Ocular Leprosy in the Canal Zone[1]

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Throughout the 70 year period of the United States' presence on the Isthmus of Panama, leprosy and its associated ocular problems have been a concern of civilian and military physicians stationed in that area. This paper presents the findings of a recent survey concerning the prevalence of eye complications among leprosy patients at the Palo Seco Hospital, Canal Zone.

HISTORY AND SUBJECTS

During the early days of the construction of the Panama Canal, Colonel William Crawford Gorgas, the conqueror of yellow fever and malaria, chose a site on the Pacific shore of the Canal Zone for the building of a medical facility for leprosy patients. When it was opened in 1907, three years after the United States had begun excavation of the canal, it was known as the "Palo Seco Leper Colony." The name was changed in 1948 to the "Palo Seco Leprosarium" and then in 1964 to its present designation, "Palo Seco Hospital."

From an initial census of nine patients, six males and three females, the number of those hospitalized increased to a maximum of 131 in 1940, but thereafter declined until in 1974 there were 48 patients, 32 males and 16 females. They ranged in age from 41 to 90 years. The average age was 66 and the average length of hospitalization was 36 years.

The distribution of patients according to place of birth is: Panama, 38; Colombia, 3; Costa Rica, 3; Barbados, 1; Jamaica, 1; Nicaragua, 1; St. Vincent, 1.

The classification of patients as to the type of leprosy is: lepromatous 39; tuberculoid 7; dimorphous 2.

In 1946 Dr. R. D. Harley reported the results of a survey at the Palo Seco Leper Colony and indicated that 90% of the patients had ocular complications ([6]).

METHODS

In 1972 a study was begun among patients at the Canal Zone institution to determine the current rate of ocular involvement. During the period 1972-1974 each of the patients was examined and, with the exception of three with bilateral enucleations and an equal number who were bedridden, all have been reexamined in the Gorgas Hospital Eye Clinic at intervals of approximately three months.

FIG. 1. Palo Seco Hospital, Canal Zone.

RESULTS

Madarosis. Leprotic involvement of the skin in the areas of the eyebrows and eyelashes may result in the loss of both due to destruction of the hair follicles. Loss of the eyebrows generally occurs before that of the lashes and characteristically begins in the outer third and proceeds nasally. Ultimately, there may be total loss of the eyebrows and eyelashes.

Madarosis was found in 35 of the 48 (73%) patients at Palo Seco and thus was the single most common evidence of the disease. This is in agreement with a recently reported study from Kampala, Uganda ([5]). Our percentage is somewhat higher than the 50% figure reported by Harley in 1946 ([6]) and markedly higher than the 16% noted by Hornblass among the Montagnard patients in South Vietnam in the years 1969-1970 ([7]). The low percentage noted there was probably related to the fact that the mean age of the Montagnard patients was 37, while at Palo Seco it was 65. Thus, the duration of the disease

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here has been considerably longer.

Dacryocystitis. Due to the nasal lesions of the disease, especially those causing periostitis and atrophy of the nasal bones, obstruction of the nasolacrimal duct may lead to dacryocystitis. In view of the gross nasal infection in many patients, Brand ([4]) noted that it is surprising that this condition is not encountered more often.

At Palo Seco a single case of bilateral chronic dacryocystitis was found (2%). Harley encountered four cases (3%) ([6]), and Weerekoon ([9]) in Ceylon found 14 cases among 630 patients (2%).

Lagophthalmos. Leprotic involvement of the facial nerve may lead to lagophthalmos with the resultant complications of exposure keratitis and corneal ulceration.

While in the survey of 30 years ago lagophthalmos was noted in 10% of the patients ([6]), in 1974 it was found in 9 of 45 (20%). Other recently reported rates of involvement are: Ceylon, 6% ([9]); Malawi, 3% ([8]); and Uganda, 6% ([5]). Although of interest, no real comparisons of significance with the Palo Seco group can be made because of marked differences in average age and type of leprosy.

Ectropion. Following leprotic involvement of the facial nerve, the patient may have a relaxation and atonicity in the tissues of the lid with a resultant ectropion. This in turn may cause exposure keratitis, excessive lacrimation and epiphora. The number of Palo Seco patients in 1974 with ectropion was 6 of 45 (13%), while in the 1946 report it was 11 of 150 (7%) ([6]).

Edema of the lids. The eyelids may be diffusely thickened due to edema or infiltration and they may show individual lepromatous nodular swellings. Three patients manifested edematous lids and four others presented lid nodules which might well have been lepromatous in etiology.

Entropion. Following resolution of a lid infiltrate which caused destruction of elastic and connective tissues one may find a flaccid entropion and consequent trichiasis in the leprosy patient. This was encountered in 20% of those at the Canal Zone Hospital.

Trichiasis. This condition may be a source of great annoyance to patients retaining corneal sensitivity and furthermore may be the cause of serious corneal injury. Trichiasis may be due to the previously mentioned entropion or to the distortion of hair follicles caused by post-inflammatory scarring. In this survey, 22 of 45 patients (49%) demonstrated and often complained about trichiasis.

TABLE 1. *General information.*

Total patients	48
Patients with ocular leprosy	46
Average age (years)	66
Average hospitalization (years)	36
Patients with lepromatous leprosy	39
Patients with tuberculoid leprosy	7
Patients with dimorphous leprosy	2

TABLE 2. *Leprotic ocular complications.*

Complication	No. of patients	Prevalence %
Madarosis	35	73
Dacryocystitis	1	2
Lagophthalmos	9	20
Ectropion	6	13
Edema of lids	3	7
Entropion	9	20
Trichiasis	22	49
Dermochalasis	19	42
Opacified corneal nerves	8	18
Superficial avascular keratitis	17	39
Pannus	19	43
Interstitial vascularization	8	18
Exposure keratitis	6	13
Corneal hypesthesia	31	69
Iridocyclitis or sequelae	28	64
Miliary lepromata of iris	4	9
Visual acuity 20/200 or less	24	50

Dermochalasis. This condition, in which the skin of the lids loses its elasticity and hangs in loose folds, may also be a sequel to inflammatory changes in the lids. While 19 of 45 patients (42%) had such changes, the significance of this is debatable since similar changes may be seen in many nonleprous patients in this age group.

Opacification of the corneal nerves. Early in the course of leprotic involvement of the globe of the eye one may detect transient edema of the corneal nerves. To the slit lamp examiner they appear to be wider and more opaque than normal nerves. Later in the course of the disease one may encounter minute irregularities along the course of the nerves which may have a bead-like appearance. Still later one may find chalky-white

"beads" in a linear arrangement with no evidence of nerve fibers between them. As Dr. Margaret Brand states, "Probably they have degenerated leaving a line of 'beads' but no 'string'" (4). The nerves which are first involved are generally those of the superior temporal quadrant, followed in order by those of the superior nasal, inferior temporal and inferior nasal quadrants.

Allen and Byers (2) described well the pathologic changes occurring in the cornea of leprosy patients. Dr. Harley wrote, "The 'beading' of the corneal nerves seen under high magnification is an interesting sight and easily overlooked. It was found in five cases but may be more common than this" (6).

Considerable searching with the slit lamp is at times needed to discover the characteristic linear pattern of fine opacities. This was found in 8 of the 44 patients examined with the biomicroscope.

Superficial avascular keratitis. This is considered as pathognomonic of leprosy. In the early stages the lesion is only visible with magnification, but later is obvious to the naked eye. With the slit lamp the early lesion is seen to be composed of a superficial, thin, patch-like or diffuse haze at about the level of Bowman's membrane. Somewhat later a number of punctate, chalky opacities begin to appear. The latter tend to develop in clusters giving the cornea a stippled appearance. The keratitis generally begins in the periphery of the superior temporal quadrant and may gradually spread to the other quadrants, but in many cases a prolonged or indefinite remission occurs after only partial corneal involvement. At times, as the active inflammation of the cornea subsides, the superficial thin, grayish-white opacity may disappear, though the chalky-white punctate opacities remain permanently. Often, however, Bowman's membrane is destroyed and replaced by veil-like scar tissue. The punctate opacities, which may slowly increase in size, are actually miliary lepromata and are at times referred to as "corneal pearls." Although the earliest opacities are superficial, one may find that with the passage of time the deeper layers of the cornea become involved, first peripherally near the limbus and then centrally.

The patients we have seen with avascular keratitis have been surprisingly asymptomatic. Often the patient is not aware of corneal involvement until it is sufficiently advanced to affect the pupillary area or the lower half of the cornea. Even at that point, he generally complains only of loss of visual acuity or "light scatter" symptoms.

Characteristic superficial avascular keratitis was noted in 17 patients (39%). Harley had previously reported the occurrence of this form of keratitis in 34% (6). Both figures are a marked contrast to the 1% occurrence rate in Uganda and to similarly low rates in Tanganyika, Ghana and Malawi (5).

Pannus. Following repeated, severe, or long-standing avascular keratitis, the blood vessels of the limbus may begin to extend onto the surface of or into the most superficial layers of the cornea. Pannus generally follows the same pattern of appearance as does corneal nerve involvement and superficial avascular keratitis: the superior temporal quadrant first and the inferior nasal quadrant last. This vascularization may be arrested at any point or it may proceed to involve the entire cornea.

Pannus was seen in 19 patients (43%). Harley found it in seven cases (5%), while in Uganda it was noted in only 5 of the 890 patients examined.

Interstitial vascularization. In a small percentage of patients a deep or interstitial vascularization of the cornea may develop. It may follow prolonged avascular keratitis after the deeper layers of the cornea have been heavily infiltrated or it may occur in some patients with a severe uveitis. Generally, it follows or accompanies pannus formation and usually begins in the upper temporal quadrant. As it spreads to other quadrants it follows the pattern previously noted.

Eight patients (18%) were noted to have interstitial vascularization, while in 1946 this problem was found in 15% of those examined.

Exposure keratitis. In patients with lagophthalmos, this condition begins insidiously. At first there is a diffuse haziness and dessication of the epithelium in the exposed area (the lower part of the cornea) with the development of a fine punctate epithelial keratitis which will stain with fluorescein. If untreated, fissures and exfoliation of the epithelium develop and eventually ulceration may result.

Exposure keratitis was found in six of nine patients who had lagophthalmos. In Malawi this corneal problem was reported in only 47 of 251 patients with lagophthalmos (8).

Hypesthesia. Due to leprotic involvement of the ophthalmic division of the fifth cranial nerve there often is corneal hypesthesia. It is an early change, appearing in some by the second or third year of the disease ([3]). It is most marked, however, in patients with long-standing lepromatous disease. The insensitive cornea may be readily damaged by dryness, foreign bodies, trichiasis and trauma.

Hypesthesia of varying degree was noted in 31 patients. The relative crudity of the cotton wisp method of testing corneal sensitivity casts some doubt upon the accuracy of this statistic.

Nodular episcleritis. This may occur during acute reactions in the course of the disease and then follow a subacute or chronic course. Thinning of the sclera may result. Although no cases of nodular episcleritis were encountered, three patients did exhibit staphylomata. These very possibly may have arisen at sites of previous nodular episcleritis.

Iridocyclitis. Lepra bacilli are thought to be present in the iris and ciliary body early in the disease, but probably lie dormant for several years. When iridocyclitis is first noted, it may be in the form of an acute, subacute, or subclinical iritis. At times a ciliary flush or a mild photophobia may be noted by the patient, but very often there are no symptoms in spite of flare and cells in the anterior chamber and fine keratic precipitates on the corneal endothelium. A chronic subclinical iritis is probably present in most patients who have had the disease for several years. It may persist indefinitely in that form or there may be clinically apparent acute or subacute exacerbations from time to time. In occasional patients an anterior chamber lepromatous nodule may appear during such an exacerbation and persist after the flare and cells in the anterior chamber have cleared. The sequelae of untreated iridocyclitis are a major cause of severe ocular damage in this affliction.

Half of the patients examined with the slit lamp during this survey were found to have an active iridocyclitis of varying intensity at one time or another. Six other patients evidenced past iritis by virtue of synechiae discovered on biomicroscopy or gonioscopy. Thus 64% gave evidence of present or past iritis. The first survey at Palo Seco reported evidence of iritis or its sequelae in over 50% of those examined ([6]). A 1969 report from Ceylon indicated that 17% of 630 leprosy patients had iritis ([9]).

Miliary lepromata of the iris. These "iris pearls" as they are usually called are pathognomonic features of lepromatous leprosy. They have their origin early in the course of the disease, developing in the iris stroma, generally near the pupillary border. They increase in size and may coalesce with neighboring pearls. As this growth process continues, they approach the surface of the iris, protrude through the surface and may eventually break loose to fall into the anterior chamber. The individual pearls containing leprosy bacilli and calcium salts are spherical, creamy-white or faintly yellowish-white and opaque. They seldom attain a diameter of more than 0.5 mm, but at times a conglomerate pearl may reach a diameter of 2 mm ([1]).

Four patients were found to have typical iris pearls. Some were fixed to the surface of the iris near the pupillary border, while others were seen in the angle of the anterior chamber during gonioscopy.

Impairment of visual acuity. As one would expect from the preceding list of ocular complications, impairment of vision is common in leprosy. Three patients in the study had had bilateral enucleations. Two others had no light perception in either eye. Altogether there were 24 patients (50%) who had a best correctable vision of 20/200 or less in one or both eyes. This corresponds closely to the 54% who were found to have the same visual deficit in 1946 ([6]).

DISCUSSION

The eye and its adnexal tissues are frequently involved in leprosy: the prevalence of ocular complications in the tuberculoid form of the disease is approximately 15%, while in advanced cases of lepromatous leprosy it approaches 100%. The specific ways in which the eyes may be affected are as follows:

1. Direct invasion of the eye by *Mycobacterium leprae.*

2. Leprous involvement of the seventh cranial nerve or of the fifth nerve at the cornea with resultant changes in the eye.

3. Damage to the structures adjacent to the eyes with secondary ocular problems.

4. Participation in the generalized allergic response encountered in lepromatous lep-

rosy known as the "reactive phase" or "lepra reaction."

In many patients, especially those with advanced lepromatous disease, all four factors may contribute to their eye problems.

SUMMARY

The results of a two year survey of eye problems among the patients at the Palo Seco Hospital in the Canal Zone are presented. Only two patients, one classified as having lepromatous leprosy and the other as having the tuberculoid form of the disease, failed to exhibit ocular complications. The high prevalence of leprotic ocular disease (96%) is most probably due to the advanced age of the patients, the lengthy duration of their illness, and the high percentage of patients afflicted by the lepromatous form of the disease.

RESUMEN

Se presentan los resultados obtenidos en un estudio de dos años de duración sobre la incidencia de problemas oculares en pacientes con lepra del Hospital de Palo Seco en la Zona del Canal. Sólo dos de los pacientes estudiados no presentaron complicaciones oculares. Muy probablemente, la elevada incidencia de afecciones lepróticas oculares (96%) está relacionada con la avanzada edad de los pacientes, con la prolongada duración de la enfermedad, y con el alto porcentaje de pacientes afectados por la forma lepromatosa de la lepra.

RÉSUMÉ

On présente ici les résultats d'une enquête sur les lésions oculaires menée pendant deux ans parmi les malades de l'hôpital Palo Seco dans la Zone du Canal. Deux malades seulement, dont l'un avait été classé comme malade lépromateux et l'autre comme tuberculoïde, étaient indemnes de complication oculaire. La prévalence élevée de lèpre oculaire (96 pour cent) est probablement due à l'âge avancé des malades, à la longue durée de leur affection, et au pourcentage élevé de sujets atteints de la forme lépromateuse de la maladie.

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