A STUDY OF MYOSITIS INTERSTITIALIS LEPROSA

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Muscular change in leprosy, which is frequently observed, is primarily an atrophic condition. As the atrophy is usually based upon the lack of motor and trophic nerve supply, it is commonly classified as (*a*) disuse atrophy, or (*b*) neuarotrophic atrophy.

Recently I have made a histologic study of the skeletal muscles in leprosy, the materials for which were biopsy specimens or tissues obtained at autopsy. Most of them showed the muscular atrophy, including neuritis and angiitis leprosa in the muscle.

Rarely, however, cases with myositis interstitialis leprosy have been encountered together with the muscular atrophy. The following four cases are examples.

ILLUSTRATIVE CASES

CASE 1. S.U., female, age 21 in 1955, moderately advanced lepromatous leprosy. The patient developed the disease in 1947, and came to the leprosarium in 1948. At the time of admission she showed lepromatous infiltrations on the face, forearms and legs, and saddle nose and a slight degree of alopecia. Between June 1948 and July 1956 the patient was treated with Promin and Promizole. During the course of the treatment, erythema nodosum leprosum reactions frequently occurred. However, the improvement of the disease was so marked that there was regrowth of the hair. In October 1956 the lepromin reaction was found to have converted from negative to weakly positive. Shortly before that the patient had noticed tumors of both calves, arranged almost symmetrical.

Examination of the left leg revealed in the calf muscle a sharply-defined, palpable tumor of the size of a pigeon's egg, cartilage-like in hardness. It was not adherent to the skin, and it could not be identified roentgenologically. A biopsy specimen was removed from this muscle tumor.

Histology.—There is marked hypertrophy of the fascia, the collagenous bundles of the fascia showing thickening, and granulomatous infiltrations are seen around the vessels (Fig. 1). The inflammatory infiltration is found mainly on the surface of the fascia, where it composed chiefly of histiocytes and lymphocytes with occasional eosinophilic leucocytes.

The muscle bundles show various degrees of degeneration. In the more severely affected areas the muscle bundles show fragmentation. Vacuolar degeneration is seen, especially marked at the margin of the fascia; the vacuoles are of various sizes. This vacuolar degeneration, it is believed, may be caused by a hydropic process due to the inflammation or to blood stasis.

The inflammatory infiltrate between muscle bundles (Fig. 2) consists of histiocytes, lymphocytes and eosinophilic leucocytes, with an occasional Langhans giant cell. Fatty degeneration is not marked, either in the muscle bundles or elsewhere. In the inflammatory infiltrates are found acid-fast bacilli, especially in the form of large round globi (Fig. 3), but not many otherwise.

CASE 2. J.S., male, age 53 in 1958, far advanced lepromatous leprosy. Admitted in June 1949. At that time there existed multiple large firm nodules on the face and extremities, with infiltrations of practically the whole skin. Alopecia, ocular phthisis, saddle-nose, ulcer of the soft palate, claw-hand on both sides, etc., were also present. Bacilli were abundant in smears from the nasal mucous membrane and the skin. Treatment was started on admission, Promin and Promizole being used, but after 1952 the patient was psychotic and refused treatment. He died of gangrene of the lung in June 1958.

Autopsy findings: The whole of the skin was involved in diffuse lepromatous infiltration. Lepromatous lesions were also found in the liver, spleen and testicles. The left lung showed gangrene.

Histology.—Examination of a specimen from the calf muscle shows that the muscle bundles are markedly shrunken and separated from each other (Fig. 4), with in places inflammatory infiltrations. The fibers exhibit, widely, effacement of the transverse striations and, in some places, vacuolar degeneration as well. Fatty degeneration is not noteworthy. In proportion to the degree of the atrophy of the muscle fiber, there is increase of the nuclei of the sarcolemma—atrophic nucleus proliferation—and occasionally the proliferated nuclei are so arranged as to form a plate or mass of nuclei. The interstitium between such atrophied muscles, especially around the vessels, is invaded by inflammatory infiltrates which are composed of histiocytes or macrophages and lymphocytes. Many acid-fast bacilli, located in macrophages, are found scattered throughout the section; and they are also located in the endothelial or adventitial cells of the blood vessels and in the nerves.

DESCRIPTION OF PLATE

(The magnifications given refer to the pictures before about 20 per cent reduction in reproduction.)

FIG. 1. Case 1. Marked hypertrophy of the fascia, and atrophy and vacuolar degeneration of the muscle bundles. The inflammatory infiltrate in the fascia and the interstitium of the muscle consists mainly of histiocytes, lymphocytes and a few eosinophilic leucocytes. Hematoxylin-eosin stain; 50x.

FIG. 2. Case 1. In the inflammatory infiltrate in the interstitium of the muscle, histiocytes, lymphocytes and an occasional Langhans giant cell are present. Degeneration of the muscle fibers is to be seen. Hematoxylin-eosin stain; 200x.

FIG. 3. Case 1. Acid-fast bacilli are found in the inflammatory infiltrates, in the form of large round globi (see arrows). Ziehl-hematoxylin stain; 200x.

FIG. 4. Case 2. Muscle bundles are markedly shrunken and more or less separated, and the nucleus prolifieration of atrophy is present. Hematoxylin-eosin stain; 50x.

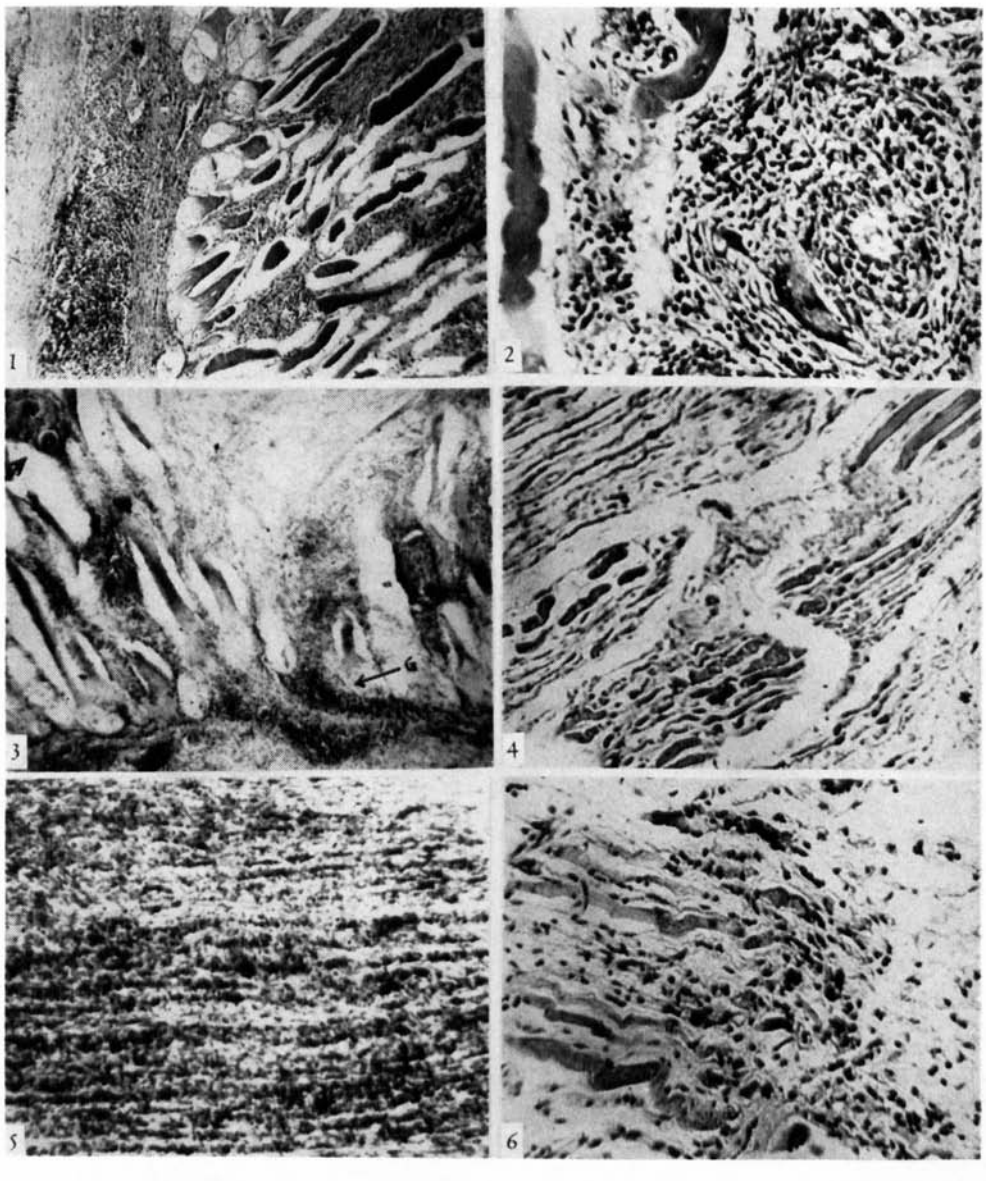
FIG. 5. Case 3. The original structure of the muscle cannot be distinguished in this preparation, but it is indicated by the arrangement of the nuclei. Acid-fast bacilli are scattered. Ziehl-hematoxylin stain; 50x.

FIG. 6. Case 4. Muscle bundles are in various stages of atrophy. Nuclei of the sarcolemma are increased, and some of them form nucleus plates. Hematoxylin-eosin stain; 50x.

Case 3. S.M., female, age 46 in 1958, far advanced lepromatous leprosy. Onset was in 1929 and admission in 1957, before which the patient had received no treatment. On admission she had marked lepromatous infiltrations throughout the body and ulcers on the ears and forearms. Alopecia, ocular phthisis, saddle nose, and claw-hands were present. The patient died of nephrosis 10 months later.

Autopsy findings: Lepromatous lesions were found in spleen, liver, adrenal gland and larynx. There was marked edema of the lungs.

Histology.—In a specimen from the calf muscle the muscle tissue is so markedly destroyed that the original structure cannot be distinguished at a glance (Fig. 5). The muscle bundles are extensively



shrunken, exhibiting widely effacement of the transverse striations. Round or oval nuclei, abnormally rich in chromatin, are arranged in lines. The existence of the muscle structure can be inferred by the arrangement of these nuclei. Cellular infiltrations, composed largely of histiocytes and lymphocytes, are found between the muscle bundles, either arranged perivascularly or distributed diffusely. In these infiltrates there are scattered abundant acid-fast bacilli, and globi are also found. Leprous changes are found in both vessels and nerves.

CASE 4. Y.K., male, age 45 in 1958, far advanced lepromatous leprosy. Onset was in 1940, admission in 1950. At that time the patient showed severe lepromatous infiltrations throughout the body. There was anesthesia of the forearms and feet, and both hands were clawed. The treatment on admission was with Promizole. The patient eventually died of tuberculosis.

Autopsy findings: Lepromatous lesions were found in the larynx, spleen, liver and testicle. There was marked tuberculosis of the lungs, and also mitral insufficiency.

Histology.—In the specimen of the calf muscle examined the muscle bundles are in various stages of atrophy, showing a certain degree of shrinkage and fragmentation, and in some of them the cross striations are effaced. Nuclei of the sarcolemma are increased, and some of them form nucleus-plates (Fig. 6). In spite of the atrophy of the muscle, fatty degeneration is not conspicuous. In the interstitium there are inflammatory infiltrates composed of histiocytes, lymphocytes and fibroblasts. Numerous acid-fast bacilli with globi are found in these infiltrates. Bacilli are also found in the fascia, nerve tissue and endothelium of the blood vessels.

DISCUSSION

In view of the findings in the cases reported, it may be said that there exists a "myositis interstitialis leprosa." There is an interesting point of difference between the Case 1 and the other three cases. In the first case tumors were present in both calf muscles. Histologically, this tumor condition resembled the so-called "acute infiltration" in the skin, because Langhans giant cells and eosinophilic leucocytes were found in the inflammatory infiltration. In spite of the finding of large globi, there were very few acid-fast bacilli elsewhere. The lepromin reaction changed from negative to weakly positive in the same year that the tumors appeared. These findings indicate that the "acute infiltration" which is often seen in the skin may also occur in muscular tissue.

The other three cases are much alike in the clinical and histologic findings. The histologic changes were diffuse inflammatory infiltrations in the muscle, and various stages of atrophy of the muscle bundles. In these infiltrations the acid-fast bacilli were numerous.

The histologic condition in all four cases may be called "myositis interstitialis leprosa," because of the infiltrations, composed of histio-

cytes, lymphocytes, and fibroblasts, with many leprosy bacilli and globi, in the interstitium of the muscular tissue, resulting in the dispersion and degenerative atrophy of the muscle bundles.

By what course can the leprosy bacilli have reached the muscle? This is an interesting problem. In Case 1, probably, the bacilli reached the muscle through the blood vessels, leading to the inflammatory reaction. In the other three cases, however, the infiltrations and bacilli affected so much of the muscular tissue that the course of the invading bacilli cannot be traced.

CONCLUSION

It is concluded from these findings that, even though it may be very rare, a condition of "myositis interstitialis leprosa" really occurs in lepromatous leprosy.

ABSTRACT

This is a report of the finding of, besides atrophic changes, lepromatous infiltrates with acid-fast bacilli in specimens of muscle tissue from the calf of the leg in four cases.

The first case reported had improved markedly under treatment, and the lepromin reaction had converted from negative to weakly positive. There had, however, developed tumors in the calves of both legs, and biopsy of one of these lesions showed a condition which resembled the so-called "acute infiltration," because of the presence of Langhans giant cells and eosinophilic leucocytes in the inflammatory exudate. Apart from globus masses, however, bacilli were few.

In the autopsy specimens of the calf muscle of the other three cases reported there were found, besides muscle atrophy, inflammatory infiltrates consisting of histiocytes or macrophages and lymphocytes, with more or less abundant bacilli in various locations.

The author concludes that, even though it may be very rare, a condition of "myositis interstitialis leprosa" really occurs in lepromatous leprosy.

RESUMEN

Versa esta comunicación sobre los hallazgos, aparte de alteraciones atróficas, de infiltrados lepromatosos con bacilos ácidorresistentes en ejemplares de tejido muscular, procedentes de la pantorrilla en cuatro casos.

El primer caso descrito había mejorado notablemente bajo tratamiento, y la reacción a la lepromina había virado de negativa a débilmente positiva. Sin embargo, habían aparecido tumores en ambas pantorrillas y la biopsia de una de estas lesiones reveló un estado que se parecía a la llamada "infiltración aguda," debido a la presencia de células gigantes de Langhans y leucocitos eosinófilos en el exudado

inflamatorio. No obstante, aparte de esas masas globulares, había pocos bacilos.

En los ejemplares autópsicos del músculo de la pantorrilla de los otros tres casos, se encontraron, además de la atrofia muscular, infiltrados inflamatorios compuestos de histiocitos o macrófagos y linfocitos, con más o menos abundancia de bacilos en varios sitios.

Dedúcese que, aunque puede ser muy raro, existe realmente un estado de "miositis intersticial leprosa" en la lepra lepromatosa.