

MINI REVIEW

The continuing challenge of ocular leprosy

The timely resurgence of interest in ocular leprosy noted in this journal a year ago continues,¹ as witnessed by the number of clinical papers from various centres which have recently appeared in the ophthalmic literature and the emphasis that is now being placed on measures for the prevention of blindness.² Apart from being one of the oldest infections known to man, leprosy is still the systemic disease with the highest incidence of ocular complications. Epidemiological studies of these have always proved to be difficult to obtain, and estimates of important aspects of the disease such as blindness rates and the prevalence of sight threatening lesions may fall wide of the mark.³

Ocular involvement in leprosy depends on a number of internal and external factors which may vary, not just within ethnic groups and geographic regions, but also from community to community. These factors include such variables as the age, race, and sex of the individual, the type of leprosy, its duration, therapy, and status. To these must be added the influence that the climate, environment, occupation, and social status have on the disease, and, most important of all, what measures to prevent eye complications are undertaken locally. Ocular leprosy is after all the archetypal preventive disease, and simple treatment at an early stage will usually avoid major irreversible damage later. This will be reflected by blindness and disability rates from different centres. Indeed, analysis of data derived from regional studies on the prevalence of eye complications often gives a very good idea of what services for leprosy are available in the community and how effective they are.

The problems of gathering information on the prevalence of eye complications in leprosy are further hindered by difficulties in estimating the actual numbers of patients with the disease. Many affected individuals do not come forward for treatment and therefore do not appear on registers until quite late in the course of disease. This is often due to ignorance, or it may reflect the social stigma that still exists throughout the world. The figures are further distorted by the WHO policy of deregistering patients once they have completed multidrug therapy and have satisfied the criteria for cure – a policy that has many pitfalls from the ophthalmic point of view. It is not therefore difficult to see why hard epidemiological data on the prevalence of ocular complications of leprosy are scarce, and knowledge on their incidence is almost non-existent.

A large multicentred study of ocular involvement in the disease, which included information on more than 3000 patients obtained by simple standardised examination techniques from over 30 centres in the world, showed that 5.5% of patients examined had bilateral corrected vision of less than 3/60 and were therefore blind by WHO definition, and 7.0% had vision less than 6/60, classified as having severe visual impairment. The same study demonstrated that 21.3% of patients had potentially sight-threatening lesions, defined as conditions likely to lead to visual loss unless preventive treatment was instituted.⁴ A parallel study on 354 patients who had been discharged from care having completed multidrug therapy, showed a prevalence of 24.3% with potentially sight-threatening lesions,⁵ and this raises the important point that in a considerable number of patients 'cure', as defined by the WHO, does not guarantee freedom from eye complications, and many patients may face increasing ocular problems

unless they are adequately supervised by leprologists after completing therapy.

Epidemiological information is therefore still lacking. But, if the global leprosy population is taken to be between 10 and 12 million,⁶ the disease ranks as one of the major causes of world blindness, likely to increase proportionally as others such as trachoma, xerophthalmia, and onchocerciasis become contained by modern therapy and public health measures.

The numbers, however, do not give the whole story. Leprosy is a unique condition, not least because of its social stigma, which evokes almost mediaeval sentiments in many countries, condemning the patient to become a member of one of the most disadvantaged groups in society. It is also a crippling disease. The nerve damage which is a feature of all forms of the condition, but particularly affects multibacillary (MB) patients, destroys tactile sensation, decreases mobility, and causes the familiar skeletal deformities. Loss of vision in these patients is a major event in this downward sequence of disability and frequently renders the patient entirely dependent on others. 'Blindness in the individual who has normal skin sensitivity is enough of a handicap, but in one who has lost that faculty it is disastrous. Few have the resources, material, mental or spiritual, to live with it.'⁷

The greatest current tragedy in this disease is that most of the ocular complications that lead to visual loss can be avoided by simple therapeutic measures requiring little more than adequate supervision, patient education and common sense. The role of the ophthalmologist should therefore be extended to one of a teacher, persuading leprologists and field workers and even fellow ophthalmologists, that leprosy is an ocular disease, that the eyes need supervision, not necessarily with sophisticated instruments, and that a mechanism for rapid referral to an ophthalmic centre should be available when serious sight-threatening complications develop.

Ocular involvement in leprosy occurs at several levels with differing effects on vision. In general the patients with the paucibacillary form, representing the tuberculoid and borderline-tuberculoid cases under the Ridley-Jopling classification,⁸ are affected through loss of lid function and corneal hypoaesthesia, whereas multibacillary patients, representing lepromatous, borderline-lepromatous, and mid borderline cases suffer complications through these as well as from direct infiltration of organisms into the anterior segment, inflammatory reactions, and damage to the adnexae.

Not surprisingly the many influential factors that determine which patients are at risk give rise to a variety of ocular manifestations, several of which do not threaten vision, but may indicate significant ocular involvement. They include such findings as madarosis, opaque corneal nerves, superficial stromal keratitis, and iris pearls.

Most of the visual impairment and blindness results from four main causes which may be isolated but often occur together: lagophthalmos leading to exposure keratopathy, corneal hypoaesthesia leading to ulceration, acute or chronic iridocyclitis, and secondary cataract.

LID INVOLVEMENT

The lower lid is affected through paralysis of the maxillary

branch of the facial nerve, giving rise to a reduction or failure of lid closure, with the consequent deleterious effect on protection of the cornea and lacrimal drainage. It occurs in all forms of leprosy, either as an acute event during a reversal reaction or over a long period of time. Immediate treatment of the acute stage with systemic steroids, thalidomide, or clofazamine will often lead to recovery or improvement of function.

In chronic cases, or in those which do not respond to medical therapy, it is often necessary to perform surgery on the lid, either in the form of a lateral tarsorrhaphy or by more sophisticated operations such as the tarsal strip procedure or temporalis transfer. Whereas tarsorrhaphy and some of the simpler lid surgery can be carried out by leprologists and field workers, procedures such as temporalis transfer require more expertise and usually involve an ophthalmologist.

The risk from lagophthalmos is always one of exposure keratopathy, particularly if the cornea already has diminished sensation. Monitoring of lid function by the leprologist and patient is an important preventive measure, and simple ways of protecting the eye can effectively reduce the incidence of corneal problems. Recent work by Hogeweg *et al.*⁹ on the relationship between the position of facial patches during erythema nodosum leprosum (ENL) reactions, and the subsequent development of lagophthalmos has considerably helped to identify those patients most at risk.

CORNEAL INVOLVEMENT

Corneal sensation is affected in all forms of the disease, and when hypoaesthesia accompanies impaired lid closure the eye is at high risk. Impaired corneal sensation predisposes to corneal damage as well as disturbing the normal epithelial metabolism, and the complications of corneal ulceration and opacification account for a significant proportion of blindness in leprosy. Diminished corneal sensation may well be an indicator of ocular pathology elsewhere as shown by the frequency with which it occurs in association with major ocular damage from the disease.¹⁰

Treatment consists of simple measures to protect the cornea at an early stage of the condition, and therefore detection of those eyes at risk is an important aspect of management. This is the combined responsibility of the leprosy worker and patient, since in the later stages the problems of dense vascularised corneal opacities may be insurmountable even with the most sophisticated surgical techniques.

IRIS INVOLVEMENT

Invasion of the iris and ciliary body by *Mycobacterium leprae* occurs exclusively in the multibacillary form of the disease. Acute iridocyclitis is often a feature of the ENL reaction and has all the signs of iritis due to any other cause except that pain and discomfort may not be so prominent. Conventional treatment with steroids and mydriatics is required, and field workers and leprologists need to be instructed in the early recognition and management of the condition.

Chronic iridocyclitis remains the enigma of this disease, though knowledge of the subject has increased considerably over the last decade. It is now accepted that organisms lodge in the iris and ciliary body at an early stage in lepromatous

leprosy, and attempts are being made to detect this involvement by measuring various types of pupil reactions such as the pupil cycle time, light/dark reactions, and pharmacological responses.¹¹ Other studies have analysed the effect of the disease on the ciliary body, manifested by a progressive lowering of intraocular pressure and alteration in postural responses.¹² Most authors now recognise that chronic iridocyclitis in multibacillary leprosy has neuroparalytic elements, with primary involvement of the autonomic nerves, particularly the sympathetic fibres in the iris and ciliary body. This may raise the possibility of pharmacological treatment at an early stage to prevent the chronic immobile miotic pupil that characterises the end stage of the disease, and which can at present be overcome only by surgical intervention or photocoagulation.

CATARACT

The question of 'leprosy cataract' has yet to be solved. The disease is endemic in most countries where cataract is a major problem, and certainly data collected in surveys show that many patients have poor vision related to lens opacities without any signs of ocular leprosy. There is however a reported high prevalence of cataract in MB cases completing multidrug therapy,⁵ and this merits further study. Sadly the stigma of the disease that still exists in many countries often excludes the leprosy patient with cataract from receiving the surgical services that are available to other members of the community.

Cochrane in 1956¹³ described leprosy as 'the most thrilling and exciting adventure on which any medical man can embark', and the hyperbole 35 years ago was understandable. Our knowledge of the intricacies of the disease as it affects the eye is still increasing, but more clinical and laboratory work is needed. As we move forward into the new century there are still research efforts to be expanded, therapies to rationalise, prejudices to overcome, and challenges to meet in this ancient disease.

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