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SYMPTOMATOLOGY OF LEPROSY.

BY

HOWARD FOX, M.D.,

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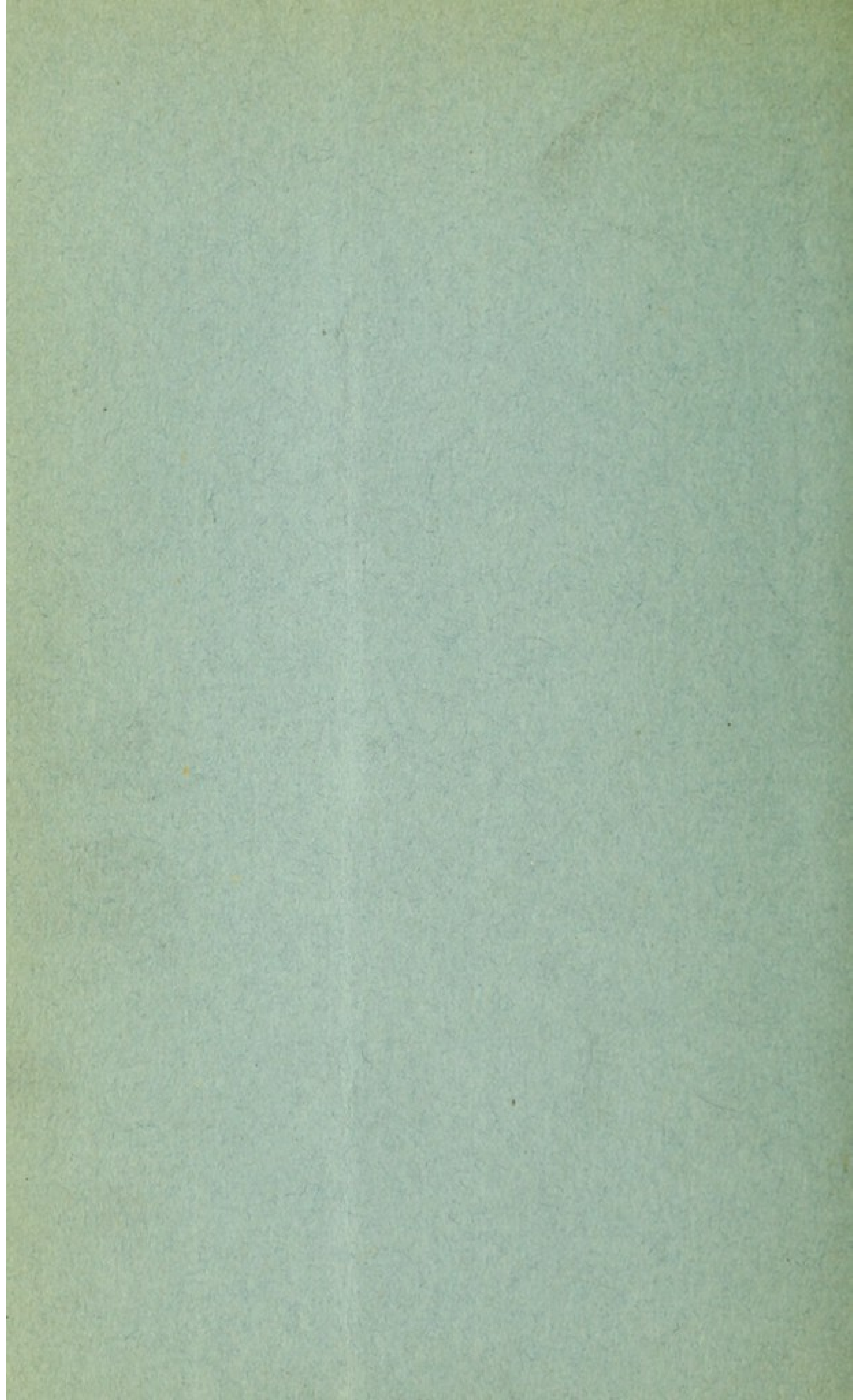
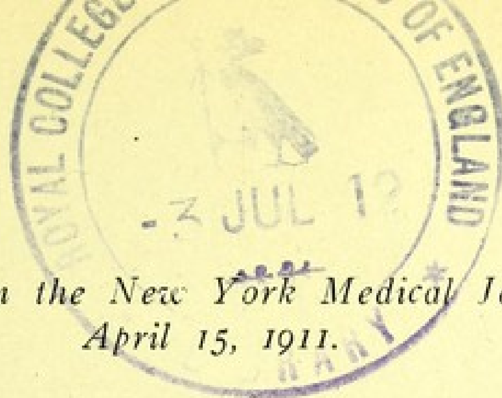
April 15, 1911.

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SYMPTOMATOLOGY OF LEPROSY.*

By Howard Fox, M. D.,
New York.

As the number of lepers in New York city appears to be slowly but steadily increasing, it would seem that a knowledge of the symptoms of the disease is becoming of practical as well as of theoretical interest to the profession of this city.

In briefly presenting the numerous and varied symptoms of leprosy it is most convenient to describe the two distinct and classical types of the disease, namely, tubercular leprosy, in which the skin and mucous membranes are chiefly affected, and anæsthetic leprosy, in which the nervous system is principally involved. Macular leprosy is described by some writers as a third type of the disease, and in a few cases the macules constitute practically the only symptom. In most cases, however, they are simply forerunners of either the tubercular or anæsthetic stages and need not be discussed as a separate type of the disease. In a large number of the cases there is a combination of symptoms constituting what is called the mixed type of leprosy. Practically all of the cases seen in New York are of the tubercular or mixed type, the cases of pure nerve leprosy being extremely rare. Indeed, among thirty cases which I have seen in New York city during the past year I can recall having seen

*Read before the New York Academy of Medicine, December 29, 1910, in a "symposium" on leprosy.

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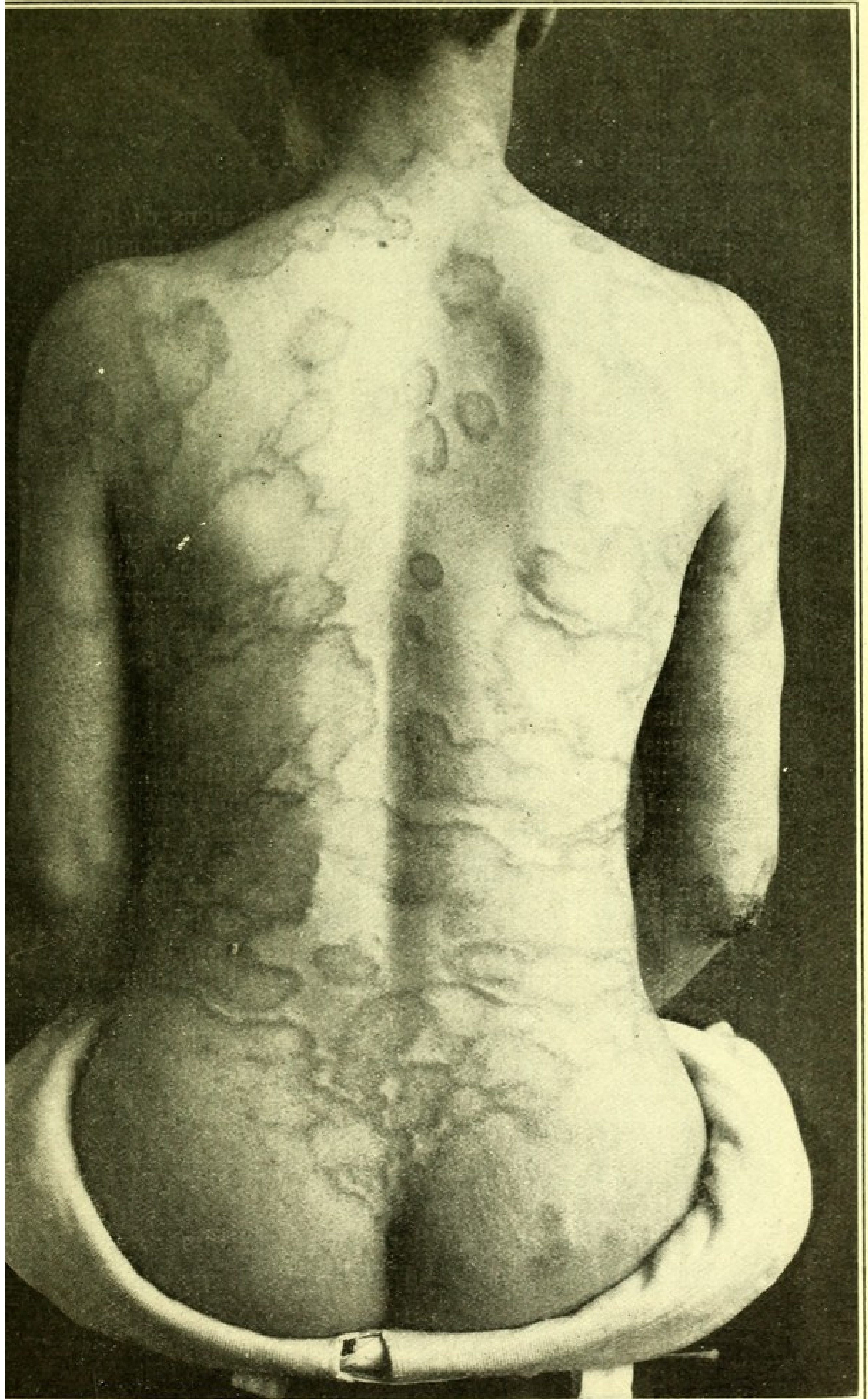
but one case of pure anæsthetic leprosy presenting well marked deformities or mutilations.

The symptoms of leprosy do not evolve with as much regularity as they do in syphilis for instance. and an artificial description of the stages of the disease seems hardly warranted. We do not know in what manner the infection occurs, as there is no primary lesion in leprosy that is comparable to the chancre of syphilis. It is therefore difficult or impossible to judge of the exact time when a certain patient has become infected. This may be estimated in certain cases in which, for instance, a person has resided for a short time in a leprous country and then returned to a nonleprous region and later presented the symptoms of leprosy.

The period elapsing between the time of infection and the first manifestations of the disease the so called period of incubation, has no parallel as regards length of time with any other infectious disease. Thus it may vary from three months to ten, twenty, or even thirty-two years, as in the case reported by Hallopeau.

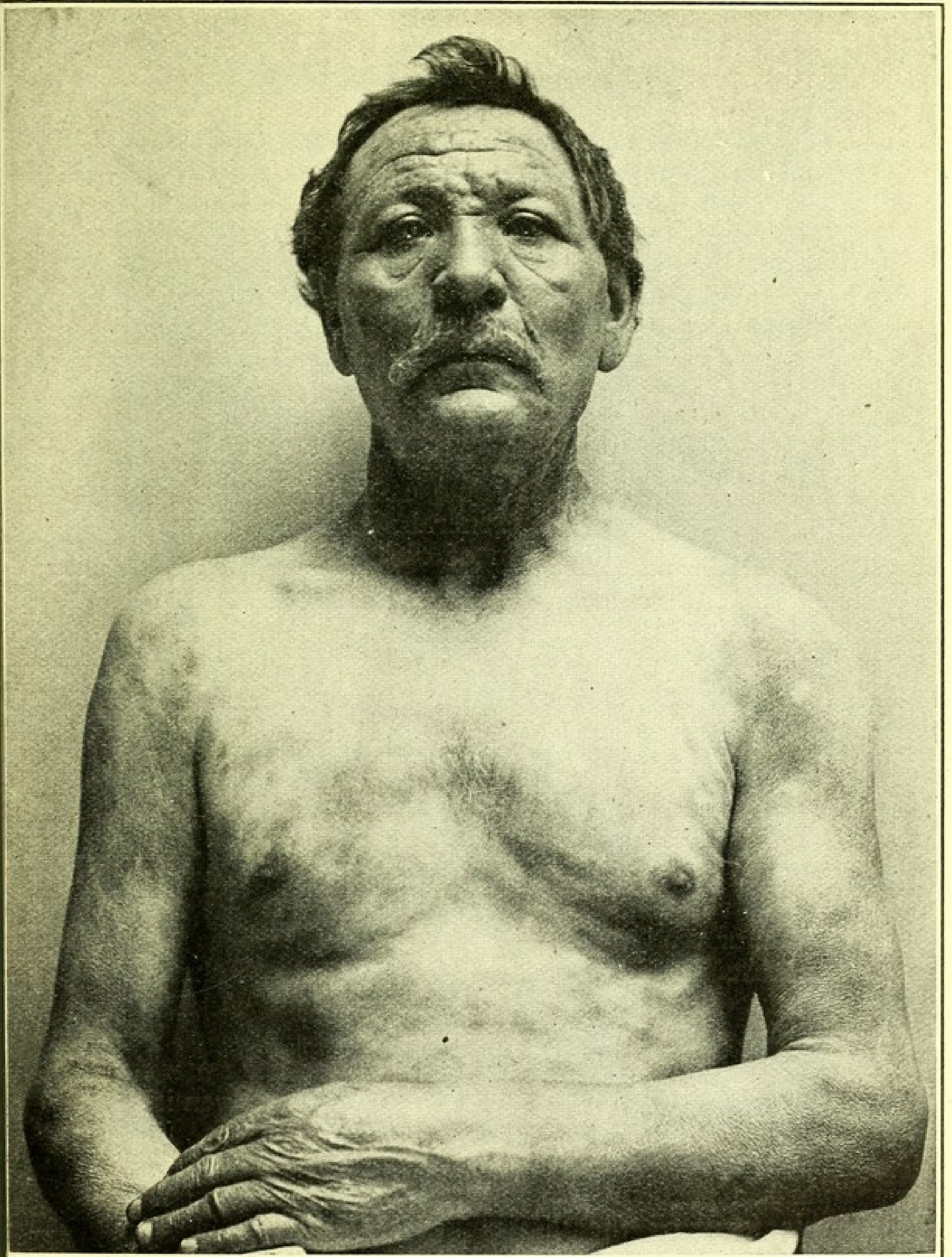
The question as to whether the period of incubation represents a true germination of the bacilli, or only a long period of latency or hibernation, is one that need not be discussed at this point.

The general invasion of the body by the bacilli or their poisonous products may be marked by a group of symptoms or prodromata, that are not in themselves in any way diagnostic of leprosy. They include a rise of temperature, which may be mild and gradual, or sudden and severe, as in pneumonia. The fever may be accompanied by general malaise, headache, vertigo, drowsiness, severe sweating, and various rheumatoid pains. The patient himself is apt to ascribe such symptoms to a "bad cold" or an attack of malaria. These symptoms may be fol-



Lazaro Hospital with an erythematous eruption that looked strikingly like an erythema multiforme. It certainly would never have suggested the diagnosis of leprosy to my mind. After a number of days or weeks the early erythematous eruption disappears and may reappear later, accompanied by fever and other general symptoms. After several repeated attacks in such a case the macules become permanent or "fixed," their color deepens and does not tend to disappear on pressure. In many cases, especially of the anæsthetic type, the macules make their appearance insidiously, without any constitutional symptoms, and are fixed and pigmented from the outset. In the majority of the cases, certainly as we see them in New York, the macules have become permanent by the time medical aid is sought.

The macules vary in size from a pea to that of the palm of the hand, or may occupy large areas of the body, especially in the anæsthetic type. Their borders may be sharply or poorly defined. They may clear up in the centre and form circinate lesions, or gyrate figures from a coalescence of several such rings. The serpiginous tendency is chiefly noted on the lower extremities. The centres of the macules may present a loss of pigmentation, while their borders are superpigmented, and sometimes upon the leprous patch a vitiligo develops in the shape of white and depressed discs. The macules may be hyperæsthetic or itchy at the outset. Sooner or later they become anæsthetic, the anæsthesia being most marked in the centre of the patches, while the superpigmented border is often, for a time at least, hyperæsthetic. The macules are roughly symmetrical, the symmetry being more marked in the anæsthetic type. The sites of predilection are the face, extensor aspects of the extremities, the buttocks, and the back. The lesions



G. 3.—Case showing nodules upon the forehead and macules upon the arms and trunk
Patient of Dr. George Henry Fox.

are very rare upon the palms and soles and are almost never seen upon the scalp. There is generally noticeable absence of sweating over the macules, and after considerable time there may be slight branny desquamation. While the macules may exist unchanged for many years they are usually followed sooner or later by the symptoms of tubercular or anæsthetic leprosy or a combination of both types.

The first appearance of tubercles and indeed the various crops of tubercles are often ushered in by febrile symptoms. In some cases they appear insidiously without such general manifestations. They may appear in the form of diffuse flat infiltrations or as circumscribed nodules, varying from a small pea to a cherry or pigeon's egg in size. They are generally situated within the skin, although at times they may be beneath the skin, as in the shotlike nodules that may occur in the lobule of the ear. The tubercles may be engrafted upon or appear between the already existing macules. Their color varies from a light red, violaceous, or yellowish hue to a dark brown or reddish brown color. The surface of the tubercles is generally greasy from an excessive sebaceous secretion; it may be hyperæsthetic or moderately anæsthetic, and shows as a rule an absence of hairs. Indeed the falling of the eyebrows due to tubercular infiltration constitutes one of the earliest symptoms of nodular leprosy. The tubercles are found in the greatest number upon the face and the extensor aspect of the extremities. Upon the face they are chiefly noted upon the forehead, nose, chin, and ears, and their presence imparts a peculiar frowning or leonine expression to this form of the disease, which when once seen, can never be forgotten. For some unknown reason the scalp seems to enjoy a remarkable immunity

to the ravages of leprosy, as it is never invaded by tubercles, and only in the rarest instances by ma-

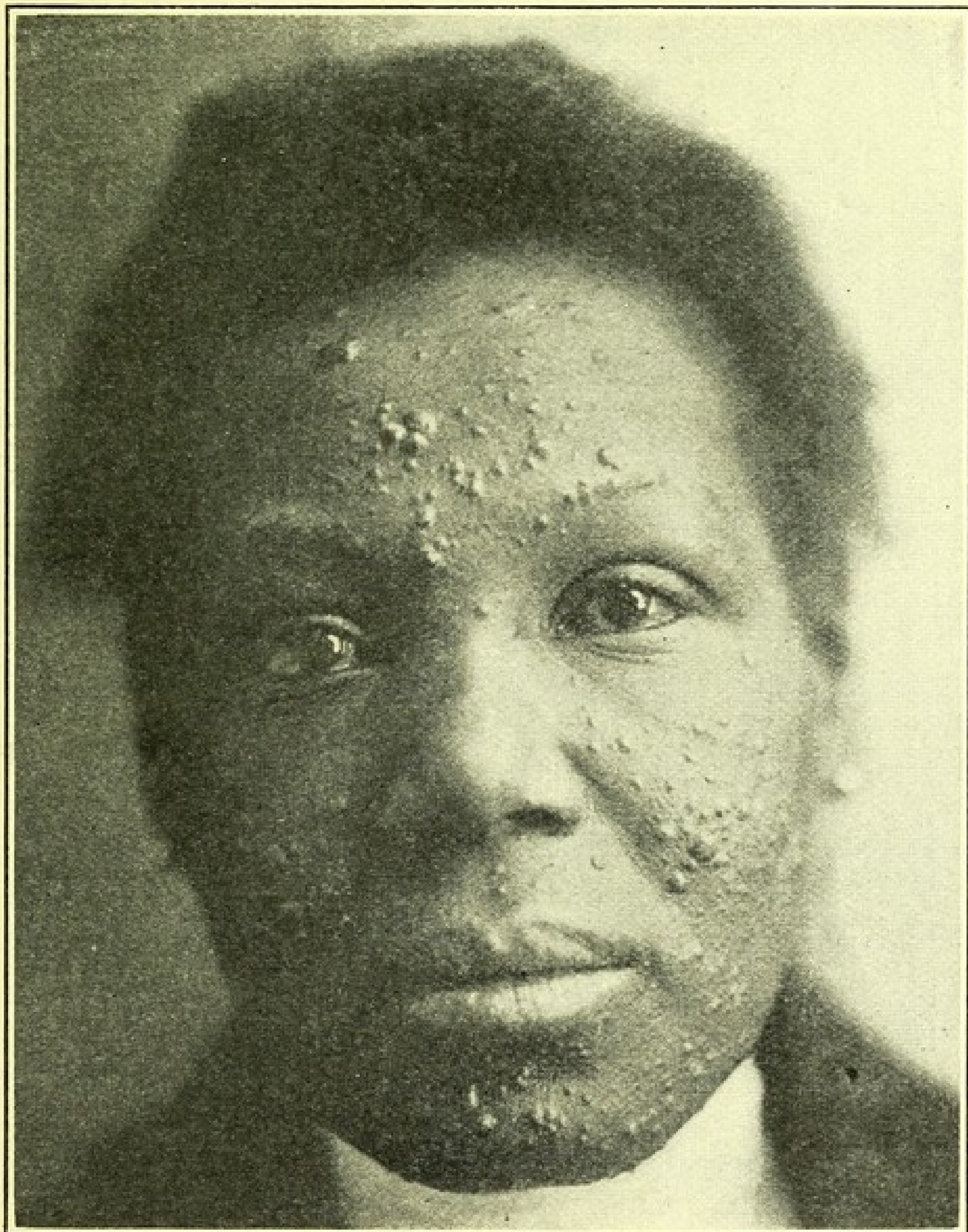


Fig. 4.—Nodular leprosy, showing lesions of a small and unusual type. Presented by Dr. J. McF. Winfield before the New York Academy of Medicine, December 29, 1910.

cules. The palms and the soles are also very rarely the seat of tubercular infiltration. The tubercles

are abundant upon the backs of the hands, elbows, knees, and about the ankles. As the last phalanx of the hand is spared for a time the infiltration of the first two phalanges gives the fingers a peculiar fusiform appearance. Upon the feet and ankles it is impossible to distinguish individual nodules, as the skin of this region is often the seat of a diffuse, hard œdema, presenting the picture of true elephantiasis. The nails, especially of the fingers, are often spared for a considerable time.

During the exacerbations of leprosy which are of frequent occurrence it is often noticed that the tubercles become reddened, swollen, and tender. At the end of these attacks some of the lesions are found to have decreased in size while new tubercles will have formed in other localities. The usual evolution of the tubercles is to slowly disappear by absorption leaving a deep stain or a slight cicatrix or to soften and ulcerate. In a few cases they undergo a fibrous transformation. Ulceration of the nodules occurs more frequently in tropical countries and among the more ignorant and uncleanly class of patients. The ulcerations cover large areas at times and encircle an entire limb. They eventually heal and leave apigmented scars that may be somewhat anæsthetic and are generally surrounded by a superpigmented border.

The pharynx, palate, uvula, epiglottis, dorsum of the tongue and less often the mucous membrane of the mouth are sooner or later attacked by diffuse nodular infiltrations. These may persist for a time or become absorbed, ulcerate or cicatrize. As a result of ulceration there is frequently distortion of the epiglottis or uvula. Perforation of the palate is very unusual. Later the interior of the larynx and even the trachea may be invaded by the leprous deposits. A peculiar pallor is shown by the mucous

membrane of the larynx and pharynx. One of the earliest symptoms of leprosy is a change in the voice, which may at first be slightly hoarse and later

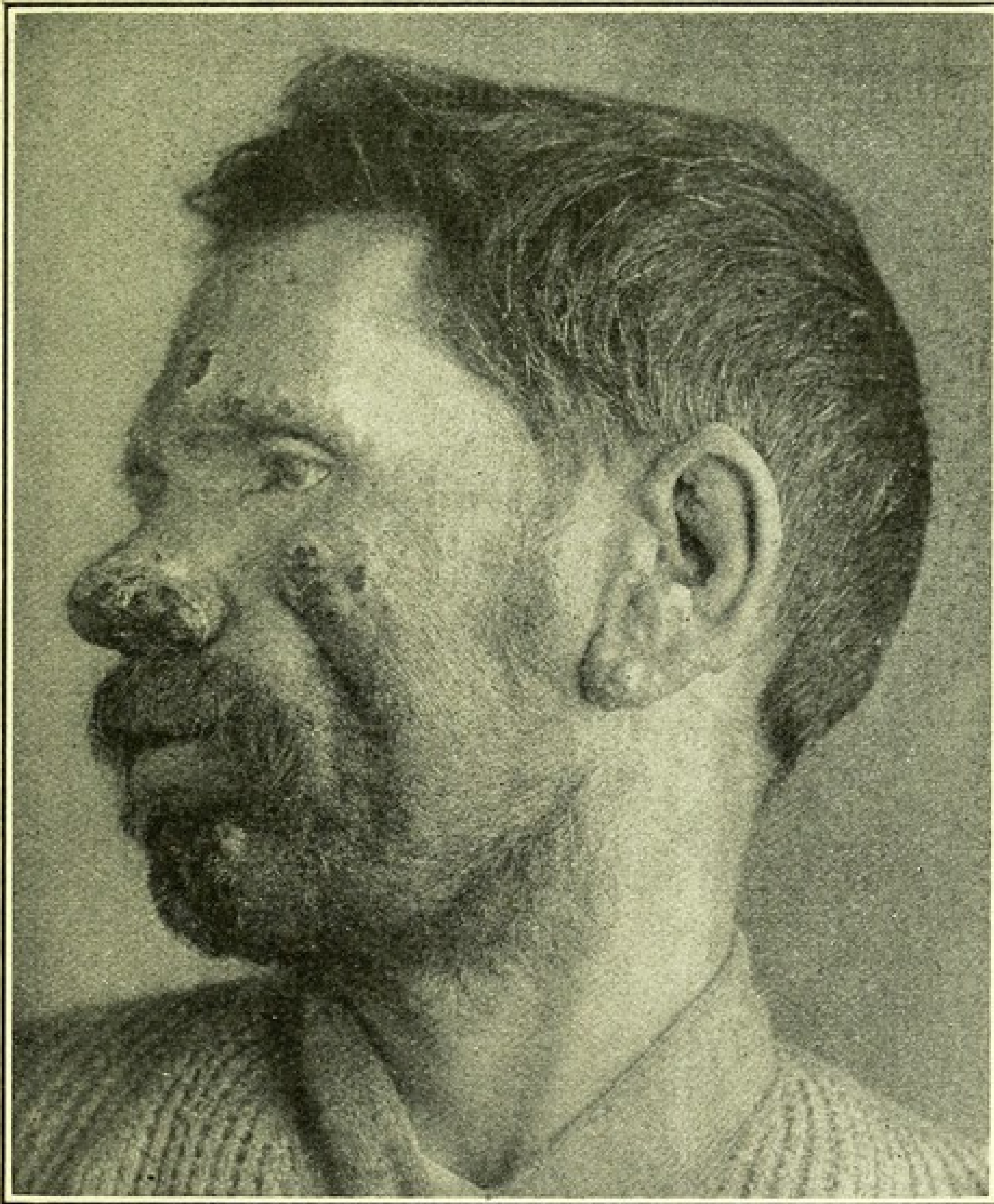


FIG. 5.—Nodular leprosy showing ulceration of lesions. Patient presented by Dr. Howard Fox before the New York Academy of Medicine, December 29, 1910.

becomes harsh, sibilant, or whispering, and often interrupted by attacks of dyspnœa.

The visual apparatus is frequently involved in

tubercular leprosy. The lesions involve the anterior segment of the globe, especially the sclerocorneal junction and the ciliary region. There may be a keratitis which gradually invades the entire cornea, or a tubercle may form at the limbus and penetrate into the anterior chamber and destroy vision. Iritis is also common and is most frequently of plastic type.

The inguinal glands and less often the axillary and cervical glands are enlarged in tubercular leprosy and increase in size during the exacerbations of the disease. They are not as firm as the glands of syphilis and do not suppurate in the latest stages of the disease.

The urine often contains albumin and casts but no lepra bacilli, the changes in the kidney being due to an ordinary nonleprous nephritis. While the bacilli are as a rule disseminated in the internal organs, especially the liver, spleen, and bone marrow, they do not produce any characteristic symptoms except at times an enlargement of the spleen or possibly of the liver. A double epididymoorchitis is fairly common and presents a compact mass with smooth or nodular surface, with or without any accompanying hydrocele. In the rare cases in which fistulæ form, there is probably a secondary infection from the tubercle bacillus. In the majority of cases the disease causes a diminution of sexual power and desire. In women the menses become irregular and finally cease. In the case of young girls this function is not established at all.

Among other symptoms of tubercular leprosy should be mentioned a general darkening of the skin, a dusky cyanosis of the fingers, the appearance of occasional flaccid bullæ, and enlargement of the ulnar nerve.

The course of tubercular leprosy is in most cases

chronic, the average duration of life being about from eight to ten years. The most favorable ter-

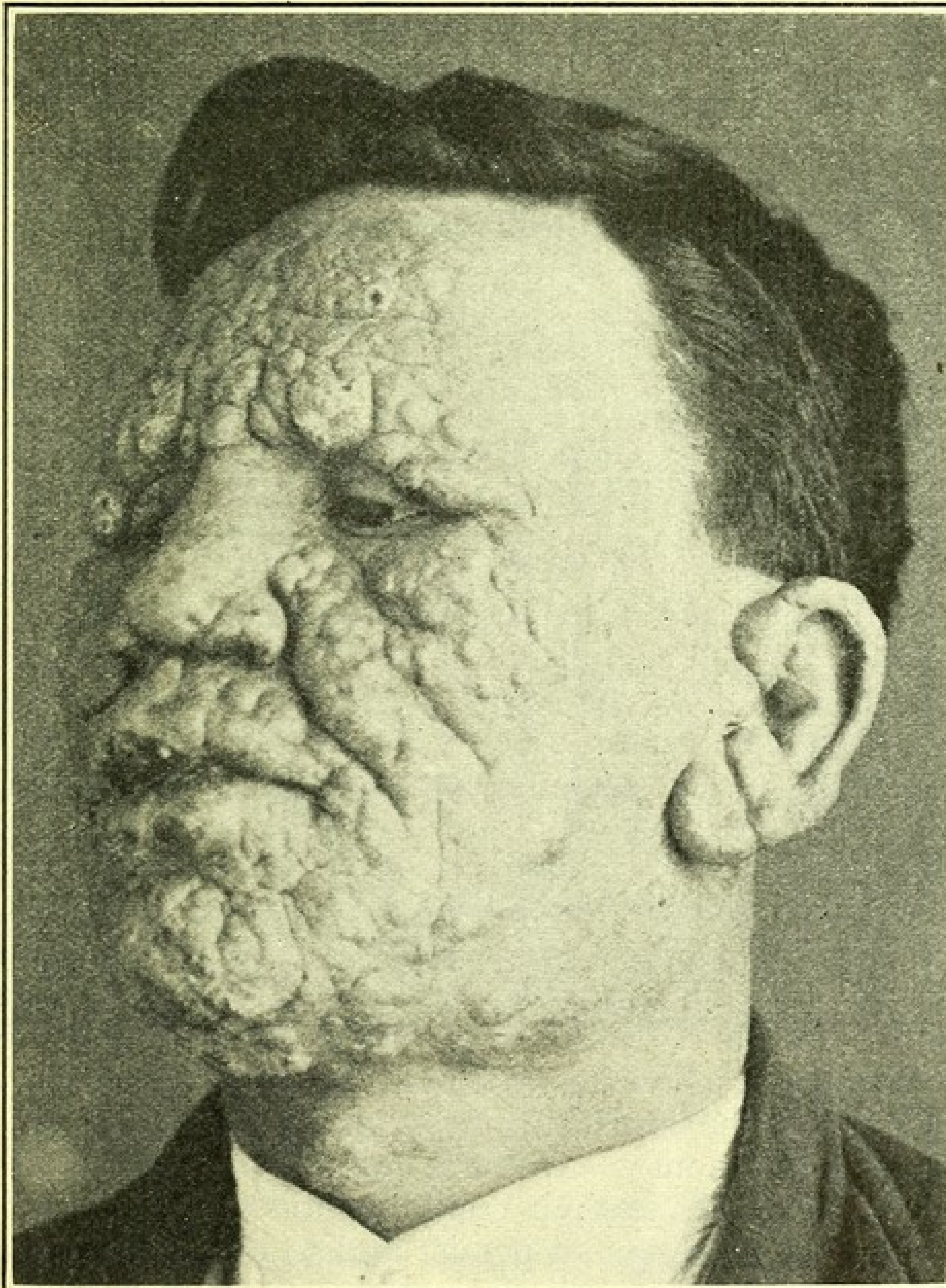


FIG. 6.—Nodular leprosy of three years' duration. Presented by Dr. Howard Fox before the New York Academy of Medicine, December 29, 1910.

mination of the form of the disease is a change to the anæsthetic type in which case the symptoms

ameliorate and the patient's life is often considerably prolonged. The tubercular leper in the terminal stages of his affliction is indeed an object of pity. The face is distorted by a mass of tubercles, many of them are covered with crusts and ulcers. There is a foul discharge from the nose and general fœtid odor from the lungs and skin. The voice is lost, sight is destroyed, and of the special senses hearing alone remains. The patient is extremely cachectic and weak and suffers from continual diarrhœa, and is robbed of his sleep by intense neuralgic pains. In spite of such a terrible condition the mind of the leper remains practically unaffected up to the time of his death, which is generally due to marasmus, diarrhœa, stenosis of the larynx, or an intercurrent disease or complication such as pneumonia or pulmonary tuberculosis.

The onset of anæsthetic leprosy may be accompanied by febrile symptoms as in the tubercular type. It is, however, more apt to appear insidiously and make itself manifest by a macular or bullous eruption or by various disturbances of sensation. The macules, according to Impey, are not due to the presence of bacilli in the skin but to vasomotor action on the terminal branches of the cutaneous nerves. The same is probably true of the bullous eruption in which the lesions are as a rule free from lepra bacilli. "While the cutaneous eruption in nerve leprosy," writes Morrow, "is not so essentially a part of the morbid process as in tubercular leprosy, the macules exhibit a greater variety of aspect, especially in their configuration and coloring." They are more apt to persist and to clear up in the centre and form vitiligoid patches, especially in the dark skinned races. The hairs upon the macules do not fall but are more apt to become white.

The bullous eruption is more often encountered

in the early stage of the disease, the lesions varying in size from a split pea to a cherry. They may appear upon any part of the body except the scalp, but are seen upon the hands, elbows, knees, ankles, and also the palms and soles. The bullæ soon rupture, dry, and form crusts followed by cicatrices.

The symptoms in general of the anæsthetic type

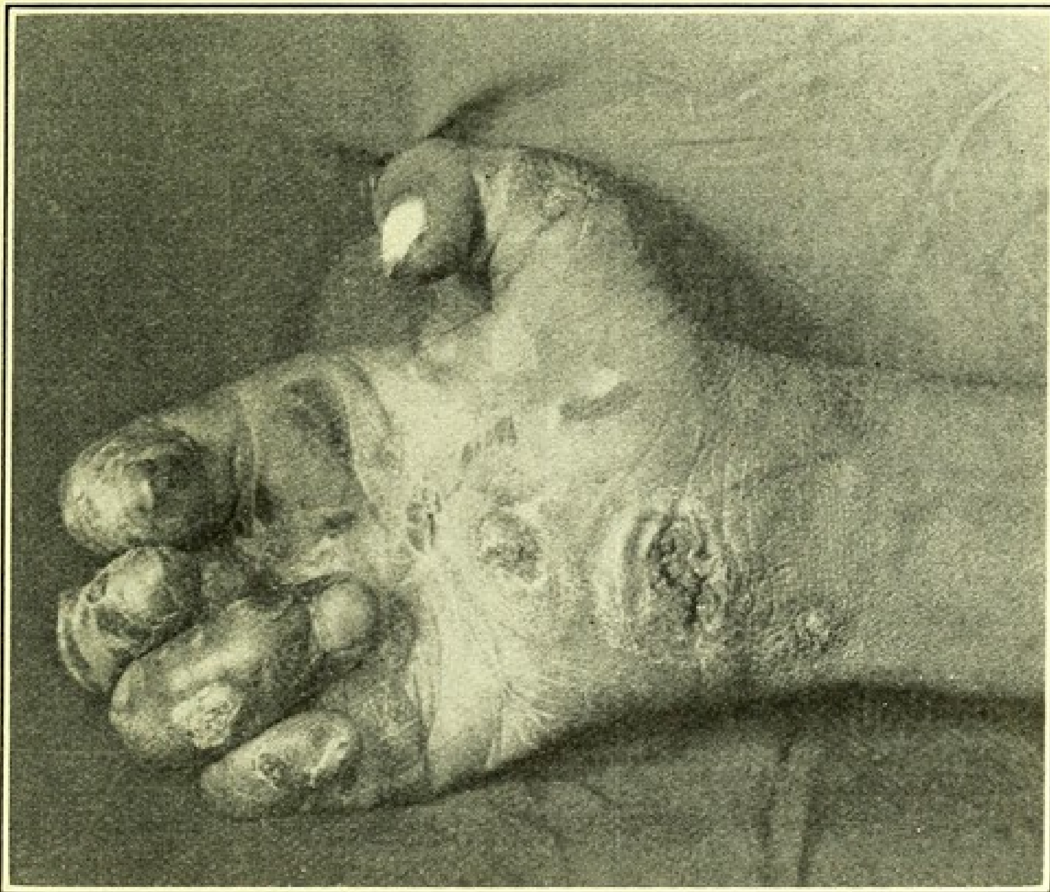


Fig. 7.—Anæsthetic leprosy showing "claw hand" and ulceration. Patient presented by Dr. J. McF. Winfield before the New York Academy of Medicine, December 29, 1910.

are those of a peripheral neuritis causing various sensory and trophic manifestations. In contrast with the tubercular type of the disease which is characterized as von Bergmann says by marked hyperplasia, the noticeable changes in anæsthetic leprosy are those of atrophy.

One of the most important and diagnostic symptoms is an increase in the size of some of the nerve trunks that occurs at a very early period of the disease. The greatest changes are observed in the nerves that are superficially situated such as the ulnar and median nerves. They are changed to cylindrical or fusiform or beaded cords and may at times attain the thickness of the little finger. The ulnar enlargement is especially characteristic and may at times be felt in its entire course from the elbow to the axilla. At the outset, the nerve trunks are painful upon pressure but later become completely insensible to pain.

The disturbances of sensation in leprosy may be confined to the macules or exist independently of them. The characteristic anæsthesia, which represents a complete disorganization of a nerve trunk, is invariably preceded by irritative symptoms. There is often hyperæsthesia which may be intense or there may be pruritus, neuralgic, or shooting pains or various forms of paræsthesia. There may be vasomotor symptoms such as cyanosis, or secrettory disturbances such as interference with sweat secretion.

Sooner or later anæsthesia makes its appearance, and is especially marked in the extremities. It is first noticed in the fingers and toes, and then travels upward toward the trunk. It is often bandlike at first, but later involves the entire circumference of a limb. Part of the area of anæsthesia is fixed, while the rest, according to Jeanselme, varies in intensity from day to day. There is often dissociation of sensation, that of temperature and pain disappearing, for instance, while the sensation of touch remains. Sensation may also be delayed, as when the prick of a pin is felt after an interval of several seconds. The anæsthesia becomes com-

plete at last, so that the patient may be able to cut off portions of the hands or feet without experiencing the slightest pain.

Muscular atrophy is one of the constant and striking symptoms of anæsthetic leprosy. There is a diminution in force in proportion to the waste of muscle tissue. At times there is a true paralysis.

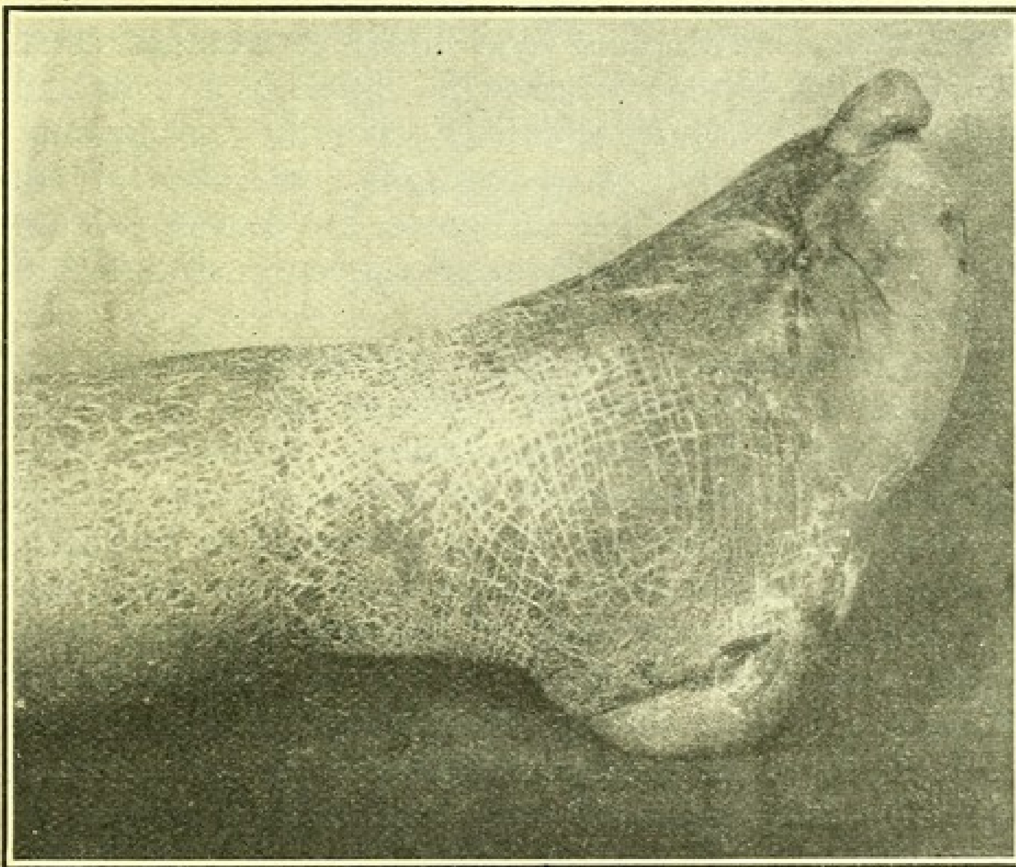


Fig. 8.—Anæsthetic leprosy, showing characteristic mutilation. Same patient as Fig. 7.

Atrophy is noted at an early stage in the thenar and hypothenar regions. From atrophy of the first dorsal interosseous a characteristic hollowing of the back of the hand is produced. One of the commonest deformities is the "leper claw" produced by tendinous retractions and muscular atrophy. It is formed by extension of the first and a flexion of

the second and third phalanges. The palm is flattened and the thumb is on the same plane with the fingers. In spite of this deformity the use of the hands is far from being lost, especially if the sensation of touch remains.

The superficial muscles of the face are frequently atrophied and can give the patient an appearance that is as characteristic as the leonine face of the tubercular leper. The eyes have a peculiar stare and the lids cannot be closed. The lips are flaccid and pronunciation of labials is difficult. On account of eversion of the lower lip, saliva may flow from the mouth. The face becomes an expressionless mask, and the patient's appearance is stupid and doleful.

In some cases the process of anæsthetic leprosy does not go beyond the production of muscular atrophy and tendinous retraction. In other cases, where the disturbance of nutrition is very great, there are bone changes which give rise to frightful mutilations. The loss of bone may occur from necrosis or from a process of interstitial absorption. In the case of the fingers the second phalanx is apt to be the first to disappear, so that the fingers appear to have only two phalanges. In some cases the nails are preserved with remarkable tenacity in spite of extensive loss of bone. Finally, the hands and feet become veritable stumps, the shape of the feet being compared to that of a pestle or drum stick. Large trophic ulcers, among them the so called perforating ulcers, may add to the patient's distress.

The mucous membranes are much less often involved in the anæsthetic than in the tubercular type of leprosy. As the lower lid is everted the conjunctiva is unprotected and soon becomes inflamed. Corneal opacities may form and destroy the sight.

Ulceration and destruction of the septum, with consequent sinking of the nose, is not uncommon. Due to loss of tactile sensation there is often difficulty in swallowing, the food being regurgitated into the nostrils.

The course of anæsthetic leprosy is decidedly chronic, the average duration of life being about fifteen years. In some cases the process appears to come to a standstill and the patient is apparently cured. A number of such cases have been reported by Dyer, of New Orleans. In a few cases the anæsthetic type changes to the tubercular form of the disease.

The termination of a severe case of anæsthetic leprosy presents a totally different picture from that of the tubercular type, but one that may be equally pitiful and distressing. The anæsthetic leper is emaciated and cachectic. He is bedridden and has to be fed by attendants. The nose is sunken, the sight is extinguished, and saliva pours from the paralyzed lips. The patient suffers from a sense of cold, loss of appetite, insatiable thirst, and severe neuralgic pains. The numerous ulcerations add to the natural disagreeable odor of the leper and the deformities of the limbs make them scarcely recognizable as those of a human being. The patient's mind is dull, but by no means lost. Death is generally due to marasmus, amyloid degeneration of the viscera, diarrhœa, or to an intercurrent disease. It is rarely due to pulmonary tuberculosis.

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