THE DIAGNOSIS OF LEPROSY — COMMON ERRORS

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WOULD the correct diagnosis of leprosy have been easier if this disease had been described in textbooks on Neurology instead of Dermatology? The emphasis on the changes in the skin of the patient with leprosy may well be the cause of misdiagnosis in many cases.

Leprosy is a chronic infective disease and it is generally accepted to be caused by Mycobacterium Leprae discovered by Dr. A. Hansen in 1873 and published by him in 1874. M. Leprae is an acid-alcohol fast bacillus. So far no artificial medium has been found in which to culture the bacillus, but it can be kept alive and will multiply in the earholes and foot pads of the mouse and hamster. The foot pads are used solely as a culture medium enabling the testing of the effect of drugs on the bacillus.

It is important to know that M. Leprae has a special affinity for the Schwann cells of the sensory nerves in which they lie, protected by the basement membrane, (only seen by electron microscopy) and if conditions are suitable multiply. Only nerves of the Peripheral Nervous system are affected in leprosy. Therefore without symptoms showing nerve involvement the diagnosis of leprosy should not be made in the absence of positive skin smears. The great auricular nerve—the ulner and median and peroneal nerves are easy to palpate and possess predominantly cutaneous sensory fibres. In the early stages of the disease the bacillary invasion is directed to the sensory fibres while later on all types of fibres are affected due to granulomatous infiltration, scarring, ischaemic damage or even possibly transneuronal spread. Autonomic nerve fibres are commonly involved early in the disease, shown by the characteristic dryness and roughness of the skin and anhydrosis.

This article is mainly written for doctors who have never worked in countries where leprosy is prevalent and therefore have some difficulty in recognising the disease. To be Leprosy-conscious is as important and essential as to be Malaria-conscious and this applies to patients of all races. Leprosy can occur in ALL races and at any age. It should be born in mind that Europeans are not exempt from leprosy and unfortunately it has happened that Europeans have been treated for many years for an unidentified skin disease which later proved to be leprosy.

The main routes of infection are:

1. Droplet infection and inhalation from Lepromatous patients who have numerous AFB in the mucous membranes of nose, mouth and larynx.
2. Via the alimentary tract by infected food, of infected articles brought to lips and mouth.
3. Close skin contact with "open" cases. The presence on the skin of bites, cuts, or abrasions facilitates entry of the bacilus to the bloodstream through which it may be carried to its target, the schwann cell of cutaneous nerves.

There seems to be a genetic susceptibility for leprosy. Figures show that 70% of the population living in endemic areas will never manifest the disease. 24% however, pass through a so called "silent" phase where there is some pathology, but no clinical signs of leprosy. In the remaining 6%. suitable living conditions in the Schwann cells give rise to multiplication of the bacillus and according to the individual resistance of the patient this 6% will produce evidence of leprosy in any of the known forms from Tuberculoid to Lepromatous Leprosy.

The Cardinal signs and symptoms of leprosy:

1. Skin lesions with loss of sensation in the diseased patch. The lesion should be tested for hypoanaethsia or anaesthesia by touching the suspected lesion lightly with a feather or a wisp of cotton wool. Lesions on the face retained sensitivity to touch longer than those on the body also no examination is complete unless the lesion is tested for impairment of thermal sensitivity by using 2 test-tubes with cool and warm (not hot) water and also for analgesia using a pin and asking the patient to differentiate the sharp end from the blunt by gently pressing the diseased patch.

   Impairment of sweating, loss of hair, reduction in the degree of pigmentation are features of leprosy.
2. The presence of M. Leprae in the lesions. In patients suspected of leprosy routine skin smears should be taken from the earlobes and the lesions. One of the most frequent errors in diagnosing leprosy is the idea that skin smears should be positive and many patients are referred to Liteta because of negative skin smears.

   A patient can have extensive leprosy without having positive skin smears just as a patient can suffer from Tuberculosis without having a positive sputum.

Continued on page 127

Continued from page 123

3. Enlargement, hardness and tenderness of one or more of the main peripheral nerve-trunks in their superficial courses and of cutaneous nerves in proximity to a skin lesion and palpability of nerve-twigs not normally palpable.

The most common nerves to watch are:

a. The Supra-orbital nerve:
By palpating a skin lesion on the forehead this nerve may be felt enlarged.

b. The great auricular nerve: (Fig 4.)
This nerve is often enlarged when there are facial skin lesions.

c. The ophthalmic branch of the Trigeminal nerve causing corneal anaesthesia.

d. The Facial Nerve:
The Facial nerve may be involved at the stylomastoid foramen and causing a complete facial paralysis. Often only the zygomatic branch of this nerve is affected and may be found enlarged and tender over the zygoma producing lagophthalmos and paralytic ectropion.

e. Ulnar nerve:
If affected causing flexion of 4th and 5th fingers with loss of sensation of 5th and half of 4th finger.

f. Median nerve:
Sensation is lost over thumb and radial two thirds of palmar surface. There is hyperextension deformity of the index and middle fingers and wasting of small muscles. The patient has difficulty with many fine hand activities. A combination of ulnar and median nerve damage is common producing claw fingers. If in addition the radial nerve is affected, then a typical claw hand results.

g. Radial nerve:
This causes a "wrist-drop". Sensory changes are minimal. Radial nerve paralysis is less common than the ulnar and median varieties.

h. Peroneal nerve:
Peroneal nerve paralysis is very common in leprosy and results in a "drop foot".

i. Posterior tibialis nerve:
Damage to this nerve causes clawing of the toes and anaesthesia of the sole of the foot and therefore these patients are liable to get plantar ulcers.

Without going into details the following classification can be used in general practice:

Indeterminate:

This may be only one or two hypo-pigmented skin lesions anywhere on the body. It may show anaesthesia but very often it is impossible to make a definite diagnosis without a biopsy. One should never start treatment for leprosy until the diagnosis is made. If a definite diagnosis cannot be made the patient should be reviewed every 3—6 months.

Tuberculoid Leprosy.
Dimorphous Leprosy.
Lepromatous Leprosy.
Neuritic Leprosy.

This latter group is very important because of the absence of any skin lesions of the body but shows only the neural signs. One nerve may be affected (mononeuritic leprosy) or more than one nerve may be involved (poly-neuritic leprosy.)

The signs of neuritic leprosy are:-

1. Anaesthesia.
2. Nerve enlargement.
3. Muscular paralysis, paresis and wasting.
4. Trophic changes.

Pure neuritic or polyneuritic leprosy is one of the most mutilating forms of leprosy and early diagnosing is most important.

CASE 1

A male patient aged about 37 was referred to Liteta on 21st April 1967. In January, 1967 he saw a red skin lesion on this right cheek. He then felt itching of the lesion and eventually the right cheek started swelling. He knew it was leprosy! He was hospitalised and discharged without improvement. He was given anthisan tablets. He then went to a leprosarium and was referred to Liteta.

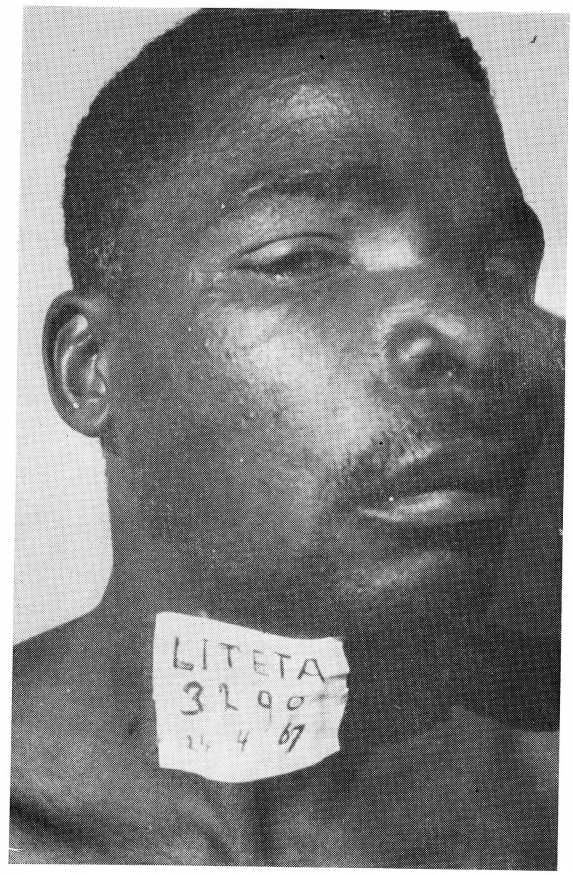

Fig. 1. Borderline tuberculoid with facial paresis and slight lagophthalmos.

On examination the patient had a very red slightly raised skin lesion covering forehead—nose—right cheek

Continued on page 129

Continued from page 127

and partly left cheek and upper lip. The edges of the lesion were flat and visible. The lesion was not anaesethetic but analgesia was present and impaired sensitivity for cool and warm.

Excessive lacrimation of the right eye with slight lagophthalmos—corneal anesthesia and slight right facial paresis were the most marked features. The zygomatic branch of the facial nerve was enlarged and very tender.

This skin lesion combined with corneal anaesthesia, facial paresis and lagophthalmos in addition to an enlarged tender nerve was not an allergic condition but a Borderline Tuberculoid leprosy. Skin smears were negative.

CASE II

A male patient aged about 25 was admitted to Liteta on 23rd July, 1967.

He noticed a few nodules on his left arm and a few weeks later some more nodules on his left hand. He attended a hospital but no specific treatment was given. His previous employer sent this patient to no less than 5 different doctors—3 times to a hospital and twice to a General Practitioner. A biopsy was taken

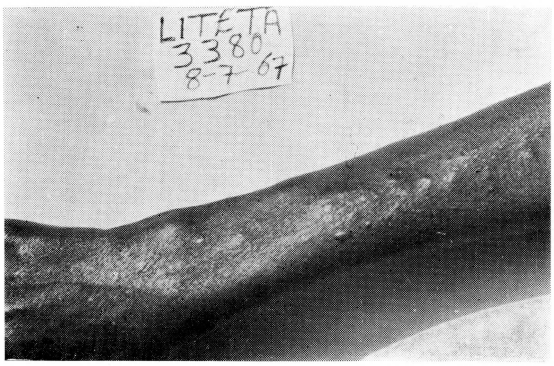

Fig. 2 Nodulation of skin with shiny patches of infiltration on the arm.

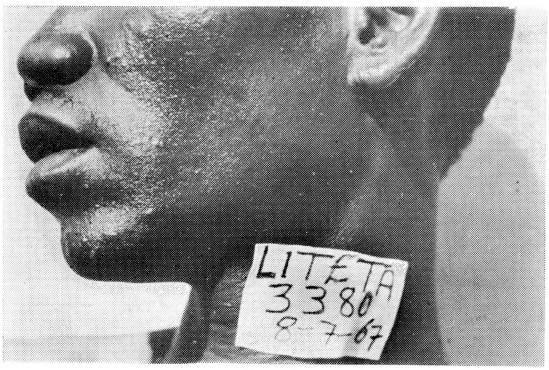

Fig. 3. Swelling of the ear lobe with slight nodulation f the skin. Thickening of the skin of cheek (orange peel appearance).

in 1966 but reported as "quite normal." Never were any smears taken.

In the meantime he served as a cook. He was told that he was allergic to fish. Two years after the first symptoms appeared he was admitted to Liteta.

On examination he had moderate nodulation of both ears, lateral maderosis thickening of entire skin of face, nodules on the chin. He had a general widespread, coppery skin rash on trunk, arms and legs.

Hands and feet were anaesthetic (glove and stocking type anaesthesia). Seven skin smears were positive with 10.4 % solids. He was diagnosed Lepromatous Leprosy.

CASE III

On 31st May, 1967 a male patient aged 46 was referred to Liteta with the following history.

His father had leprosy. In March, 1967 he noticed numbness of his right hand and right foot. He felt as if "something was moving under the skin of his right arm" and he felt prickling pains in his fingers and in his right foot. He knew it was leprosy and tested the skin of his hand with a piece of grass but did not feel the touch. Two months later his right hand as well as his right foot started swelling. He went to a clinic and was told that he suffered from rheumatism.

His condition worsened and as he was convinced that he had leprosy he went to a hospital and was referred via a leprosarium to Liteta. It is remarkable how many patients know when they are suffering from leprosy.

He had no skin lesions suggesting leprosy but widespread hyperpigmented areas caused by fire when rescuing a child from a burning hut.

His right hand was oedematous and anaesthetic with wasting of right hypothenar. His grip was poor and he had difficulty in holding objects. Right ulnar and median and superficial radial nerves were enlarged and tender. The right foot was oedematous and the lateral aspect anaesthetic. His right peroneal nerve was enlarged and tender. Routine smears were negative. Lepromin tests + +. Diagnosis : Polyneuritic leprosy.

CASE IV

On 27th June, 1966 a 49 year old white Canadian male patient was referred to Liteta with a letter from a colleague who admitted that he had never seen leprosy in a white skin but that the skin lesion and the anaesthesia of hands and feet in this patient were suggestive of leprosy and asked for confirmation of his diagnosis.

This patient arrived in Zambia in 1951 and working as a Missionary in the bush he was in close contact with the people. He was not conscious of having been in contact with a leprosy patient but he had been told that a cook once employed by him was supposed to have leprosy.

In 1959 the patient noticed stiffness in the 4th and 5th fingers of his left hand and swelling of the right index. At the same time he saw redness of the skin of the forehead—left cheek and legs. He also saw some

Continued on page 131

Continued from page 129

white skin lesions on the chest. From 15th October, 1959 until 25th October, 1960 he went to Canada on leave, and on his return while in London he again felt stiffness in his fingers of the left hand and burned his 4th and 5th fingers on a tea-pot without feeling pain. He returned via Rome and there his legs became swollen and painful.

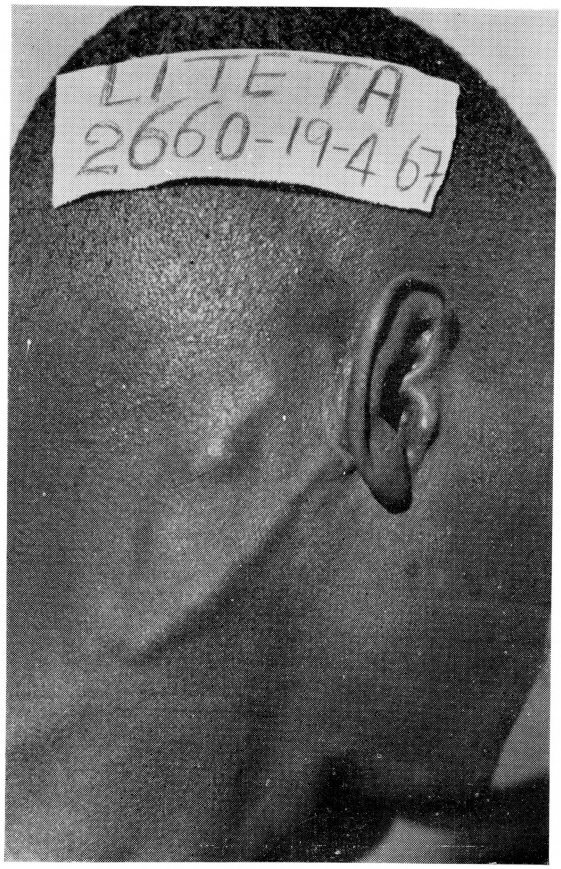

Fig. 4. Enlargement of the great auricular nerve.

On return to Zambia in November, 1960, he was examined by a doctor and treated with cortice-steroids for 3 weeks. His skin condition was thought to be an allergy to nylon. The patient was used to wearing nylon shirts and stockings. The cortice-steriods brought temporarily relief but from 1960—1966 he continuously felt "pins and needles" in arms and legs. It is amazing that this patient never saw another doctor but accepted the diagnosis of an alergic skin rash without questioning.

In 1966 he became apprehensive that something seriously was wrong. He could not feel the handlebars of his bicycle and he had to look to see that his fingers gripped the brakes. in other words his hands were anaesthetic.

On examination (Fig. 5)—the most striking features

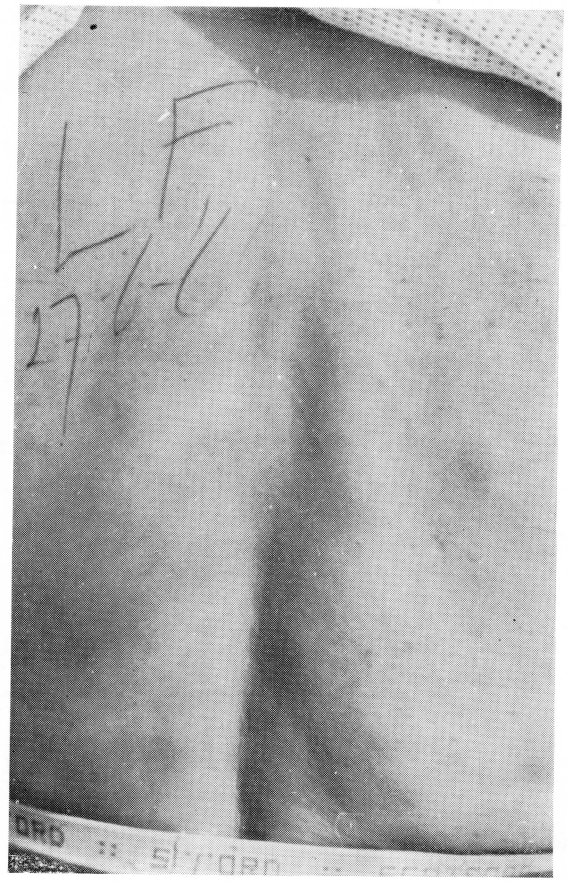

Fig. 5. Large and small widespread brownish slightly raised skin lesions without definite edges and scattered nodules in the skin. (A white male patient with lepromatous leprosy).

were the widespread dark-brownish skin lesions—some large—some small—slightly raised—without edges on chest—back—arms and legs. His earlobes were thickened and a few small reddish nodules present. The skin of forehead and cheeks was thickened and reddish. On the left forearm there were 3 red raised nodules (a biopsy of one of these nodules was taken).

NERVES: Left and Right auricular nerves slightly enlarged and tender.
Left and Right ulnar nerves enlarged and tender.
Left and Right median nerves enlarged and tender.
Left and Right peroneal nerves enlarged and tender.

Hands and feet: anaesthetic (glove and stocking type)—analgesia and impaired sensitivity for cool and warm present.

Seven smears were positive for M. Leprae with 15.1 % solids. The lepromin test was negative.

Continued on page 133

Continued from page 131

Diagnosis: Lepromatous leprosy of many years duration.

Report on the biopsy by:

Dr. D. J. Harman. M.B., Ch.B.,
The Leprosy Study Centre,
57a Wimpole Street,
London, W.I.

"This is a biopsy for an infiltrated and raised lesion in a case of active and advanced lepromatous leprosy".

There is little doubt that the signs of stiffness in 4th and 5th fingers of the left hand the patient experienced in 1959 (ulnar neuritis) combined with the red and white skin lesion were due to leprosy.

Thanks are due to Dr. D. J. Harman for his biopsy report and my apologies for not publishing his excellent report in full.

SUMMARY

Nerve involvement is one of the main symptoms of leprosy and it is essential that the skin lesions suspected of being leprosy should be tested for sensory changes.

The nerves of the peripheral nervous system should be examined for enlargement and (or) tenderness and their distribution tested for sensory and motor changes.

Routine smears should be taken in suspected patients. One should never treat an unidentified skin disease without having ruled out leprosy either clinically or by biopsy.

1965. It stresses the importance of visualising an appreciable length of proximal bowel above the site of obstructed herniae.

Case Report. A young man of about 30 years was admitted on 25th April, 1967 with a history of painful right inguinal swelling, pain in the lower part of the abdomen and vomiting. The symptoms were of about 4/½ hours duration. On examination he had a tender, non-reducible, indirect Inguinal Hernia with no cough impluse. He had mild tenderness on deep pressure on the lower part of his abdomen. Operation was performed an hour after admission. The sac contained sero-sanguinuous fluid, caecum and appendix and 8—9 inches of terminal ileum. The contents were viable. Ileum proximal to the obstruction was brought down and showed about 3 feet of ischaemic, angry looking ileum. It recovered its normal colour on compression with warm moist packs. The contents were reduced. The terminal portion of the appendix appeared gangrencus due to kinking of meso-appendix and appendicectomy was performed. The operation was then completed. The patient was discharged 7 days later following an uneventful recovery.

Discussion: In the common type of obstructed hernia, the strangulated bowel lies in the sac of the hernia. In the type described above, the bowel strangulated is the ileum lying proximal to the loop of bowel lying in the sac and is entirely intra-abdominal. In Philip's Series[1] this type of strangulation was found in 14.2% of obstructed herniae.