

A Forgotten Disease “Leprae” and Limb Lesions

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Abstract

Leprae, which is a forgotten infectious disease, presents with primary findings of skin lesions and peripheral nerve lesions. Neuropathic ulcers are frequently found and may cause deformities, particularly in the extremities. It should not be forgotten that malignant tumors seen on the floor of chronic ulcers can also be seen on the floor of neuropathic ulcers depending on lepra. In our study, a 61-year-old male patient with an ulcerated and unhealed lesion in the volar surface of the second finger of his right hand that existed for approximately 6 months is presented. Although lepra is not common these days, early defect repairs will prevent osteomyelitis and sequelae of amputation, depending on the malignancy in these patients.

Keywords: Leprosy, auto amputation, skin defects

INTRODUCTION

Leprosy is a rare infectious disease caused by *Mycobacterium leprae*. It shows various clinical and histopathological symptoms. Although it primarily affects the skin and peripheral nerves, it can involve almost any organ. Neuropathological ulcers are commonly encountered in leprosy.¹ Due to their chronic progress, they may undergo malignant transformations, as is also the case for other ulcerated lesions.² In our study, we present a case of a patient who had been diagnosed with lepromatous leprosy (LL) and who was regularly monitored; the patient presented to our clinic with tissue defects in two digits of the right hand and widespread digital deformities.

CASE PRESENTATION

A 61-year-old male presented to our clinic with a 6-month history of non-healing ulcers with discharge in two digits of his right hand. His medical history revealed that he had been diagnosed with LL 40 years ago, after which he had undergone treatment for multibacillary leprosy and thereafter monitored periodically. Physical examination revealed sensory loss on both hands, congestion in the right eye, lagophthalmos, bilateral loss of eyebrows and eyelashes (madarosis), and the following deformities on the third, fourth, and fifth digits: 3×2 cm ulcerated lesions on the volar surfaces of two digits on the right hand and auto-amputation of the third digit (Figure 1). The right and left gluteal regions and left knee contained an atrophic scar of approximately 4×3 cm, which was pale at the center and hyperpigmented at the periphery. Upon consultation with an infectious disease specialist, the patient was prescribed a multiple antibiotic treatment. The scar culture revealed no reproduction, and no acid-resistant bacteria were found after specimens obtained from the lateral part of the eyelids and the ulcerated lesion were stained using the Ehrlich–Ziehl–Neelsen (EZN) method to confirm the diagnosis of leprosy. The results of two incisional biopsies taken from the margin of the ulcer were consistent with chronic inflammatory granulation. Magnetic resonance imaging did not yield any findings related to osteomyelitis. Surgical treatment included debridement of defect margins under axillary block anesthesia and suturing of the full thickness skin graft obtained from the flexor surface of the wrist to the defect area (Figure 2).

DISCUSSION

Leprosy is a chronic granulomatous infectious disease caused by *M. leprae*; it is particularly encountered in socioeconomically underdeveloped regions.¹ Because it affects the sensory and motor fibers of the peripheral nerves, patients are susceptible to traumatic injuries, and this in turn facilitates ulcer development. Chronic peripheral neuropathy can also lead to neuropathic ulcers. Neoplastic transformation is rarely encountered in ulcers; however, epidemiological studies demonstrate that the risk is higher than expected.^{2,3}

In leprosy, lepromas can readily settle in extremities, more specifically in the knees, elbows, and extensor surfaces of wrists and ankles. Protruding from the skin, these copper-colored hemispherical nodular lesions can also assemble to form infiltrative plaques. When they form large masses, they may exert pressure on the circulatory system, causing elephantiasis in distal regions.⁴

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Atrophic scars may emerge in the knees and elbows as a result of healing lepromas in these regions or due to regeneration of deep ulcerations formed by the bursting of bullae previously developed under the influence of various traumatic and thermic factors. Localized atrophic scars on the knee and elbow are significant in that they are indications of permanent and lifelong leprosy. In particular, in tuberculoid leprosy, paralysis caused by affected peripheral nerves may interfere with hand at foot function, resulting in various deformities. In the hands, disappearance of opposition, hyperextension at the metacarpophalangeal joints, and hyperflexion at the interphalangeal joints cause the digits to flex toward the palmar surface. Grasping ability is also lost and claw hand deformity may develop. The effects of the disease on hand bones lead to resorption mutilation and loss of digits. In this resorption mutilation process, the unaffected fingernails that attach to the tip of the phalanx lying underneath the phalanx that has been lost form a deformity, which is an invariable characteristic of this condition (Figure 3).⁴

Research demonstrates that among leprosy patients, malignancy occurs most frequently in the age group of 60–69 years, in borderline tuberculoid (BT) or borderline lepromatous (BL) leprosy diagnosed patients who have been monitored for the last 30–35 years. Severe nerve damage occurring in BT and BL may enhance ulcer formation. The most common finding in tumors is cauliflower growth. These are usually asymptomatic but may be accompanied by mild pain and easily triggered bleeding. Direct graphy findings may indicate osteomyelitis, non-specific reaction of the periosteum, or osteolysis. Local enlargement of lymph nodes may also occur indicating infection or metastasis.⁵ Our patient had been diagnosed with LL and was monitored for 40 years. The lesion was on the palmar surface of the digits, which is a common location for neuropathic ulcers. In one study, biopsies taken from 416 chronic ulcers that had developed on the extremities of leprosy patients revealed 102 (24.5%) squamous cell carcinomas, of which 83.4% were localized in the lower extremity, mostly on the plantar region.⁶



Figure 1. Preoperative appearance of the ulcerated lesion on the palmar surface of the second digit of the right hand



Figure 2. Early postoperative appearance after repairing the tissue with a graft



Figure 3. Auto-amputation and claw hand deformity in the third digit of the right hand

In a study of leprosy with plantar ulcers, patients were classified in two groups based on the existence of squamous cell carcinoma. The comparison of the groups yielded no correlation between squamous cell carcinoma development and race, profession, type and duration of leprosy treatment, locational origin of malignancy, existence of bone involvement, and type of ulcer treatment. However, interestingly, the group with malignancy presented a significantly decreased ulcer duration.⁷

An early diagnosis and treatment of chronic ulcers in leprosy is important to prevent deaths from violent occurrences of the disease. It is also important to provide proper care to the ulcer and avoid trauma. Biopsies should be taken from persistent ulcers; it is suggested that multiple biopsies be taken from various areas of the suspicious lesion to confirm diagnosis.²

In treatment of squamous cell carcinoma occurring in conjunction with chronic ulcers, planning should be done in accordance with each patient. The clinical appearance of the tumor and existence of osteomyelitis affect decisions related to treatment. In case of early diagnosis, simple resection is sufficient, while delayed diagnosis may require amputation and result in loss of the extremity. Surgical treatment involves excision and grafting in well-differentiated, small, and non-invasive tumors and amputation in moderately and poorly differentiated tumors, as well as large, invasive, and well-differentiated tumors.^{2,8}

CONCLUSION

In neuropathic ulcers observed in leprosy, it is important to bear in mind the risk of malignant transformation. We propose that periodic clinical observations, and biopsy applications when necessary, are important in reducing mortality and morbidity and that early treatment of ulcerated lesions should decrease the number of amputations necessitated by osteomyelitis, and thus the loss of extremities.

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