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Ocular leprosy in West Malaysia

Search for a posterior segment lesion

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The publication of an earlier communication by the author (1969) on ocular leprosy inadvertently posed a problem, that of the posterior segment lesion. Suggestions had been put forward previously by other workers, notably Chatterjee and Chaudhury (1964a, 1964b), but it was not till Choyce (1964, 1967, 1969) made rather more emphatic statements that the problem was brought into the arena of discussion. Choyce's arguments, however, do not appear to be very strongly substantiated nor were they supported by any large series of figures, though he does maintain that 4 per cent. of leprosy patients show the specific lesion which he described as:

> "a heaped-up, highly refractile hypopigmented area at the periphery of the fundus, particularly the temporal periphery . . . "

On the other hand, Ticho and Ben Sira (1970), working in Malawi on 8,325 leprosy patients, stated that only occasionally was even a non-specific choroiditis found in their large series, but they did not supply any figures of this number.

Though it might be regarded as of mere academic interest, it does appear that, if such a lesion could be demonstrated in the fundus in an early stage of the systemic infection, it could serve to initiate further investigation of suspected leprosy just as a specific retinopathy does in diabetes and hypertension.

The opportunity arose in 1970 when facilities were made available for further studies on this subject at the Sungei Buloh leprosarium.

Sungei Buloh Leprosarium

The need for a modern leprosarium in Malaysia was first mooted by Sir George Maxwell, subsequently Chief Secretary, Federated Malay States, as long ago as 1919, and his ideas were realized in 1930 when the buildings of the leprosarium were finally completed in the valley of the Sungei Buloh (*sungei* = river) about 14 miles from Kuala Lumpur. With its opening it took over from other smaller leper asylums that had existed previously in Taiping, in Pangkor Island, and in Kuala Lumpur itself down the Circular Road. The original idea of having a pleasant modern settlement for isolating leprosy cases soon developed into the open colony system with no signs either manifest or implied of a prison atmosphere. In fact, patients today are encouraged to leave the colony once they have been declared non-infective, generally a matter of 6 months from the commencement of treatment, and as dapsone is readily obtainable at many centres throughout West Malaysia there is no necessity for further incarceration.

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The settlement now forms part of a much wider Leprosy Control Centre for the whole of West Malaysia, a project that was started early in 1967.

The leprosarium as it stands has, in addition to hospital wards and facilities (including reconstructive surgery), accommodation for 2,500 inmates in chalets and dormitories, physiotherapy and occupational therapy units, and an artificial limb centre. There is a school for affected children, and a Baby's Home, the babies being separated immediately from their parents and kept here for a short period before being adopted by relatives or well-wishers outside the leprosarium.

A feature of the settlement is that the inmates control most of their activities through a Council, only two members of which are officials of the hospital. A community centre and a cinema complete the picture. Most of the inmates eventually become self-supporting, running their own shops, and engaging in some occupation such as kitchen gardening, and goat, poultry, or pig keeping. Prominent among the occupations available to the inmates are appointments to the nursing and para-medical staff of the hospital, almost the entire complement being inmates of the settlement trained at the Nurses' Training School within the settlement (Kim, 1967).

Scope of the work

It was impossible within the period of one year to examine all the inmates, who numbered nearly 2,500. The primary purpose was to examine as many fundi as possible, the choice of patient being left to the hospital assistant who would collect from within the colony up to twenty patients, either tuberculoid or lepromatous, for each visit. In addition, any case referred for opinion regarding ocular diagnosis and treatment was added to the list for the day.

When full dilatation had been achieved, the fundus was examined minutely and systematically for any abnormal features. Slit-lamp microscopy was carried out with the aid of a Hobbs's hand slit lamp in the first instance, and the patient was subsequently referred for more detailed examination on the table model if a lesion was detected.

A second and incidental part of the programme consisted in selecting cases of active iritis for treatment with subconjunctival rifamycin in an attempt to assess the value of this drug as a local therapeutic agent.

A third part of the programme covered a general survey of the cases in the hospital wards to classify the ocular lesions. Here all patients were examined whether their disease was tuberculoid, lepromatous, or indeterminate.

A total of 444 cases (888 eyes) was examined and these constitute the basis of this report.

Findings

INCIDENCE OF OCULAR INVOLVEMENT

A general impression was created earlier (Mann, 1966) that leprosy in Malaysia does not cause ocular involvement as frequently as it does in other countries. This does not appear to be borne out by our figures (Table I, overleaf), which show that 230 (51·8 per cent.) of the 444 patients examined had some ocular or adnexal lesion that could be considered as being undoubtedly leprotic in origin. It is admitted that 'active' cases, mainly those involving the iris and sclera, were not numerous, but longstanding ocular involvement of every possible kind was rather common.

Table I *Analysis of 444 cases of leprosy*

Total no. of patients examined			444
Patients with eye conditions undoubtedly due to leprosy		*No.*	130
		Per cent.	51·8
Conditions due to leprosy	Absence of lashes		111
	Seventh nerve involvement		209
	Lid nodules		7
	Lid erythema		3
	Anaesthetic patches on lid		1
	Scleritis		6
	Episcleritis		4
	Episcleral nodule		2
	Corneal anaesthesia		2
	Corneal nebula		14
	Exposure keratitis		6
	Corneal vascularization		2
	Calcification		2
	Corneal staphyloma		1
	Interstitial keratitis		1
	Adherent leucoma		1
	Corneal macula		2
	Active iritis		3
	Chronic iritis		43
	Iris nodule		1
Conditions probably leprotic	Phthisis bulbi		4
	Fundus (without other underlying aetiology)		16
	Ciliary staphyloma		2
	Paralytic squint		1
Conditions not considered leprotic	Cataract		111
	Fundus (with underlying aetiology)		30
	Vitreous		3
	Concomitant squint		4
	Disc lesion		5
	Conjunctiva		32
	Sac		1
	Entropion of upper lid		4

(Multiple lesions have been classified separately)

Fundus conditions

Of the 444 cases examined, 290 (513 eyes) had fundoscopies carried out under mydriatic, and the discs and fundi were scrutinized. In 67 eyes fundoscopies were not possible either because the ocular media would not permit a clear view of the fundus or, as was found in four cases, the inflammatory condition had proceeded to a phthisis bulbi. The remainder were examined without dilating the pupil.

Abnormal fundus conditions were detected in 51 eyes (Table II, opposite), of which 35 were considered as being due to causes other than leprosy. Of the sixteen eyes in which the aetiology was unexplained, and therefore left unclassified, leprosy as the responsible agent was considered. None of these sixteen conditions corresponded in any degree with the lesion described by Choyce, and in one case the exact opposite was found. The eyes showed a highly pigmented irregular area with a narrow hypopigmented almost greenish-yellow border lying alongside the nasal edge of each disc and about the size of

Table II *All fundus conditions*

Condition	*No. of eyes*
Choroidal atrophy	1
Central myopic degeneration	1
Myopic sclerosis	3
Choroidal sclerosis	5
Tigroid fundus	2
Macular degeneration	7
Retinal dysplasia	1
Diabetic retinopathy	2
Renal retinopathy	2
Hypertensive retinopathy	2
Hypertension with colloid degeneration at macula	4
Optic nerve conditions	5
Conditions with underlying aetiology unknown:	
Hypopigmented patch on nasal side of each disc	2
Colloid degeneration centrally	6
Pepper-and-salt degeneration	4
Drusen (solitary & multiple)	4
Total	51

the latter. It was noted at the time that the lesion was quite unlike that produced by a resolving choroidal haemorrhage.

Four other cases (6 eyes) showed waxy exudates in the central area, rather like colloid degeneration, but more extensive and symmetrical. They were large irregular waxy exudates arranged radially round the macula, with rapid decrease in individual size of each exudate as the lesion was traced outwards. The affected area was limited to the macular and para-macular region and did not extend as far as the equator. In one of these cases the waxy exudates were small, the largest being the size of a pin-head, whereas the commoner finding was that the more centrally placed exudates were larger, being 3 to 4 mm. in diameter. The visual acuity was generally unaffected, and the condition was bilateral except in two cases. Clinical details of these are included. below

Case 1, a man aged 25 years, a Malay with lepromatous leprosy.

The visual acuity was 6/18, 6/6. Bilateral lesion: discrete waxy exudates clustered round the macula. The largest were 3 to 4 mm. in diameter nearest the macula and smallest further out and about the size of pin-head, but a few smaller exudates were also found in between the larger ones. All investigations were negative (blood pressure, urine, blood Wassermann reaction, chest *x* ray).

Case 2, a man aged 31 years, a Malay with lepromatous leprosy.

Visual acuity 6/6; 6/9. Bilateral lesion: brownish discoloration of both maculae with waxy 'colloid' degeneration (discrete exudates) in the macular and para-macular regions, each about 3 mm. in diameter. Investigations negative.

Case 3, a man aged 59 years, a Chinese with lepromatous leprosy.

Visual acuity 6/12 in affected eye. Unilateral lesion: limited 'colloid' degeneration in paramacular area. Left eye unaffected. Investigations negative.

Case 4, a woman aged 45 years, an Indian with lepromatous leprosy.

Visual acuity 6/36 in affected eye. Unilateral lesion: punctate waxy exudates in macular region of right eye. Left eye unaffected. Investigations negative.

Two other cases (4 eyes) showed a similar though more limited colloid degeneration in the macular region with good vision in each eye. They were found to have an associated hypertension and have been excluded from consideration, even though the lesion appeared typically 'waxy' rather than hypertensive. Both these patients also had lepromatous leprosy.

Of the remaining abnormal fundi, four eyes showed some type of pepper-and-salt degeneration particularly in the lower temporal quadrant of the fundus. These patients were in the younger age group, under 19 years of age, and no other eye lesion was demonstrable.

Four other eyes showed 'drusen' scattered either singly or in groups over the fundus. In the absence of a discoverable cause, these retinopathies cannot with justice be left unconsidered in this search for a posterior segment leprotic lesion. Macular degenerations were excluded.

Adnexal conditions

Involvement of the lashes to a greater or lesser extent appears to be a common manifestation of ocular leprosy. A total of 111 cases (222 eyes) were involved in this way. They were more often sparse than totally denuded. An absence of brows, especially over the lateral apsect, was noted to be almost twice as common (219 cases), but this has not been included as an adnexal structure.

Paralytic lid conditions were next in order of frequency, 209 being the result of involvement of the seventh nerve with varying degrees of palsy, from a mild eversion of the punctum to a marked ectropion, from a mere weakness of the orbicularis (as evidenced by poor resistance to forced opening of the eyes) to an early but definite lagophthalmos, and finally a severe lagophthalmos with or without exposure keratitis. Other lid conditions (lid nodules, erythema, anaesthetic patches) were infrequently seen. The two cases of entropion reported were probably the result of associated trachoma rather than of leprosy. Seventh nerve involvement accounted for 23·5 per cent. of lesions in the eyes examined.

Cornea

This was involved in thirty eyes (3·3 per cent.). Strangely enough, anaesthesia of the cornea occurred in only two of them. There were six cases of exposure keratitis, and the majority of the remainder showed some form of corneal opacity. No case of active keratitis was noted in this series.

Sclera

Episcleritis and scleritis which occurred in ten cases were the cause of repeated attacks that brought the patient to the clinic again and again. Often the condition was bilateral. There were only two cases of episcleral nodule, which may be considered surprisingly low.

Uvea

This was involved in a large number of cases, 47 eyes (5·3 per cent.); 43 showed evidence of chronic iritis, while only three eyes showed signs of active inflammation at the time of examination. Only one iris nodule was demonstrable in the whole series.

Other findings not considered as being due to leprosy

These have not been taken into consideration here, though one such case might be mentioned.

A Chinese male aged 18 years was referred from the research ward with bilateral papilloedema and visual acuity of 6/6 in each eye. He had developed erythema nodosum leporsum (ENL) 5 years previously and when this was initially treated with ACTH he developed severe hypertenison, papilloedema, and hypertensive encephalopathy. From that time onwards he had received almost continuous treatment with prednisolone in a dosage of 10 to 20 mg. daily for most of the time; and in the proceeding 2 years thalidomide had been added to this treatment. It was considered that the papilloedema was a complication of the steroid therapy rather than in any way secondary to the leprotic condition (Pearson, 1971).

INCIDENCE OF BLINDNESS

In a statistical survey of 2,235 patients at the Sungei Buloh leprosarium, Kim (1967) found 66 blind persons in the Decrepit ward and fifteen in the East and Central sections of the settlement, a total incidence of 3·6 per cent. He did not however mention the visual criteria he employed in his assessment of the blindness. In his series, 48 patients were binocularly 'blind'. In our total of 888 eyes examined, sixty (6·7 per cent.) were found to have less than 3/60 vision; while only nine had both eyes so affected. Excluding senile cataracts (22), myopia (6), retinal dysplasia (1), optic atrophy (1), and surgical anophthalmos (1), 31 eyes (3·5 per cent.) owed their defective vision to the involvement of the eye by the leprotic condition. In thirteen (42 per cent.) of these 31 eyes the lesions were the result of some form of uveal involvement (Tables III and IV, overleaf).

ETHNIC AND SEX INCIDENCE

According to the Annual Report (1969) of the National Leprosy Control Centre, Sungei Buloh, there would have been at the beginning of the following year, 2,410 leprosy patients within the settlement, of whom 1,924 (79·8 per cent.) were Chinese, 272 (11·2 per cent.) Malays, and 204 (8·5 per cent.) Indians. The incidence of ocular leprosy in our series has not shown any significant variation from these proportions: Chinese 168 (72 per cent.), Malays 46 (20 per cent.), Indians 16 (8 per cent.).

The ratio of male to female in the Sungei Buloh population was, according to the same Report, in the proportion of 2·7 : 1. Similarly, in our series, twice as many males as females have ocular leprosy (Table V, overleaf).

CLINICAL TRIALS WITH RIFAMYCIN*

Three eyes with active iritis and three patients (4 eyes) with active scleritis were given rifamycin by subconjunctival injection (1·5 ml. equivalent to 75 mg. rifamide) in an attempt to assess the value of this drug as a local therapeutic agent. Its effectiveness against *Mycobacterium leprae* when given systematically has been convincingly demonstrated by other workers, notably Rees, Pearson, and Waters (1970), but the drug had not been used previously as a local agent. It was therefore suggested by Hobbs (1970) during his visit to Malaysia that trials might usefully be carried out at the leprosarium.

*Rifamycin, rifamide, for subconjunctival injection was provided by Lepetit Pharmaceuticals Limited, Slough, Bucks, as 3 ml. ampoules equivalent to 150 mg. rifamide

Table III *Incidence of blindness (visual acuity less than 3/60)*

Sex	*Total cases*	*Blindness*	
		Uniocular	*Binocular*
Male	33	27	6
Female	18	15	3
Total	51	42	9

Table IV *Causes of blindness in sixty eyes*

Cause	*No. of eyes*
Myopia	6
Iritis	7
Secondary cataract	5
Gelatinous exudate in A.C.	1
Senile cataracts	22
Phthisis bulbi	4
Calcified cornea	2
Anterior staphyloma	1
Vascularized cornea	2
Interstitial keratitis	1
Corneal nebula	1
Adherent leucoma	1
Corneal macula	2
Corneal degeneration	1
Surgical anophthalmos	1
Scleral nodule	1
Retinal dysplasia	1
Optic atrophy	1
Total	60

Table V *Ethnic and sex incidence of ocular leprosy*

Sex	*Total cases*	*Race*		
		Chinese	*Malay*	*Indian*
Male	160	111	34	15
Female	70	57	12	1
Total	230	168	46	16

All those to whom the drug was administered responded favourably and signs of inflammation had disappeared when they came up for review in the week after the injection. However, the impression was that the response was no more rapid than when the condition is treated with standard remedies, *viz.* local steroids. Mydriatics were used as adjuvant therapy in all cases. It should perhaps be mentioned that in one case of iritis the vision improved from 6/24 to 6/6.

The cases are too few for any conclusion to be drawn, and although the results are encouraging this method requires more extensive trials. The injection is quite painless, though rather bulky. Rifamide solution for intramuscullar injection was administered by the subconjunctival route in one case and proved to be extremely painful, as it usually is when given intramuscularly. The lids, conjunctiva, and orbital tissues became markedly oedematous, and caused a temporary but alarming proptosis and chemosis.

Conclusion

Choyce's lesion, as described by him, occurs primarily at the temporal periphery of the fundus, and consists of an area of heaped-up highly refractile waxy exudate. Other fundus lesions as described by Chatterjee and others (1964a,b) also comprise hypopigmented patches in the choroid. The dominant lesion in the present series, however, appears to favour the central region and, with one exception, the lesion so far observed does not appear to affect visual acuity. The condition closely resembles a colloid degeneration. Another striking feature is that the lesion does not appear to be associated with any gross leprotic affection of the eyes. There was no involvement of the iris or ciliary body anteriorly, while the ocular adnexae, except for one case of lagophthalmos,

were exempt from the disease. All the cases of this fundus lesion were found to be amongst lepromatous patients; none was tuberculoid.

It would appear then that, if the lesion as described here is leprotic in origin, it is an isolated and non-specific lesion, not due to contiguous spread from the ciliary body, as is obviously the case in Choyce's lesion. The question of a blood-borne bacillary embolism to this area of the fundus (as in the case of iris and ciliary body lesions) cannot thus be postulated, the cluster arrangement and the bilaterality of the lesion being more suggestive of a uveal tract sensitivity reaction concentrated on the macular region. Unfortunately no histological specimens were available in this small series.

Summary

Leprosy in West Malaysia as might be expected takes the same course as it does elsewhere. Seventh nerve involvement appears to be the commonest ocular complication, with anterior uveal complications as the commonest intraocular lesions. A serious degree of blindness was found among those patients who had suffered from the disease before therapy with sulphones became routine.

A posterior segment lesion resembling a colloid degeneration is described and it is tentatively suggested that, though perhaps not specific of the disease, it is probably a reaction of sensitised uveal tissue. It is in any case extremely uncommon, being found in only six eyes in this series of 888 eyes examined, a percentage of 0·67.

Local treatment with rifamycin by subconjunctival injection was given to a limited number of cases of active anterior segment inflammation. It appeared to be as effective as the standard therapy with steroids.

My thanks are due to the medical assistants of the Sungei Buloh Leprosarium, without whose ready help in the wards and at the clinics this work would not have been possible; to Lepetit Pharmaceuticals for a generous supply of Rifamycin; to Dr. J. M. H. Pearson, M.R.C.P., Leprosy Research Unit, Sungei Buloh, for permission to refer to one of his cases; to Dr. M. K. Bhojwani, M.R.C.P., Consultant Leprologist and Director, National Leprosy Control Centre, Sungei Buloh, Malaysia, for his permission to use the clinical material at the Leprosarium, and for his unfailing courtesy at all times.

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