A Case of Leprous Nodular Interstitial Myositis

W. H. JOPLING
Consultant Leprologist, Hospital for Tropical Diseases, London, N.W. 1.

H. D. MEHTA
Senior Registrar, Hospital for Tropical Diseases, London, N.W. 1.

Although the presence of lepromatous granulomata in skeletal muscles is by now well known, these usually produce no symptoms or clinical signs. This is a report of a patient who developed firm and well defined nodules within the muscles of the calves and thighs while under treatment with dapsone. The histological findings are described.

Case Report

Mr B. L., an Indian from Surat District, came to England in 1964 at the age of 18, having spent the previous 2 years in Fiji. In 1968 he noticed swelling of his ankles and lower legs in the evenings and puffiness of the face in the mornings. About this time, he began to experience bouts of fever associated with epistaxis, redness of the eyes, and erythema of the skin of the nose and cheeks. Early in 1970 he noticed dryness of the skin of the trunk and limbs; in August of that year he was admitted to the Hospital for Tropical Diseases, London.

On examination, he was found to have oedema of the hands and feet, and ichthyosis of the trunk, arms and legs (Figs 1 and 2). There was a faint erythema of “butterfly” distribution in the central region of the face, but there were no macules, papules or nodules anywhere on the skin. The ear lobes were normal, and there was no diminution of hair of the scalp, eyebrows or body (Fig. 3). Slight enlargement of the right great auricular nerve and the right superficial peroneal nerve was noted, while examination of the eyes revealed superficial punctate keratitis and iridocyclitis. There was no impairment of sensation or of vibration sense; muscle power was normal, and the deep reflexes were active and equal.

Investigations

Histamine tests on various parts of the body gave a normal flare response. Skin smears from 8 random sites all contained granular acid-fast bacilli and the Bacterial Index was 3.6 on the Ridley scale (i.e., there were slightly fewer than 10 bacillary fragments in an average oil-immersion field). Biopsy of normal-looking skin showed lepromatous leprosy in regression, with considerable numbers of bacilli in the cutaneous nerves. Results of the Heaf and lepromin tests were negative. Blood count was as follows: Hb. 74% (10.8 g %); red blood cells 4,000,000 per mm$^3$; P.C.V. 35%; M.C.V. 88 mm$^3$; M.C.H.C. 31 g %; white blood
cells 6600 per mm$^3$. (P. 71%, L. 20%, M. 5%, E. 4%). The E.S.R. was 53 mm in 1 h. The erythrocytes showed moderate anisocytosis and hypochromia. Electrophoresis of serum proteins showed a slight decrease in albumin and a slight increase in alpha and gamma globulins (A-G ratio 3.3:3.9). The following tests were negative: antinuclear factor, Rose-Waaler, slide latex, Wassermann, smooth-muscle antibody, mitochondrial antibody, thyroid and gastric antibodies. Tests of liver and kidney function were normal, and X-ray of the chest was normal.

Progress

Treatment with dapsone, 10 mg daily, was begun on 21 August; a month later, he complained of difficulty in walking because of stiffness and discomfort in the legs and thighs. On palpation there were firm and well-defined masses of various sizes within the muscles of the calves and thighs, each mass tending to be elongated in the line of the muscles. The largest nodule measured 3 by 1.5 cm; it was not tender, and the skin over it was freely mobile and showed no signs of inflammation. Dorsiflexion of the ankles was limited by stiffness and pain in the calves. Body temperature was normal and there were no visible nodules on the skin, nor was there any sign of erythema nodosum leprosum (ENL). Blood examination at this time showed an increase in polymorphonuclear leucocytes to
85% of 8500 white cells per mm³, and a further increase in the E.S.R. to 80 mm, and of serum globulin to 4.7 mg%. An electromyogram revealed large areas in the calves devoid of muscle tissue; the needle electrode was very difficult to insert. In other areas, the motor units were broken up and reduced in number. An intramuscular nodule was removed from one calf and one thigh, and Dr D. S. Ridley reported as follows:

"There is extensive leprous interstitial myositis. Quite large foam cell foci are seen between the muscle bundles, and they appear to be followed by fibrosis, which is also extensive. The lepromatous foci are in a regressive phase, and the muscle itself is normal except where it is strangled by fibrosis. Non-solid bacilli (with club forms) are numerous in the lepra cells of one specimen but are not present in the muscle fibres (Figs 4, 5 and 6).

Treatment with dapsone was continued, and in addition he was given intramuscular injections of long-acting ACTH, 40 units twice a week. Physiotherapy was instituted to mobilize his ankle joints, and the ichthyosis was treated by daily bathing followed by inunction of Calmurid cream (10% urea in a stabilizing emulsified base). The intramuscular nodules slowly regressed over the next 2 months, and he was discharged on 27 November, 1970, feeling well able to walk normally, and with the ichthyosis much improved.

Fig. 3. Facial appearance on admission. Photo: U.C.H. Medical School Photographic Department.
Fig. 4. Low-power view of a muscle showing cellular infiltrate between muscle bundles and a surrounding zone of fibrosis. H & E stain

Fig. 5. The same as Fig. 4, under higher magnification, showing foam cells between muscle bundles. H & E stain

Fig. 6. Foam cells shown in Fig. 5 under highest magnification and stained to show acid-fast bacilli. The latter are in various stages of degeneration (fragmented, granular and club forms).
Discussion

Our first impression on studying the results of skin biopsy and smears was that the patient had been taking dapsone secretly, but he denied any previous history of leprosy or of self-medication. We considered the possibility of Lucio leprosy because of the absence of skin lesions, especially as patients with this type of diffuse lepromatous leprosy have been known to present as cases of ichthyosis (Frenken, 1963), but we excluded it on the strength of normal eyebrows, normal body hair, normal histamine test results, and the presence of superficial punctate keratitis.

Atrophy of skeletal muscles is well known in leprosy and is usually attributed to disuse or to peripheral nerve damage, but direct invasion of muscles by lepromatous granulomata, forming palpable intramuscular nodules, is quite rare. Ishihara (1959) reported the finding of nodules in the calf muscles of a patient under treatment, and on post mortem examination of 3 other patients he found lepromatous interstitial myositis. Convit et al. (1960) described nodules in the skeletal muscles of 4 patients undergoing lepra reaction, and histological examination of the nodules revealed lepromatous granulomata with fragmented leprosy bacilli. Lyer and Nath (1965) and Oderiz et al. (1965) described interstitial myositis in biopsy specimens of skeletal muscles taken during lepra reaction. Job et al. (1969) reported on the histological and electron-microscopic appearance of biopsy specimens from smooth and skeletal muscles in lepromatous leprosy. Pearson et al. (1970) made a study of skeletal muscles in leprosy and suggested that they were invaded by Mycobacterium lepra at an early stage in the disease. They found leprosy bacilli within muscle cells; in all other reports, including ours, cellular infiltrate and bacilli have been found between the muscle bundles and not within muscle cells.

Interstitial myositis is now a well recognized pathological feature in lepromatous leprosy, especially during lepra reaction, but this usually produces no symptoms or clinical signs. A finding of clinically palpable nodules within skeletal muscles is decidedly rare, and we expect that more cases will come to light if muscles are systematically palpated.

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References