Dermatology Case Reports

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Diffuse Necrotic Ulcerations Revealing Lepromatous Leprosy with Lucio's Phenomenon

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Abstract

We report a 62-year-old farmer who consulted with multiple chronic wounds over his limbs and chest. Physical examination revealed multiple ulceration, madarosis and infiltration of ear lobes. Slit skin smear showed numerous acid-fast bacilli consistent with a diagnosis of lepromatous leprosy associated with Lucio's phenomenon. Administration of thalidomide together with MDT resulted in a huge improvement.

Keywords: Lepromatous leprosy; Lucio's phenomenon; Mali

Introduction

Leprosy or Hansen's disease is a chronic infectious disease caused by *Mycobacterium leprae* (*M leprae*). Peripheral nerves (Schwan cells), and skin (macrophages) are the predilection sites of *M leprae*. The clinical manifestations of leprosy depend on the immune response (Th1) developed by the host after exposure. Based on this response, five types of leprosy have been described on a spectrum varying from tuberculoid leprosy to lepromatous versions [1]. Lepromatous leprosy is characterized by the occurrence of acute inflammatory episodes known as "leprosy reactions". Lucio's phenomenon is an unusual type of reaction initially described in 1852 by Lucio and mostly encountered in South America [2]. Patients usually present with multiple necrotizing skin ulcerations that may result in death. So far, only one case was reported in West Africa in a Senegalese patient [3]. We report here the first Malian case of Lucio's phenomenon.

Case Report

Mr. S.F, a 62-year old farmer was referred to our department of Dermatology for diffuse chronic wounds. The onset of the disease was traced back to two years prior to the referral. The disease started with a spontaneous depigmentation of his face, which has steadily evolved with tiny red spots that ulcerated later on. At the physical examination, we found numerous ulcerated lesions on his chest and limbs (Figure 1) along with madarosis, infiltrated earlobes, enlarged nose, congestive nasal mucosa and a swelling of the wrists. Examination of peripheral nerve found a hypertrophy of the trunk of the radial, ulnar, fibular and great auricular nerves. The rest of the physical examination was normal. Except an anemia with 9 g/dl, the renal function, liver enzyme and blood sugar tests were normal. Serological tests for HIV and syphilis were also negative. The Fit Faraco slit smear skin test, revealed numerous acid-fast bacilli at various sites (bacillary index 5+ on Ridley scale) (Figure 2). The histopathology of ulceration also showed numerous acid-fast bacilli associated with polymorphonuclear infiltrate and histiocytes in the dermis. Due to the lack of equipment for the isolation of DNA, the identification of the *Mycobacterium* species was not performed. The diagnostic of the lepromatous leprosy with Lucio's phenomenon was mentioned. While the 15 days treatment with antiseptic plus antibiotics was unsuccessful on the ulcerations, a huge improvement was noted within ten days with Thalidomide plus WHO Multidrug therapy (MDT) for multibacillary leprosy that contains Rifampicin, Clofazimine and Dapsone.

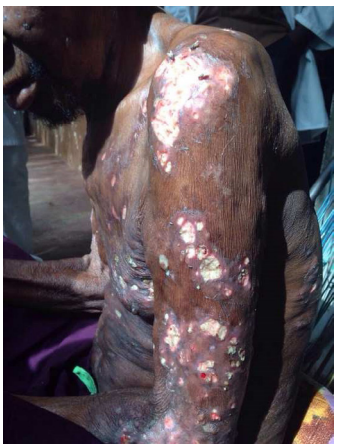

Figure 1: Ulcerated lesions on his chest and limbs along with madarosis, infiltrated earlobes, enlarged nose, congestive nasal mucosa and a swelling of the wrists.

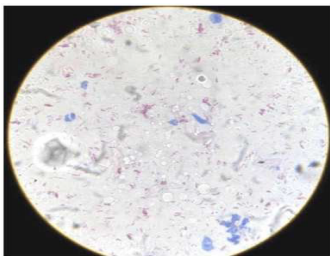

Figure 2: Fit Faraco slit smear skin test, revealed numerous acid-fast bacilli at various sites (bacillary index 5+ on Ridley scale).

Discussion

We report a case of non-treated diffuse lepromatous leprosy who presented multiple spontaneous skin ulcerations, numerous acid-fast

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bacilli in skin slit smear test. Despite the lack of histological evidence of thrombosis and endothelial cells invasion by *M leprae* that characterize the Lucio'phenomenon (LP), the clinical features presented by our patient met this disease diagnosis criteria. These symptoms cannot be attributed to any disease other than leprosy. The inefficacy of antibiotics also ruled out the possibility of common bacterial infection initially suspected.

Lucio's phenomenon or Lucio reaction is a severe form of lepra reaction type 2. Initially described in Mexico, it has rarely been reported in Africa [3,4]. As observed in our case, it affects patients with diffuse lepromatous leprosy without nodules and begins clinically with erythematous spots of limbs leading to bulla, epidermal necrosis and ulceration [2]. Systemic symptoms are sometimes associated. Age and gender are not considered as risk factors. In a Brazilian cohort, five of the 12 cases were male with an age range from 27 to 67 years old; death occurred in four cases indicating that lethality is not uncommon in LP. Additional cases were reported from India and Sri Lanka [4-6].

The pathophysiology of LP is not well understood. The clinical feature is thought to be the results of heavy bacterial colonization of the capillary endothelium resulting in the formation of immune complexes, necrotizing vasculitis of superficial and medium size vessels and dermal necrosis seen in histological staining [7,8]. These features were absent in our patient because the skin biopsy was likely focused on ulceration rather than erythematous lesions.

Until recently, *M leprae* was considered as the sole causative agent of all form of leprosy [5]. In 2008, a new mycobacterial species named *Mycobacterium lepromatosis* was identified as the causative agent of diffuse lepromatous leprosy in a Mexican patient. This bacterium, different but very close to *M leprae*, has also repeatedly been reported in patients with Lucio's phenomenon supporting the evidence that LP results in M lepromatosis infection. For technical reasons, this investigation was not performed in our patient [8,9].

Due to its rarity, the treatment of LP is not well-codified. In our patient, Thalidomid associated with multidrug therapy (MDT) for multibacillary leprosy were used with satisfactory outcomes. Unlike the controversies on corticosteroid use, a strong agreement exists on the use of MDT. Pentoxifilin, which improves blood flow and prevents from platelet aggregation has been proposed.

Conclusion

The number of leprosy patients worldwide contrasts with the rarity of LP. Besides typical cutaneous lesions, necrotic wounds must be among the cutaneous signs that may reveal leprosy. Further studies focusing on DNA isolation from the causative agent in patients suspected of LP or diffuse lepromatous leprosy are appealing.

Conflict of interest

The authors declare they have no conflict of interests.

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