, erosion of nasal cartilage or chronic laryngitis, the diagnosis is simple. In these patients smears invariably show many bacilli, and biopsy shows characteristic histology. Lepromin test is invariably negative (Figs. 4 and 5).

Trophic lesions due to neural involvement are seen in any type of leprosy. Destruction of nerve components results in contractures, drop wrist, drop foot, muscle wasting, perforating ulcers of extremities, decalcification and absorption of bone, paralysis and analgesia and anaesthesia of areas supplied. Typically, nerve lesions in lepromatous leprosy tend to occur late and are symmetrical, while in tuberculoid types and in indeterminate and borderline groups nerve lesions may occur early and are asymmetrical.

Cases of pure nerve leprosy have been seen on two occasions only. Both the patients were young males and had a bilateral involvement of the ulnar nerves without any other signs of leprosy. One of these patients developed a left ulnar nerve abscess which required surgical treatment.

The Symmetry of Lesions

I now refer to a striking feature of Lovaland leprosy which merits emphasis—the symmetry of lesions. Some writers refer to this subject (Arnold, 1953; Muir, 1948; Rogers and Muir, 1946; Cochrane, 1947), but as recently as the Havana Congress in 1948 the official description of the three classes of leprosy then defined omitted any adequate reference to this
feature of leprosy lesions. The Madrid Congress classification of 1953 has remedied this and clearly indicates that in lepromatous leprosy "peripheral nerve trunks become manifestly involved . . . habitually in symmetrical fashion," while in tuberculoid cases "it is often asymmetric and unilateral."

(b) Erythema Nodosum Leprosum: Now commonly seen as a response to sulphone treatment and of good prognostic significance.

2. In Tuberculoid Leprosy

Reaction often develops suddenly—existing lesions become succulent, fresh lesions appear, smears become positive; eventually lesions desquamate and go on to complete healing. Acute leprous neuritis occurs and is most distressing. These cases require skilful handling if permanent nerve damage is to be avoided.

Complications

Reactions

1. In Lepromatous Leprosy

(a) Lepra Fever: In ten years' experience here, not more than two or three cases annually have been seen. This fact is referred to later in connection with the epidemiology of leprosy in this area.

Polyneuritis

This occurs in all types and groups of leprosy and results in contractures, ulceration, bone
absorption and trophic losses—occasionally, when neglected, going on to gangrene and loss of limbs.

LEPROUS PHARYNGITIS AND LARYNGITIS
Commonly seen in advanced lepromatous cases.

LEPROUS IRRITIS AND KERATITIS
Not at all common here.

SULPHONE DERMATITIS
Has occurred in 4 per cent. of patients on D.D.S. (Fig. 7.)

SULPHONE PSYCHOSIS
Only one case has occurred.

A few other rarer complications have been seen, none of them worthy of special note.

Fig. 7—Sulphone dermatitis.

THE CLINICAL COURSE OF LEPROSY

It is essential to remember, especially in assessing the results of treatment, that much leprosy is self-limiting in nature. Once the invading bacilli have gained entrance to the body their fate is determined by the resistance of the tissues of the host. It is not at all uncommon to see single abortive skin lesions that have become completely inactive and residual without any treatment. The two extremes of tissue resistance, i.e., energetic tissue response to the bacillary invasion and no resistance at all, produce the two polar types of leprosy—tuberculoid and lepromatous.

Nerve involvement occurs in all forms of leprosy, but its effects are often worst in the tuberculoid forms, where asymmetrical peripheral nerve trunk lesions commonly result in contractures, trophic ulceration and eventually bone absorption and loss of digits. In lepromatous leprosy the symmetrical nerve trunk involvement is late in the disease, and its effects are usually less crippling than those seen in tuberculoid leprosy. Severe nerve involvement may occur in the indeterminate group and also in the borderline group of cases.

Leprosy is rarely a killing disease; it attacks its victim; may find the host tissues uncongenial and die out without causing permanent effects; it may provoke a violent tissue response which in itself may produce permanent crippling lesions, or it may find no resistance at all and steadily progress, producing the repulsive, disfigured features and deformities seen in some advanced lepromatous cases, to whom life becomes a misery and whose lesions are constantly discharging bacilli and thus keeping the infection alive in the community.

TREATMENT

In 1893 William Tebb wrote, “leprosy is absolutely unamenable to therapeutic treatment.” In a popular book on leprosy published in 1938 it was stated, “it is scarcely justifiable to speak of ‘curing’ leprosy” (Weymouth, 1938).

In 1945 an American opinion was “an ample well-balanced diet and hygienic surroundings are probably more effective than any drug in the treatment of leprosy. It is doubtful if there is a truly specific method of therapy” (Manual of Tropical Medicine, 1945).

As recently as 1948 Muir wrote that “up to the last few years leprosy was considered an incurable disease and any treatment given was only in the nature of palliatives” (Muir, 1948).

When hydnocarpus oil and its derivatives were introduced in treatment by Sir Leonard Rogers in 1915, it was hailed as a great advance, but to-day practically all authorities agree that these preparations have little specific action against the Mycobacterium leprae. The beneficial effects from their use were mainly mechanical in nature, resulting from the irritation
induced—especially by intradermal injections, which undoubtedly gave good results in the hands of experts.

*Sulphones* have now gained general accept­ance as the treatment of choice for all forms of active leprosy. They were first introduced at Carville, U.S.A., in 1941 in the form of the complex sulphone “Promin” (Arnold, 1953; Lowe, 1950).

In 1949 Cochrane was the first to report the use of the parent sulphone, diamino-diphenyl sulphone (Cochrane et al.). Lowe (1950), in his original report in January, 1950, paved the way for the general introduction of oral diamino-diphenyl sulphone.

Since 1950 some 2,401 patients at Chitokoloki Leprosy Settlement have received oral diamino-diphenyl sulphone and, in general, the results of treatment have been excellent. In lepromatous patients the exhibition of D.D.S. results in the steady subsidence of activity—the nodules, placques, skin thickening and infiltrations slowly shrink and all the indications at present are that most cases go on to ultimate arrest of the disease and the dying out of the infection. In a small proportion of severe lepromata, D.D.S. has failed to arrest the disease, but other factors, such as poor nutrition, possibly liver damage and intercurrent infections, have complicated the clinical picture.

In tuberculoid cases the cessation of activity is more rapid, but progress must be gauged by clinical observations alone, as bacteriological checks are not applicable to these cases.

Once nerve lesions have become established the response to sulphones is not so dramatic and deterioration in function is progressive in most cases, in spite of prolonged treatment.

It is still too soon to draw any final conclusions as to the permanency of improvement seen here in all forms of leprosy under D.D.S. treatment, as the treatment has been in use for some six years only, but there is already overwhelming evidence that sulphone treatment is by far the most effective treatment yet used for leprosy here in Northern Rhodesia.

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