Orthopaedic complications of leprosy

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Worldwide leprosy remains a common problem with 750,000 new cases being diagnosed each year. About 30% of patients have established nerve damage at the time of diagnosis and orthopaedic problems may therefore develop even after successful antibiotic treatment. Orthopaedic complications in leprosy include altered bony architecture and stress leading to repeated local trauma which, in combination with nerve damage, causes plantar ulcers.

Leprosy still causes considerable long-term morbidity in both the developing and developed world. We discuss the orthopaedic complications and management of leprosy.

Leprosy is an infectious disease caused by Mycobacterium leprae. It is characterised by skin lesions and peripheral nerve damage. It may cause physical disability and disfigurement. The global health costs cannot be easily calculated.

It was Hansen in 1873 who discovered M. leprae, the first bacterium to be identified as causing disease in humans.

There are over 4 million individuals who have or are disabled by leprosy worldwide and about 120 cases in the United Kingdom under regular observation at The Hospital of Tropical Diseases, with from 15 to 20 new cases diagnosed per year. With increasing migration, it has become more important to understand this disease since patients may present with the disease or its complications outside endemic areas. The diagnosis may be delayed in non-endemic areas like the United Kingdom and this may lead to permanent nerve damage and disability.

M. leprae is an acid-fast rod and grows best in cooler tissues (the skin, peripheral nerves, anterior chamber of the eye, upper respiratory tract, and testes).

Patients with untreated lepromatous leprosy discharge bacilli from the nose. The principal portal of entry into the human body is the upper respiratory tract. M. leprae cannot traverse through intact skin in either direction, and the infection is not spread by touching. The incubation period is two to five years in tuberculoid disease and eight to 12 years in lepromatous disease. In contrast to tuberculosis, co-infection with HIV has no strong effect on the development of leprosy.

Pathology

M. leprae has a predilection for Schwann cells and skin macrophages and is taken up by these cells early in infection (Fig. 1).

In many cases (indeterminate leprosy), early lesions heal spontaneously with eradication of bacilli. If the bacilli persist and multiply in the...
skin and/or nerves, established leprosy develops. In tuberculoid leprosy there is involvement of skin and nerves. Lymphocytes breach the perineurium, and destruction of the Schwann cells and axons may be evident, resulting in fibrosis of the epineurium, replacement of the endoneurium with epithelial granulomas, and occasionally caseous necrosis. Acid-fast bacilli are few or absent. Such invasion and destruction of nerves in the dermis by T lymphocytes is pathognomonic for leprosy. Sometimes a caseous abscess may form inside the perineural sheath causing paralysis of the nerve.

Lepromatous disease manifests with generalised involvement of skin, nerves and mucous membranes. The damage and hypertrophy of nerves resulting from bacillary invasion tends to be symmetrical and is more insidious and extensive than in tuberculoid disease. The granulomatous lesions contain macrophages and abundant bacilli. In skin lesions, the small dermal sensory and autonomic nerve fibres are damaged. This can lead to glove and stocking paraesthesiae and loss of sweating. Damage to peripheral nerves leads to sensory loss and paralysis. Nerve damage occurs across the leprosy spectrum.

Acute nerve pain with associated loss of function may occur as part of a leprosy reaction, an acute immunological reaction, which presents with inflamed skin lesions and neuritis.

Immunology
The patient’s immune response to M. leprae determines the features of the disease: the two poles are tuberculoid (paucibacillary) and lepromatous (multibacillary) leprosy. At the tuberculoid pole, well-expressed cell-mediated immunity and delayed hypersensitivity control bacillary multiplication, which limits the disease to a few well-defined skin lesions or nerve trunks. Few mycobacteria are found in the lesions. In the lepromatous form, there is cellular anergy towards M. leprae, resulting in abundant bacillary multiplication. There is uncontrolled proliferation of bacilli with many lesions and extensive infiltration of the skin and nerves.

Between these two poles lie different groups. These forms are characterised by a progressive reduction in cell-mediated immunity from borderline tuberculoid to borderline lepromatous leprosy in cellular responses, associated with an increasing bacillary load and increasing number of skin and nerve lesions.

The intra-osseous lesions of leprosy are characterised by granulomatous tissue reaction that lead to trabecular destruction. The lesions are evident in the epiphysis and metaphysis of tubular bones, and direct involvement of the medullary canal can also occur.

Classification
Classification of patients according to the Ridley and Jopling scale is clinically useful with a spectrum from tuberculoid to lepromatous leprosy.

There is also a simpler classification determined by the number of skin patches: single skin lesion (one patch), paucibacillary (two to five patches) and multibacillary (more than five patches). Patients with multibacillary leprosy are more likely to have nerve damage.

Clinical manifestations
The signs of leprosy are skin lesions, which are typically anaesthetic at the tuberculoid end of the spectrum, thickened peripheral nerves and eye involvement, which may lead to blindness. In lepromatous leprosy, the skin lesions are nodules, papules, or plaques with predilection for the face, wrists, elbows, buttoks, and knees. Involvement of major nerve trunks is common and there is also glove-and-stocking anaesthesia in the extremities.

Patients with borderline disease have multiple skin and nerve lesions. The pathological effects of leprosy on the skeleton are primarily a consequence of neuropathy leading to denervation, direct bone changes, secondary infection and the sequelae of trophic ulcers (Fig. 2).

The following superficial peripheral nerve trunks may be palpably hypertrophied: facial, great auricular (neck), ulnar (elbow), median (wrist), radial cutaneous (wrist), lateral popliteal (neck of fibula), and posterior tibial (medial mal-
leolus). The most commonly-affected is the posterior tibial nerve followed by the ulnar nerve at the elbow, whose involvement results in clawing of the fourth and the fifth fingers, and loss of dorsal interosseous musculature and loss of feeling in the ulnar nerve distribution (Fig. 3). Median nerve involvement impairs thumb opposition and grasp, while radial nerve dysfunction, though rare in leprosy, results in wristdrop.

Peroneal nerve palsies may result from leprosy itself or from one of its reactional states. It leads to a partial or complete foot drop, which causes an uneven distribution of weight on the plantar surface and hence a predilection to ulceration (Table I).

Repeated trauma in the absence of protective feeling may cause trophic ulcers which may become secondarily infected. In severe cases osteolysis of the terminal phalanges can occur.

Plantar ulceration, particularly at the metatarsal heads, is probably the most frequent complication of leprous neuropathy, and secondary infection leads to cellulitis and osteomyelitis.

In the face, hands and feet, there may be direct osseous involvement due to extension of infection from overlying dermal or mucosal areas. The periostium is infected initially (leprosy periostitis) and subsequently the cortex and medulla are infected (leprosy osteitis and osteomyelitis). At the fingers and the toes it may appear as dactylitis.

Inflammatory, symmetrical peripheral polyarthritis of insidious onset with a pattern of exacerbation and remission, is occasionally seen. The wrist, metacarpals and proximal interphalangeal joints of the hands, the knees and the metatarsophalangeal joints are involved with morning stiffness lasting up to one hour.

Patients with leprosy can present with skin lesions, peripheral neuropathy, ulceration, soft-tissue infections, and osteomyelitis, deformities of the hands and feet, and joint involvement. They can present to a dermatologist, neurologist, rheumatologist or an orthopaedic surgeon.

**Diagnosis.** The principal criteria for the diagnosis are a hypopigmented patch of skin with sensory impairment, thickening of the peripheral nerves, the identification of *M. leprae* in slit-skin-smears, which are graded on the Ridley and Jopling bacteriological index as 1+ to 6+, and characteristic histopathological changes in a biopsy specimen of a skin lesion or a peripheral nerve.

**Bone changes in leprosy**

These can be divided into two groups, specific and secondary changes. Specific lesions are due to the direct involvement of bone by the organism, whereas secondary lesions are the result of trauma and infection (Fig. 4) imposed upon denervated tissues. Secondary bony changes are commonly observed and are therefore discussed first.

**Secondary bony change.** These are common in leprosy. Localised osteoporotic changes result from immobilisation, most frequently because of disuse associated with fixed contractures of the fingers.

Motor denervation is sometimes associated with absorption of the cancellous bone and the development of a concentric type of bone atrophy. It affects the length, the width or both. The most common changes in leprosy however are those due to combined absorption of length and width of bone. The result is a tapered appearance at the end of the bone, termed ‘licked candy stick’.

Changes due to distal absorption affect the ends of insensitive fingers and toes. When this process is complicated by infection, it may be followed by progressive absorption with loss of the digits and the development of the so-called ‘mitten hand’.

Absorption secondary to trauma and infection has three common sites of predilection in the insensitive foot. The first is the distal type affecting the tips of the toes. In the second type the metatarsophalangeal joint is at risk of damage and the third involves the tarsus.

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**Table I. Peripheral nerve involvement in leprosy**

<table>
<thead>
<tr>
<th>Nerve</th>
<th>Motor loss</th>
<th>Sensory loss</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ulnar nerve</td>
<td>Clawing of ring and little fingers, loss of intrinsic muscle function</td>
<td>Anaesthesia over the medial two fingers of the hand</td>
</tr>
<tr>
<td>Median nerve</td>
<td>Loss of thumb opposition and grasp</td>
<td>Anaesthesia over the thumb</td>
</tr>
<tr>
<td>Radial nerve (rare)</td>
<td>Wrist drop</td>
<td>Anaesthesia of first web space</td>
</tr>
<tr>
<td>Common peroneal nerve</td>
<td>Foot drop</td>
<td>Anaesthesia over the lateral malleolus</td>
</tr>
<tr>
<td>Posterior tibial</td>
<td>Clawing of toes</td>
<td>Anaesthesia of the sole of the foot</td>
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![Fig. 3](photograph-showing-clawing-of-fingers-and-a-trophic-ulcer.png)
The common paralytic deformities of the foot, which are potentially damaging to the metatarsophalangeal joints, are clawed toes and drop foot. Static deformities such as hallux valgus, metatarsus primus varus and pes planus are also associated with plantar ulceration, which may lead to bone absorption. Tarsal disintegration is not infrequent and may involve one or more tarsal bones. The most common tarsal disintegration affects the medial arch. The lateral arch is less commonly involved and occurs as a late complication of a rigid deformed foot.

Specific bony changes. Specific bony lesions in leprosy are rare with an incidence of between 3% and 5% among hospitalised patients, and are mainly confined to the small bones of the face, hands and feet. These lesions are characterised by granulomatous tissue reactions, which are destructive and manifest radiographically as focal areas of increased rarefaction. The margins are thin, but may be sclerotic. Obliteration of the cavity may result from its collapse with flattening of the articular surface.

In the hands and feet, the disease mainly involves the proximal and/or the middle phalanges. It may present as thinning of the endosteum with corresponding widening of the medullary canal localised to the area of the metaphysis. Fusiform swelling of the soft tissues overlying the corresponding part of the affected digit is more common. This is occasionally associated with enlarged nutrient foramina. Leprous osteitis in the hands commonly involves the distal ends of the proximal and middle phalanges, whereas it usually affects the metatarsal heads in the feet. Because of weight-bearing forces, there may be comminuted pathological fractures of the metatarsal heads.

Should the disease progress and the trabeculae be destroyed, the radiographs reveal a ‘honeycomb and cystic’ appearance. With healing, radiographic changes become sharply defined as cysts with sclerotic margins. The articular surface can also be involved and the intrinsic forces in the hand may result in fracture, subluxations and rigid clawing of the fingers.

Prevention and treatment
It is vital to prevent the development of nerve palsies and ulcers. Primary prevention is by early detection and the use of antileprosy drugs. Secondary prevention is by health education and self-awareness of the patient; hygiene of the anaesthetic foot with attention to cracks and fissures, early detection of tenderness and adequate rest; awareness of potential injury mainly during work and cooking; appropriate foot wear.

Antibacterial. Multidrug therapy is the mainstay of treatment. The current multidrug therapy recommended by the World Health Organisation in adults with multibacillary disease is rifampicin, 600 mg orally once a month, dapsone 100 mg orally daily, and clofazimine, 300 mg orally once monthly and additionally 50 mg daily. The WHO recommends that it be continued for 12 months but patients with initial high bacterial loads may need longer treatment. For paucibacillary disease (bacteriological index, 2+) the regimen is rifampicin, 600 mg orally once monthly, and dapsone, 100 mg orally daily for six months.

Steroid treatment for nerve damage. Any patient who develops peripheral nerve damage during the last six months of treatment should receive a four- to six-month course of oral steroids.

Physiotherapy. There is usually stiffness of the fingers. Physiotherapy with active exercises, massage, passive stretching and wax baths and splinting should be undertaken before giving consideration to tendon transfer procedures.

Monitoring of nerve damage. Impairment of nerve function can occur before diagnosis and during or after multidrug therapy. Patients with multibacillary leprosy and pre-existing nerve damage are at the highest risk of impairment of nerve function during and after treatment. These patients should be under surveillance for two years from diagnosis. Regular clinical evaluation should include nerve palpation, motor testing of the small muscles of the hand and recording of feeling in the hands and feet using the Semmes-Weinstein nylon monofilaments. If new nerve involvement is detected steroid treatment should be started.

Surgical treatment. Surgery has an important role in the management of motor imbalance, chronic ulceration, chronic infection and the correction of soft-tissue and bony deformities. Decompression of nerves, such as the ulnar or common popliteal in the early stages may prevent the development of paralysis. Steroids may also be given to reduce inflammation.

Fig. 4
Radiograph showing osteomyelitis of the calcaneum secondary to plantar ulceration.
Permanent claw-hand deformities can be treated by tendon transfers. These procedures should not, however, be undertaken until six months after the start of antimicrobial therapy and the conclusion of episodes of acute neuritis.

The treatment of plantar ulceration includes debridement of devitalised tissue, non-weight-bearing by means of a total-contact cast or bed rest and the vigorous treatment of secondary infection, which is most commonly caused by *Staphylococcus aureus* (Figs 5 and 6). The ulcers should be examined for infection and probed to detect any bony involvement. If there is slough or necrotic material, it should be thoroughly debrided. Bony involvement may require multiple debridements and sometimes excision. Leprous ulcers heal well if weight-bearing is prevented as they have good vascular supply unlike those in diabetic neuropathy. Once healing takes place, walking must be limited and only increased slowly. Extra-depth shoes or custom-made shoes with moulded inserts are required to prevent recurrence. Areas of bony prominence which may lead to ulceration should be removed surgically. Amputation may be indicated if there is gross destruction of bones and joints.

Foot drop can be initially treated by simple splints. Subsequent tendon transfers may be required. A neuropathic foot may require more extensive surgery in the form of corrective osteotomy, arthrodesis or amputation.

**Conclusion**

Leprosy is rare in the United Kingdom. Early diagnosis and treatment is required in order to prevent nerve damage. A history of travel from an endemic region, a skin rash and/or neurological symptoms, and possibly a history of joint symptoms should raise the suspicion of the diagnosis of leprosy.

**References**

1. World Health Organisation. [http://www.who.int/lep](http://www.who.int/lep) (accessed 15/01/05).