THE MACULAR SYNDROME IN NIGERIA

G. A. RYRIE

During a recent tour of Southern Nigeria the author made a preliminary study of the macular leprosy which forms so considerable a percentage and so marked a feature of the disease in this area. These observations are presented, despite their incompleteness, in the hope that information may be forthcoming as to the existence of this form of leprosy in places other than Southern Nigeria.

The word " macule " is frequently used in the literature of leprosy to describe almost any circumscribed lesion of the skin. In this article " macule " retains its strict dermatological meaning —a small discoloured patch on the skin not elevated above the general surface. The onset of the macular syndrome may be insidious or relatively abrupt. As a rule there is the appearance of an initial or primary single macule which may vary in size from a pin head to a circumscribed area of more than five centimetres in diameter. Or there may appear multiple macules on the trunk. It seems possible that the appearance of multiple macules however is essentially a secondary phase. The primary lesion may not have been noticed by an unintelligent patient, it may have been obscured by dirt or concomitant skin disease, or it may have occurred on some area of the skin not normally seen by the patient himself. However, although the appearance of a single primary lesion may be more in line with our general ideas on the onset of leprosy, there is a not infrequent history of a more or less abrupt onset of multiple macules.

The early macule may be rounded, oval or roughly irregular. At this stage the edge is well defined. The lesion is depigmented: on the African skin it is usually a cafe-au-lait colour. It may, however, vary from a colour like that of a new leather sole, through varying shades of a milk caramel appearance. Very careful examination will usually shew a faint erythema underlying the depigmentation, though the erythema may not be so marked at this stage as later on in the syndrome. There is of course no raised edge. There are two points of note—(1) There are no bacilli to be found; (2) *There is no tactile anæsthesia, no thermal anæsthesia and no evidence of any functional skin paresis.*

This lesion should not be confused with " juvenile leprosy " (Muir, 1936) or with the prelepromatous macule (Cochrane, 1946). The appearance and clinical course of the macular syndrome is quite different.

The lesion of the macular syndrome is differentiated also from the tuberculoid or neural macule by the absence of any loss of sensation. It is frequently held that a definite diagnosis of leprosy is unjustified without the elicitation of sensation-loss, or the presence of bacilli. Here, however, we encounter a lesion which shews no sensory deprivation, no bacilli, and which in appearance and in subsequent course is quite unlike juvenile leprosy or the neural macule, which in other words does not correspond with these declared and recognised lesion types.

1. The early macule may disappear or fade clinically with the appearance of a fresh macule at a later stage.

2. New solitary lesions may appear at intervals.

3. The macules may slowly spread centrifugally with a tendency towards coalescence.

4. There may occur the remarkable phenomenon which I will term macule reaction, and which appears to be analogous to lepromatous reaction or lepra fever, and tuberculoid reaction. *The reaction of the macular syndrome consists essentially of the outbreak of a crop of macules almost overnight.* These occur commonly on the trunk. There appears to be little constitutional disturbance. At present nothing is known of the blood sedimentation index or of plantar hyperalgesia during this phase. Indeed the main justification for applying the term reaction to this condition is its peculiarly eruptive nature. Successive reactions may leave successive crops of macules.

From this point certain changes may occur in the appearance of the macules. They may remain stationary or multiply, but in either case retain their clear cut edge. Or the edge may become diffuse and hazy; the lesions may spread and coalesce; they may go through a bacteriologically positive phase. Thus we see on the one hand in the discrete clear cut macule an analogy to the neural lesion, on the other hand in the hazy-edged macule an analogy to the lepromatous lesion.

The coalescing lesions may spread until practically the whole skin surface is involved, the patient looking like a Eurafrican with two slanting strips of black skin along the groin to shew the original colour of the skin (Davey, 1942). *Let it be noted that through all the phases outlined above, the lesion remains free from any sensation loss.*

The prognosis seems to be good on the whole. It is difficult to say if there is a tendency towards self-healing, but the response to treatment appears to be good.

There is no involvement of local nerves running proximal to the lesion as there is in tuberculoid leprosy. On the other hand, the ulnar and personal nerves may be frequently thickened.

In the later stage the macular syndrome may sometimes become lepromatous, or more rarely tuberculoid. Or perhaps it may be more correct to say that there is a tuberculoid or lepromatous superimposition on the original macular type. Then small raised lepromatous areas may occur in the centre of a macule; or nodules may appear on the ears and alae nasi, while the macules remain unchanged on the trunk.

Where there is a tendency for tuberculoid change the macules may lose thermal sensitivity, this being perhaps the first sign of neural involvement. Loss of the sense of heat and cold is apparently commoner than the loss of the touch sense, though the latter may also become paralysed. These tuberculoid or lepromatous superimpositions are, however, deviations from the macular syndrome and cannot be regarded as typical.

Some further features of the macular syndrome:—

1. There is a tendency for macular lesions to appear on areas which are relatively immune to tuberculoid or lepromatous leprosy. Lesions on the area between the nose and upper lip are uncommon in early tuberculoid or lepromatous leprosy; they are frequently seen in the leprosy of the macular syndrome. Again in the spreading stages macules are often seen on the lower abdomen, a site which has considerable relative immunity from tuberculoid and lepromatous leprosy.

2. The lips themselves may shew depigmented lesions, i.e., macules appearing on the modified mucous membrane of the outer parts of the lips.

3. Macules of this syndrome are frequently treated locally by caustics—trichloracetic acid, carbolic acid or, in native treatment, by caustic leaves. The lesion may then be obliterated, leaving a scar. Later the lesion may re-appear as an aureola, and spread with an ever widening margin round the scarified area. A similar phenomenon has of course been observed both in tuberculoid and lepromatous leprosy.

The essential features of the macular syndrome therefore appear to be:—

(a) The absence of any sign of sensory deficiency in the lesions.

(b) The essentially macular structure which dominates the whole course.

(c) The marked depigmentation of the macules.

(d) The underlying erythema, faint at first, but variable and perhaps most marked in the hazy-edged macular phase.
(e) The phenomenon of macule reaction with the abrupt appearance of fresh crops of macules, but without any marked signs of inflammatory response.
(f) The frequent presence of thickened ulnar and peroneal nerves and the absence of thickened nerves running proximal to the skin lesions.
(g) The absence of bacilli except during a possibily temporary late phase or when the case has become lepromatous.

It will at once be seen that this syndrome presents marked deviations from either tuberculoid or lepromatous leprosy as seen in other countries. In many parts of S. Nigeria this syndrome is by far the commonest manifestation of leprosy.

The place of the macular syndrome in the classification presents some difficulties. In the first place the whole syndrome can be labelled " Uncharacteristic." This has the justification that the typical histological features of tuberculoid and lepromatous leprosy are probably absent, though there is no proof of this at present. But then the macular syndrome is not a histological picture; it is an observable and clear cut sequence of pathological events which recur with sufficient regularity and frequency to merit recognition as a clinical entity. Secondly its inclusion under the term " Uncharacteristic " would tend to confuse it with borderline or intermediate leprosy, the various forms of early macule, neural leprosy, residual leprosy or any one of the other forms of which uncharacteristic leprosy appears to be a conglomerate.

Secondly, an attempt may be made to split up the macular syndrome into components which fit into either the tuberculoid or lepromatous concept. Davey has made a most interesting analysis of these macules, and an inclusion of them under a broad hypothesis in which tuberculoid and lepromatous leprosy is utilised (Davey, 1946). But we must remember that the absence of any anæsthesia of the lesions makes it difficult to fit them into the neural-tuberculoid group. Equally the diffuseness of some of these lesions, and the occasional presence of bacilli, does not necessarily make them lepromatous. Nor indeed does the clinical appearance of the macular syndrome suggest to an experienced observer any phase of tuberculoid or lepromatous leprosy.

Is the macular syndrome then a type sui generis and in consequence (accepting the polar concept) must we regard leprosy in Nigeria as tripolar?

The macular syndrome differs from neural tuberculoid leprosy in the absence of the relatively massive and specific cellular response of the latter, the absence of anæsthesia and the absence of any marked thickening of local nerves proximal to the lesion.

Yet the depigmentation of the lesions and the frequent thickening of ulnar and peroneal nerves in the macular syndrome suggest that the macular syndrome and tuberculoid leprosy are two great branches which spring from neural leprosy as the parent trunk.

The above description is necessarily incomplete and may be erroneous in certain details. The writer was unable to make direct investigations on the lepromin test, histology and bacteriology of this condition. Sufficient, however, has been outlined to enable readers to identify the condition if it exists elsewhere. It is in the hope that others may identify the condition that this article has been written.

REFERENCES.

MUIR, E. (1936), Juvenile Leprosy, *Int. Jl. Lep.* **4**, 45.
COCHRANE, R. G. (1946), The Classification of Leprosy, *Lep. Rev.* **17**, 39.
DAVEY, T. F. (1942), Masked Lepromatous Leprosy, *Lep. Rev.* **13**, 3.