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Case Report

A Rare form of Hansen's Disease Presenting as Hypertrophic Scar

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Abstract

Hansen's disease or leprosy is a chronic disease that mostly affects the skin and peripheral nerves which results in neuropathy and with associated long-term consequences like deformities and disabilities. The proposed portals of entry are the skin and upper respiratory tract. The importance of the skin as a probable route of entry of *Mycobacterium leprae* into the body and the possibility that the organism may survive in the environment for several days may augment that it is likely that the bacilli may enter the skin by inoculation such as vaccination, tattooing, or trauma to the skin.

We report an interesting case of leprosy in a Filipino, arising from a burn scar ten years ago. Skin punch biopsy on two sites revealed hypertrophic scar and Hansen's disease, borderline tuberculoid. Patient was started on recommended standard regimen for paucibacillary leprosy with noted clinical improvement.

Keywords: Hansen's disease; *M. leprae*; Upper respiratory tract; Skin

Introduction

Leprosy is a chronic granulomatous disease caused by the obligate intracellular pathogen *M. leprae* [1-4]. The pathogen principally affects the skin and peripheral nervous system. Advanced disease is characterized by disfiguring mutilations [5]. There are two systems to classify patients, one is the Ridley-Jopling classification system which uses clinical and histopathological features and the Bacteriological Index (BI) to identify five forms of leprosy or the WHO classification which classifies patients as Paucibacillary (PB) or Multibacillary (MB), based in the number of lesions, presence of nerve involvement and identification of bacilli on slit-skin smear. The recommended standard regimen for paucibacillary leprosy is rifampicin 600 mg plus dapsone 100 mg daily, to complete 6 blister packs. The recommended standard regimen for multibacillary leprosy is rifampicin 600 mg once a month plus dapsone 100 mg daily plus clofazimine 300 mg once a month and 50 mg daily to complete 12 blister packs [1,2].

Transmission of *M. leprae* is assumed to be mainly from untreated MB patients. The route of transmission is not however definitely known. The proposed portals of entry are the skin and upper respiratory tract [2]. The importance of the skin as a probable route of entry of *M. leprae* into the body and the possibility that the organism may survive in the environment for several days may augment that it is likely that the bacilli may enter the skin by inoculation such as vaccination, tattooing, or trauma to the skin. Inoculation leprosy does not appear to be of much significance in nature because of its sporadic reports [3]. However, the occurrence of leprosy lesions localized to a previous burn scar can supplement the importance of the skin as a portal of entry for *M. leprae.*

Different cases of inoculation leprosy have so far been reported. In one report of indeterminate leprosy developing at a traumatic site, authors suggested the likelihood of contamination of the wound with *M. leprae* in the leprosy hospital environment, or through flies attracted to the wound site. There is also a report of a surgeon from a non-endemic area acquiring leprosy accidentally from a patient during an operation. Therefore, it seems more likely that multiple factors might be contributing to the development of a leprosy lesion after the initial exposure to the organism [6].

Case Presentation

A 18-year old, Filipino female presented with a ten year history of multiple, skin-colored to erythematous hypertrophic plaques on the right side of the chest and back due to a scald burn injury which was managed as a case of hypertrophic scar secondary to second degree burn and given with topical and intralesional steroids with moderate improvement. Seven months prior to consult, the patient noted appearance of new lesions seen as erythematous patches and plaques on the previous burn scar on the chest which expanded to involve the right side of the face, neck, upper arm and back associated with hyposthesia. Patient was then referred to dermatology service for further evaluation and management. There were no other family and household members with similar lesions.

Skin examination revealed multiple erythematous to hyperpigmented patches and plaques on the right side of the face, neck, upper arm, axilla, chest and back (Figure 1A-D). There was no thickening and/or tenderness of any peripheral or local cutaneous nerve. Caloric test was done which showed sensory deficit on the

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lesional skin on hot, cold, pinprick at 80%, 80% and 90% respectively. Primary clinical impression was hypertrophic scar with post inflammatory hyperpigmentation and to consider Hansen's disease. Laboratory tests did not identify any hematological or biochemical abnormalities. Slit skin smear revealed a negative result.

Skin punch biopsy on 2 sites was done. First specimen showed normal epidermis and the dermis revealed thickened collagen bundles and few dilated blood vessels (Figure 2A-C). The second specimen showed thinning of the epidermis and the dermis revealed pigment-laden macrophages and a dense, pandermal inflammatory infiltrate of lymphocytes, foamy and epitheloid histiocytes and multinucleated giant cells (Figure 3A-C). Modified Acid-Fast stain (MAF) was negative.

Histopathological diagnosis of the first specimen was hypertrophic scar and the second specimen was Hansen's disease, borderline tuberculoid.

Upon clinico-histopathologic correlation, our final diagnosis is Inoculation leprosy and Hypertrophic scar. Patient was started on Multi-Drug Therapy (MDT) for paucibacillary leprosy for 6 months with remarkable improvement (Figure 4A-D).

Discussion

Despite the elimination of leprosy as a public health problem (defined as achieving less than 1 case per 10,000 population) globally in 2000 and at a national level in most countries by 2005, leprosy cases continue to occur. Over 200,000 new leprosy cases were reported in 2016 [1]. The development of leprosy lesions at the site of tattooing, wounds or minor abrasions suggests the skin as a possible portal of entry for *M. leprae*. Limited is known about this occurrence due to its sporadic reports and it has been described variously due to lack of consensus about its nomenclature. The term "inoculation leprosy" was retained as described by earlier workers [3]. The incubation period varied from 15 days to 20 years before the onset of first leprosy lesion. The earliest case of tattoo inoculation tuberculoid

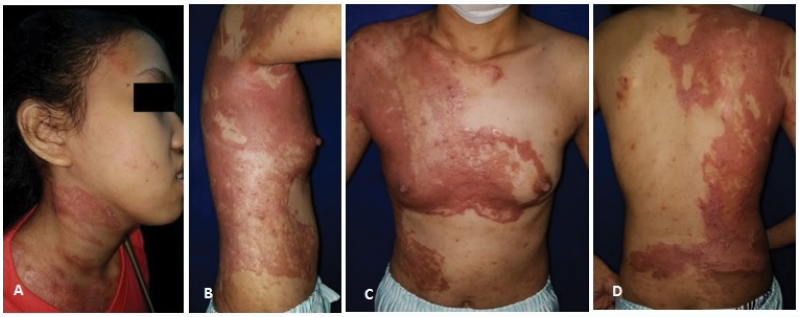

Figure 1: Multiple erythematous to hyperpigmented patches and plaques on the right side of the face, neck (A), upper arm (B), axilla (B), chest (C) and back (D).

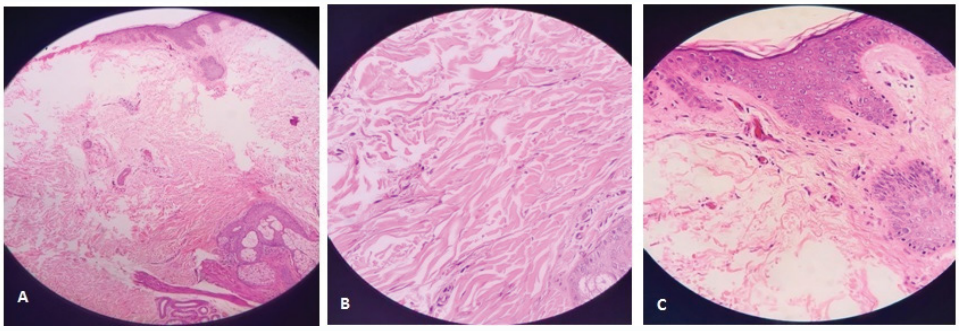

Figure 2: H & E of first specimen, (A) Normal epidermis and the dermis revealed thickened collagen bundles and few dilated blood vessels (H & E × 100). (B) Thickened collagen bundles (H & E × 400). (C) Dilated blood vessels (H & E × 400).

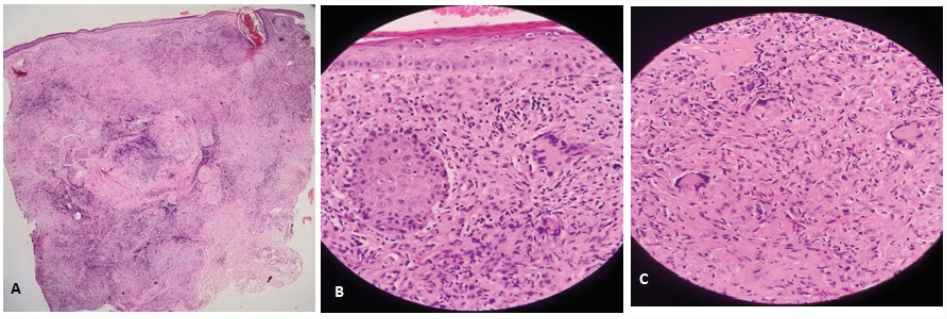

Figure 3: H & E of second specimen. (A) Thinning of the epidermis and the dermis revealed a dense, pandermal inflammatory infiltrate of lymphocytes, foamy and epitheloid histiocytes and multinucleated giant cells (H & E × 100). (B) The dermis showed pigment-laden macrophages, inflammatory infiltrates of lymphocytes, foamy and epitheloid histiocytes (H & E × 400). (C) Multinucleated giant cells (H & E × 400).

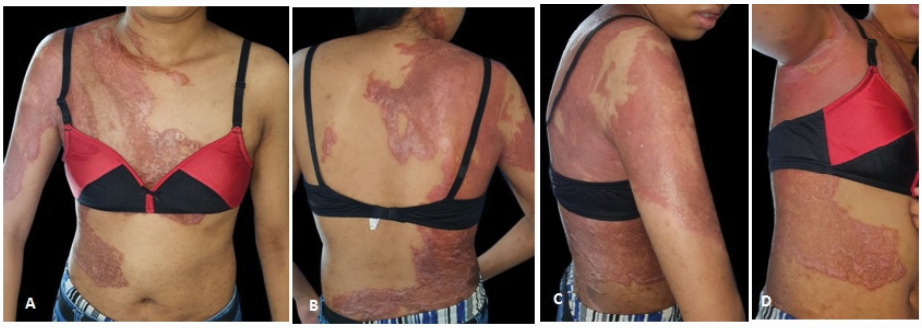

Figure 4: Multiple erythematous to hyperpigmented patches and plaques on the right side of the face, neck (A), upper arm (B), axilla (B), chest (C) and back (D).

leprosy developing 6 years after tattooing was reported by Lowe and Chatterjee in 1939. Subsequently, several cases of tattoo inoculation leprosy were also reported, mostly from India. In Chhattisgarh state, large numbers of leprosy cases are reported and about 35% of them belong to multibacillary type. Ignorance about sterilization practices and hygiene are common. Tattooing here is done for a group of patients using sewing needles tied together by the untrained, illiterate tattoo artist, usually an elderly lady who moves with her basket from one village to another. There is likelihood, that along with the needle the lepra bacilli might be getting inoculated from a multibacillary case into the skin of some subsequent subjects. There are other cases reported at the site of roadside injuries, vaccinations and dog bite [7].

The clinical appearance of inoculation leprosy manifests itself either in the form of Tuberculoid (TT), Borderline Tuberculoid (BT), or Maculoanesthetic (MA). Usually, the lesions are well-defined hypopigmented macules or papules. Erythema or pigmentation may also be seen. Lesions are hairless and anhidrotic with demonstrable sensory deficit of temperature, touch, and pain. Nerves proximal to the lesions may be thickened and/or tender. The lesions usually develop on and around the site of inoculation [3]. The development of leprosy lesions in this patient after, and at the site of burn scar, and its exclusive localization to that site, suggests that this could be a case of inoculation leprosy.

Inoculation alone or subsequent infection of the burn wound may not be the only factors responsible for the leprosy lesions, as the incubation period was long. *M. leprae* might enter the body primarily through the upper respiratory tract, and could be subsequently drawn to the site of injury to produce skin lesions depending on the local factors, local temperature and possible local factors intrinsic to a wound [6]. However, no other family and household members with neither similar lesions nor any exposure to an infected person with leprosy were noted.

Slit-skin smear of the patient revealed a negative result. Slit-skin smears to demonstrate acid-fast bacilli are of little value in scarred/unscarred leprosy following inoculation. Nevertheless, demonstration of the bacilli forms the essential diagnostic component [3]. The diagnosis of leprosy is based on the presence of at least one of three cardinal signs: (1) definite loss of sensation in a pale or reddish skin patch; (2) thickened or enlarged peripheral nerve with loss of sensation and/or weakness of the muscle supplied by that nerve; or (3) presence of acid-fast bacilli in a slit-skin smear [1].

Patients belonging to the Tuberculoid (TT) and Borderline Tuberculoid (BT) are termed Paucibacillary (PB) and those assigned to the Mid-Borderline (BB), Borderline Lepromatous (BL), and Lepromatous (LL) are termed Multibacillary (MB). This is used as the basis of guiding Multi-Drug Treatment (MDT) [2]. Taking all this into consideration, on clinical, histopathological and bacteriological assessment of the patient, the working diagnosis of the patient was Hansen's disease, borderline tuberculoid and hypertrophic scar. Inoculation leprosy should be treated by the recommended standard regimen for PB leprosy [3]. In cases of positive slit-skin smears, patients will be diagnosed as MB leprosy and can be started with standard regimen for MB leprosy [7]. The patient was given MDT-PB blister packs and silicone gel for the hypertrophic scar with noted improvement of lesions after completion of 6 blister packs.

Conclusion

There are various evidences to indicate that transmission of leprosy may occur through inoculation. Diagnosis should include the clinical, bacteriologic, histopathologic, and immunologic features. It is equally imperative to obtain a history of injury or trauma in the patient [3]. The importance of asepsis and sterilization should be emphasized to tattooers and vaccinators. Inoculation leprosy, although rare, may occur in a setting of second degree burn.

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