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REVIEWS

Nail Involvement in Leprosy[☆]

I. Belinchón Romero,^{a,*} J.M. Ramos Rincón,^{b,c} F. Reyes Rabell^c

^a Sección de Dermatología, Hospital General Universitario de Alicante, Alicante, Spain

^b Unidad de Enfermedades Infecciosas, Hospital General Universitario de Elche, Alicante, Spain

^c Gambo General Hospital, Shashemane, Etiopía, África

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Abstract Leprosy, a disease caused by *Mycobacterium leprae*, primarily affects the skin and nerves, but the nails are also involved in as many as 3 out of 4 patients. The factors that trigger nail changes in leprosy are numerous and include repeated trauma, neuropathy, vascular impairment, infections, lepra reactions, and the drugs used to manage the disease. The changes most often reported include subungual hematomas, onycholysis, onychauxis, onychogryphosis, pterygium unguis, and onychoheterotopia, most of which can be attributed to nerve damage and trauma. Furthermore, the acro-osteolysis that occurs in the advanced stages of the disease may present with brachyonychia, racquet nails, or even anonychia. Infections of the nail bed leading to paronychia and onychomycosis should also be taken into account in leprosy. Other typical changes include longitudinal striae, pitting, macrolunula, Terry nails, leukonychia, hapalonychia, and Beau lines. In this review, we describe the principal nail changes associated with leprosy. These changes, which are highly varied and diverse in origin, are in fact a reflection of the significant morbidity caused by *M leprae* infection.

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Las uñas en la lepra

Resumen La lepra es una enfermedad causada por *Mycobacterium leprae*. Afecta principalmente a los nervios y a la piel, y hasta en tres de cada cuatro pacientes también a las uñas. Las causas desencadenantes de las lesiones ungueales en la lepra son múltiples, y de ellas destacan los traumatismos repetidos, la neuropatía, la insuficiencia vascular, las infecciones, las leprorreacciones o los fármacos utilizados en el tratamiento. Entre los cambios más destacados se encuentran los hematomas subungueales, la onicolisis, la onicauxis, la onicogriphosis, el *pterygium unguis* dorsal o la onicoheterotopia, y en su mayoría pueden atribuirse al daño nervioso y a los traumatismos. Por otro lado, la acroosteolisis que se produce en estadios avanzados puede cursar con braquioniquia, uñas en raqueta o incluso llegar a la anoniquia. Las infecciones de las uñas, con la aparición de paroniquia y onicomycosis, constituyen otro de los capítulos a tener en cuenta en la lepra. Además hay otras alteraciones características como las estrías longitudinales, los *pits*, la macrolunula, las uñas de Terry, la leuconiquia, la hapaloniqualia o las líneas

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* Corresponding author.

E-mail address: belinchon_isa@gva.es (I. Belinchón Romero).

de Beau. A lo largo de esta revisión se describen los principales cambios que se producen en las uñas por esta enfermedad, que son muy variados y de origen muy diverso, y de hecho son el reflejo de la amplia morbilidad que causa la infección por *M. leprae*.

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Introduction

Leprosy, a disease known since antiquity and about which many notions have formed over the course of history, is caused by *Mycobacterium leprae*. Although not fatal, this infection significantly limits the physical, psychological, and social well-being of patients¹ and while its prevalence is now extremely low in many parts of the world it remains a major public health problem in certain areas.² We have known how to treat this disease since the end of the 1970s, yet full understanding did not come until the introduction of shorter duration multidrug therapy at established doses in 1982.³ With wide application of multidrug therapy and early diagnosis of leprosy, the prevalence of the disease has decreased worldwide and functional morbidity and mortality have diminished.²⁻⁶

M. leprae infection mainly affects nerves and skin, but additional damage arises as other organs and structures (eyes, mouth, joints and more) become involved.⁴ Functional limitations and partial or full disability can develop, with dire consequences on the patient's quality of life.

The number of new cases of leprosy around the world fell from 620 000 in 2002 to 250 000 in 2009, according to the World Health Organization. Most new cases occur in India, Indonesia, Brazil, Nepal, Bangladesh, and certain African countries, such as Nigeria, Angola, and Ethiopia.² In recent decades, fewer cases have emerged in the local population in Spain, but we are now diagnosing leprosy more often in immigrants as a result of global migration from regions where the disease is more prevalent.^{7,8} Furthermore, a fair number of young Spanish doctors are now traveling to work with organizations providing medical care in low-income countries, where they see patients with leprosy.^{9,10} This disease has been discussed in a few review articles in the recent Spanish medical literature,¹¹⁻¹⁴ but as far as we can tell the effects of leprosy on the nails have not been addressed. Therefore, based on our experience treating patients with leprosy in Ethiopia,^{15,16} we aim to review the effects of this disease on the nails for the benefit of interested clinicians.

The Nails: Development and Anatomy

Nails are cutaneous adnexal structures that protect the tips of fingers and add a degree of precision that improves our ability to pick up objects and perform other delicate maneuvers.¹⁷⁻¹⁹ These structures are derived from the primitive epidermis, as are hair and the stratum corneum.¹⁷ The nail unit consists of the plate, matrix, bed, and proximal and lateral folds. The outer layer is the plate, which covers the matrix and the bed.¹⁹ The nail matrix is divided

into the dorsal, middle, and ventral portions. The dorsal portion of the matrix forms the outer layers the nail plate, the middle of the matrix gives rise to the deeper layers of the plate, and the ventral matrix contributes to the formation of the nail bed. The proximal fold is the continuation of the skin of the finger, which folds back upon itself at the dorsal part of the matrix. The hyponychium, located just under the free margin of the nail plate, represents the transition from the epithelium of the nail bed.²⁰

Causes of Nail Damage in Leprosy

Nail changes may derive from diseases specific to the unit itself or may be part of the general clinical picture of a skin disease. In addition, nail changes may occur during the course of a variety of systemic diseases, and most such changes are nonspecific.¹⁷⁻¹⁹

Examination of the nails is usually of great help in the clinical diagnosis of certain systemic diseases, such as dermatomyositis. Nail changes that occur in both the early and late (mutilating) stages of leprosy can affect the plate, the matrix, the bed, and the folds. Observation of these changes can aid diagnosis and help in the evaluation of the repercussions of leprosy on the patient's physical, psychological, and social well-being.

As many as 3 out of 4 patients with leprosy have nail involvement. The associated factors are many and include repeated trauma, neuropathy, vascular impairment, infections, or adverse effects of drugs used in treatment.^{17,21-23} The causes of leprosy nail damage are summarized in Table 1. The main factor is neuropathy, which in addition exacerbates the negative effects of all the others.²¹⁻²³ Therefore, peripheral neuropathy in leprosy might be assumed to lead to changes of the type seen in diabetic neuropathy, but this is not the case; nail problems in leprosy patients have been seen to be more common and more severe than in patients with diabetes.²²

Nerve damage leads to loss of sensation and deformities in fingers and toes; autonomic dysfunction also develops, leading to anhidrosis and dry skin and fissures that affect the hands and feet in particular. With the numbness at the distal ends of deformed fingers and toes, any slight mechanical trauma or exposure to heat will predispose the patient to further injury, particularly if the insult is often repeated. Infections will then develop, leading to lysis of the phalangeal bone, thinning and finally loss of the tips of fingers and toes along with their nails.^{5,6,24} Nail changes support the hypothesis of Baran and Juhlin²⁵ that nail development is tied to that of the underlying bone; thus, anonychia and hyponychia occur when the

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