

A FURTHER REPORT OF POLYPOID LEPROUS LESIONS OF THE FUNDUS ¹

OBSERVATIONS OF SIX CASES

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Distinctive changes on the fundus of the eye which we believe are due to leprosy, seen in a single case, have been described in a previous report (3). Since then five other patients with similar lesions have been seen in this hospital, making a total of six.

Four of these six cases, including the one first reported, were classified as lepromatous. One of the others was of essentially the same category except more advanced, and therefore called "mixed"; the remaining case was classified as of tuberculoid type.

All of the retinal lesions seen in these patients are, in our opinion, leprous nodules or polyps, for they are morphologically similar to the pearl-like nodules of leprosy so often observed on the iris. On the fundus these pearl-like nodules seem to be projected toward the observer on a single polypoid structure of faintly yellowish-gray color, and they seem to have a waxy surface. They vary in size; a few are smaller in diameter than a retinal artery at the disc, but more commonly they are one or two times the diameter of the artery. In only one case, No. 3, did they fuse to present a conglomerate picture, though even then there were many single polyps on and above the fused base of the waxy, hobnail mass. These fundus polyps were demonstrated to the medical staff with the regular May ophthalmoscope.

LITERATURE

In the previous report certain more or less recent publications were referred to, none of which described lesions such as had been observed in our case. Little that is pertinent has been found since then.

Regarding retinal changes Klingmüller (5) refers only to Bull and Hansen (1) as finding small nodules containing globi with slight tissue changes in the neighborhood of the ciliary body. However, they said that such lesions hardly extend beyond the ora serrata, according to Jeanselme

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(4). He cites Uchida (7) as finding most of the retinal changes at the level of the ora serrata, gradually decreasing as they go farther back and seldom reaching the equator of the eye; yet infiltrations were found in the posterior part of the retina in 9 (14%) of 64 eyes examined. These infiltrations were often found in the neighborhood of the blood vessels.

Jeanselme says that clinical examination of the retina is often impossible because of corneal opacity or other lesions in the anterior portions of the eye, and that he had found only a single instance of the examination in a living case, that of Trantas (6). Cochrane (2) is also of the opinion that the lack of observations of fundus changes in the literature is due to the fact that their visualization is prevented by the degree of change usually found in the anterior portion of the eye.

The case of Trantas, according to Jeanselme, was one of the neural type in which, among other things, there were "... round points, having hardly the dimensions of the head of a pin ... which are beside the walls of the small macular veins. Not far away, on the temporal side, there is an alteration of the pigment of the fundus. ... Furthermore, there are also one or two elongate yellowish spots toward the ora serrata, above and below. The visual acuity is almost normal."

The lesions which Bull and Hansen are credited with recording were evidently not the same as those here described; at least their location was different. Uchida clearly dealt with a histological condition, but infiltrations such as he described may very well be the starting point of the outgrowths which we have observed. While we concur in Cochrane's explanation of the rarity of the observations of fundus changes, the six cases reported here illustrate the fact that some patients may have such changes in an earlier phase, when they can be seen.

REPORT OF CASES

The previously reported case is summarized as "Case 1" below, with later observations. The other cases are the new ones.

CASE 1.—An ophthalmoscopic examination showed twelve pearl-like polypoid lesions studding the left fundus, 24 hours after reported difficulty of vision in that eye. One of these pedunculated "pearls" came dangerously near the macula, and it doubtless caused the acute disturbance of vision which brought the patient to examination. In the course of the next five months these lesions faded out or tended to fuse, while others appeared in new locations. The patient was receiving routine "promin" therapy and gave no evidence of recrudescence of leprosy.

This patient again reported visual disturbance five months after the first episode, and was then found to have fundus lesions in the right eye, which had been free of them on the previous occasion. A typical "pearl" type lesion was seen, located laterally to the macula. Like the others it seemed to be a waxy, creamy white, pedunculated polyp extending out into the vitreous humor, although it did not show any evidence of movement. At this time the original lesions, in the left eye, were still observed although their number had been reduced to seven and there was a slight clouding of the vitreous. Now, after a year, there still are lesions on the right fundus (Figs. 1-3).

CASE 2.—The patient, a white male, aged 34, discharged as an arrested case of leprosy, complained of a "spot" in the right eye on July 19, 1948. This spot moved in his visual field.

On examination a solitary but distinct pearl was seen on the fundus at 1000, about one disc-diameter from the head of the optic nerve. This polyp seemed to project from just beneath an artery. It was definitely anterior to the retina, though seemingly attached to it. The past history of this patient indicated a slight but similar disturbance of vision in the left eye many years ago. Since that time the left vitreous humor has been cloudy, and it still contained particles. Each iris was now the site of pearl-like lesions, being studded with two or more large formations together with fine seed pearls deep in the iris. There was also diffuse keratitis in the superior temporal quadrant of each cornea.

This patient had been without treatment for one year, although two weeks before the onset of the visual disturbance he had been admitted to the hospital for 72 hours complaining of a general malaise and a fever of undetermined origin. Promin treatment was begun after the fundus lesion was reported. When seen in the latter part of October 1948, the single pearl on the fundus had subsided and no further disturbance of vision was reported. The lesions in the iris were unchanged.

CASE 3.—The patient, a white male, age 60, was first examined in July 1948, when he had had leprosy for about 10 years. He has a lifetime history of myopia, with a vision of 20/200 corrected to 20/20. About 10 years ago he was examined by an oculist in Rangoon because he complained of seeing spots before the right eye, and he was told that he had retinitis, no further diagnosis or explanation being given.

Examination revealed madarosis of more than 50 per cent of each brow and all cilia, with thickening of the margins of the lids of both eyes. There was moderate invasion of the cornea, with capillaries observed extending from the limbus in each eye. Pigment granules were embedded on the posterior surface of the cornea. Leprous beading of the corneal nerve was seen only in the right eye. There was slight keratitis in each eye, confined to the superior temporal quadrant. The media were clear. No changes on the iris were seen.

The fundus, best observed with a minus 4.00 lens, was seen to be studded with a lumpy, waxy mass which approached the head of the optic nerve to within one disc-diameter. A few nodules protruded above the others in a polypoid form, but no single, symmetrical, tear-drop polyp was seen in either eye. This confluent, hobnail, waxy surface, which extended laterally, nasally, inferiorly and superiorly, left only the macular and disc areas free.

The patient had a distinct contraction of visual fields; whether or not this was present in 1939 has not been determined. Perimeter readings showed this constriction, using a 2-degree test object, to be within 35 degrees superiorly, 40 degrees nasally, 50 degrees inferiorly, and 70 degrees laterally, while color discriminations of 1-degree test objects of red, green, and blue were restricted to 25 degrees in all directions except laterally where it was limited to 50 degrees.

CASE 4.—The patient, a Mexican male age 41, was admitted in March 1947 with lepromatous leprosy. He showed complete madarosis of the eyebrows and cilia; lagophthalmos of both eyes; beading of the corneal nerves of both eyes; posterior synechia at 0600, left eye; filaments

floating in anterior chamber, right eye; deep vascularization of the cornea in the superior temporal quadrants, each eye; vision 20/30.

On the fundus of the right eye a small polypoid pearl leproma was seen at 0300, just below an artery. The lesion had a waxy surface and in form was similar to the pearls seen on the iris.

This patient died on September 9, 1948 and was autopsied on the following day. The right eye was removed.² Syphilis aortitis was found—this being the only case in the autopsy records of this institution in which anatomical evidence of syphilis was demonstrated. Many authors have contended that the lesions on the iris might be either tuberculous or syphilitic, and this particular case might possibly have had a luetic basis for the ocular lesions. However, in form and type they were precisely the same as those observed in the five other cases. Likewise of negative value were the autopsy findings with respect to tuberculosis; careful search of the lungs, glands and gastrointestinal tract failed to reveal any evidence of that infection.

CASE 5.—The patient is a negro male, age 50, with leprosy of neural type probably of 15 years duration. He has complete madarosis of both brows and cilia. He was first observed in January 1947, and also in a review examination in May 1947; no evidence of lesions on the fundus were found then. On January 30, 1948, the patient complained of blurred vision. He was then hospitalized because of a fever of undetermined origin; he presented no general manifestations of leprosy activity.

Examination showed six pearl-like, waxy lesions confined to the retinal area above the disc in the right eye between 1000 and 14000 (Fig. 5), while in the left eye there were at least 15 lesions scattered over the entire fundus (Fig. 4). These small polyps were all posterior to the equator, and some were observed between the disc and macula. When seen four months later the lesions were still present, although the vision was not as much disturbed as before.

CASE 6.—The patient, a white male age 29, was admitted to a general hospital in November 1948, with a blotchy, purplish generalized skin disease and had been presented as a typical case of erythema multiforme. A pathologist familiar with leprosy visited the hospital and commented upon the "typical tuberculoid leprosy case," and this diagnosis was sustained by biopsy. The patient was admitted to the National Leprosarium in January 1949.

A tuberculoid lesion extended into the left eyebrow, although no madarosis or other lesion of the ocular adnexa was found. On the globe and laterally each eye presented, beneath the conjunctiva and at the limbus, a localized increase in vascularity suggestive of the typical episcleral leproma in its earliest form. Grossly the corneas were clear, but the slit-lamp examination showed some increase of deep vascularity in the left eye and the corneal nerves were prominent though not beaded. This condition was restricted to the superior temporal quadrant. The iris of each eye showed fine wisps on the surface and several attachments of the dilator musculature were free of their pupillar insertions. Pigment particles

² "Army Institute of Pathology, March 9, 1949, No. 217655. Lymphocytes and plasma cells infiltrate the iris and ciliary body. With special stains acid-fast bacilli are demonstrable within large macrophages with foamy cytoplasm."

were seen on the anterior surface of each lens and the posterior surface of the cornea.

The left fundus was negative, but that of the right eye showed three small, white waxy pearls lying between the vessels extending toward 0200. The first nodule was approximately one disc-diameter upward from the optic nerve, and the second larger one less than three disc-diameters toward the periphery.

The epidemiological history of this last case is most interesting. Born in a nonendemic state of northwestern United States, the patient served in the Armed Forces stationed on a Pacific Island for 25 consecutive months, leaving that endemic leprous area in January 1944. Individuals afflicted with leprosy were observed by the patient while on duty there, but no other contact can be determined. Before concluding that his infection was acquired in adult life from that exposure, an intensive investigation of his family history should be made. The patient's mother died of "scarlet fever" when the patient was six years old and similarly afflicted. A family connection may exist with a French-Canadian or Minnesota focus.

The following tabulation summarizes the serological and x-ray findings on these cases. That the serology in leprosy is notoriously unreliable in the differential diagnosis of syphilis hardly needs comment here except for the fact that—unique in the history of this institution—the one case in the present series which came to autopsy showed anatomical evidence of lues. The history and physical findings of the other five patients do not warrant the diagnosis of syphilis, although the Kahn and other tests are positive in three of them.

Case	Kahn		Mazzini		Tuberculosis
	Admission	Present	Admission	Present	X-ray chest
1	No report	Positive	None	Negative	Negative
2	Positive	Positive	Positive	Positive	Negative
3	Positive	Positive	Positive	Positive	Negative
4	Negative	Negative	Negative	Negative	Negative
5	Positive	Positive	Positive	Positive	Negative
6	Negative	Negative	Negative	Negative	Negative

SUMMARY

In summary, six leprosy patients, all males, have been observed who showed similar lesions of the fundus. Three of the cases had the condition in both eyes, while in the other three it was restricted to one eye. One of the latter patients had a single

lesion on the fundus of the right eye, while his past history and the presence of floating particles in the vitreous humor of the left eye are strongly suggestive of its prior involvement. Of these six men, five admittedly have had leprosy for at least fifteen years. The diagnosis in the sixth case was established only within the past year, and the history is nebulous. The first ocular manifestations reported were the blurring of vision or observation of particles in the patient's visual field. In two discharged cases an unexplained febrile reaction followed by the ocular manifestation were the only precursors of a reactivation of leprosy.

CONCLUSIONS

1. Leprous lesions occur on the fundus oculi in certain cases.
2. Any case may pass through a phase of this relentless disease when these distinctive pearl-like lesions can be clearly seen.
3. The findings and histories of four of the six cases observed suggest that the condition is frequently bilateral.
4. Positive results of serological reactions in three patients, and the finding of luetic aortitis in one of them, do not in our opinion warrant the assumption that the fundus lesions observed were other than leprosy in character.
5. The fundus lesions observed were identical in all of these cases, although they were classified variously, as lepromatous, "mixed neural," and tuberculoid.

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REFERENCES

1. BULL, O. B. and HANSEN, A. G. The leprosy diseases of the eye. Christiania, 1873 (cit. Klingmüller).
2. COCHRANE, R. G. Practical Textbook of Leprosy. Oxford University Press, 1947, p. 115.
3. ELLIOTT, D. C. Report of leprosy lesions of the fundus. *Internat J. Leprosy* **16** (1948) 347-350.
4. JEANSELME, ED. La Lèpre. Paris, G. Doin & Cie, 1934, pp. 339-340 and 352.
5. KLINGMÜLLER, V. Die Lepra. *Handb. Haut-u. Geschl.-Krank.*, vol. 10, pt 2, Berlin, Julius Springer, 1930, p. 436.

6. TRANTAS. Lèpre oculaire. *Rec. d'Ophth.* **20** (1898) 452 (cit. Jeanselme); (also cit. Duke-Elder, *Textbook of Ophthalmology*, C. V. Mosby, 1942, vol. 2, p. 2322).
7. UCHIDA, M. On the clinical and histological study of leprous eyes and their symptomatic treatment. *La Lepro* **2** (1931) 34-72 (English abst., suppl. 35-39, with 8 photomicrographs).

DESCRIPTION OF PLATE

PLATE 14.

FIGS. 1 to 3. Numerous pearl-like lesions on the fundus of the right eye, Case 1. These lesions are still present, one year after the first observation. Black central spot and surrounding light area is reflection of carbon-arc light used in Nordsen Retinal camera. For orientation, the same lesion is indicated by an arrow.

FIG. 4. Showing the left disc and vessels, with numerous smaller polyps scattered over the fundus, Case 5.

FIG. 5. Numerous waxy lesions just above the disc of the right eye, Case 5.

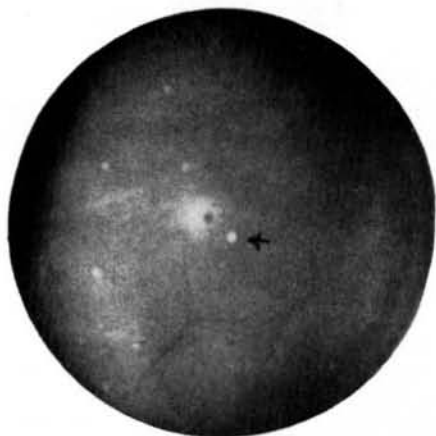


FIG. 3

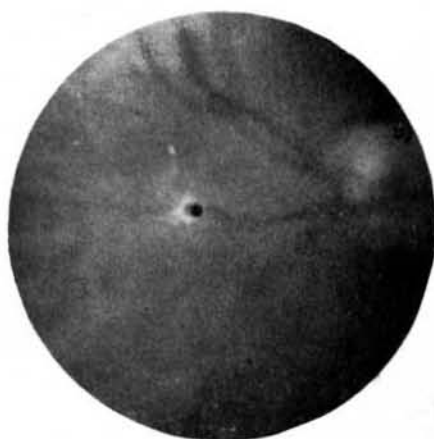


FIG. 5

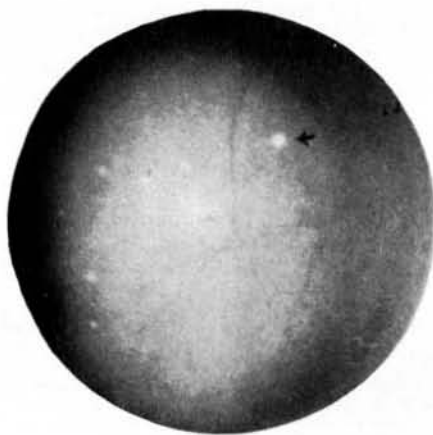


FIG. 2

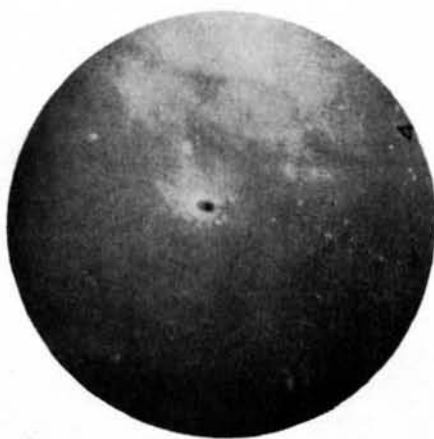


FIG. 4

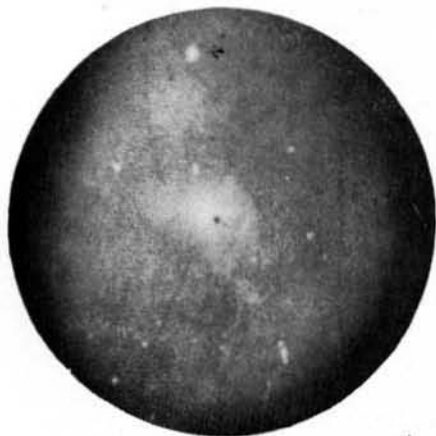


FIG. 1