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Necrotic Ulcerations Unveiling a Lucio's Phenomenon in a Moroccan Patient

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Abstract

Lucio's phenomenon is an atypical manifestation of leprosy, few cases have been described around the world, predominately in Mexico. It's characterized by purpuric lesions and necrotic ulcerations of the skin, described as a vasculitis like condition. The clinical diagnosis is challenging in a non-endemic area. It's potentially fatal. We report a case of Lucio's phenomenon in a previously undiagnosed leprosy Moroccan patient whose acute manifestations were conductive to the leprosy's diagnosis.

Keywords: Leprosy; Lucio's phenomenon; Necrotic ulcerations

Introduction

Leprosy is an infectious disease that may present different clinical forms depending on host immune response to *Mycobacterium leprae*. During Hansen's disease an acute inflammatory condition may occur before, during or after the treatment called leprosy reaction. Lucio's phenomenon (LP) is an uncommon form considered as a leprosy reaction for some authors while others consider this entity a separate entity, called Lucio's leprosy due to a novel discovered species; *M. lepromatosis*. We present a rare occurrence of LP in undiagnosed Moroccan patient with lepromatous leprosy [1].

Case Presentation

A 58-year-old Moroccan patient had 5 years history of dilated cardiomyopathy, chronic rhinitis, chronic pleuritis, hemodialysis for kidney failure. No risk behavior or a history of bowel, skin or joint affections has been reported. He developed 14 years ago non painful, non pruritic vesiculo-bullous eruption and swelling of the lower limb. He consulted several private practitioners who prescribed some oral and topical treatments (nature unknown) with no improvement. 4 years ago, the cutaneous lesions evolved to painless ulcers on lower and upper limb.

Physical examination showed afebrile patient, asymptomatic ulcerations mostly below elbow and knees, areas of necrosis involving the dorsal surface of the extremities of both upper and lower limbs as well as the pinnae. The skin was dry to touch. Hertoghe signe was positive. There was an infiltration of both ear lobes, madarosis, enlarged nose with obstruction, crusting, bleeding and hyposmia. We observed unilateral gynecomastia, bilateral epididymo-orchitis with achromic macules, joints deformities, amputation of the distal phalange of the right little toe and onychodystrophy (Figure 1-3). Respiratory examination found few bilateral crepitations. Examination of the abdominal revealed splenomegaly. Neurological examination demonstrated hypoesthesia in the glove and stocking areas of the distal extremities and the absence of nerve thickening. Generalized diffuse lymphadenopathy was found. Blood analysis revealed aregenerative normocytic normochromic anaemia haemoglobin 9g/dl, hyperleukocytosis with a predominant neutrophillic count. Serum ferritin, Erythrocyte sedimentation rate and C-reactive protein were highly elevated. Serum protein electrophoresis demonstrated beta-gamma bridging and markedly diminished albumin levels. Liver, lipid, blood glucose tests were normal. Anti-nuclear antibodies, rheumatoid factor, anti-neutrophil cytoplasmic auto antibodies P and C and complement 3 and 4 levels were normal while type III mixed cryoglobulinemia was positive. Hepatitis C antibodies VDRL, Hepatitis B surface antigen and HIV antibody were negative.

A chest x-ray showed pleural effusion and cardiomegaly, c-TAP showed axillary, mediastinal and below the diaphragm lymph nodes associated with splenomegaly. Leg ulcer biopsy disclosed epidermal ulceration associated with inflamed granulation tissue, leukocytoclastic vasculitis, superficial and deep infiltrate containing histiocytes while Ziehl-Neelsen staining revealed no evidence of microorganisms (Figure 4). Lymph node and bone marrow biopsies revealed dense inflammatory infiltrate with numerous neutrophils and histiocytes, specimen was found positive to Ziehl Neelsen. Direct examination was positive to an atypical mycobacteria infection, PCR for M. tuberculosis was negative. Due to the lack of equipment neither the isolation of DNA for the identification of the *Mycobacterium leprae* nor the culture were performed.

The diagnosis of atypical mycobacterial lymphadenitis was initially made. The patient received 6 months multidrug treatment including clarithromycin, Ethambutol, Rifabutin, ciprofloxacin and azithromycin with mild improvement and relapsing while tapering of the rifabutin. Though leprosy was highly considered in the differential diagnosis, the uncertain nature of this specific manifestation didn't directly lead us towards its diagnosis. Further investigations were made strengthening our suspicion as Electroneuromyography examination showed mononeuritis multiplex in the arms, the lower limb wasn't examinated due to ulcerations. Endoscopic endonasal biopsy demonstrated an inflammatory infiltrate with numerous histiocytes and was found positive to Ziehl Neelsen (Figure 4). In context of the clinic-pathological features the diagnosis of LP secondary to lepromatous leprosy was favored associated with type III mixed cryoglobulinemia. The multibacillary multidrug antileprosy therapy, as per the WHO recommendation was started plus wound dressing. Clinical improvement was noticed after 1 month.

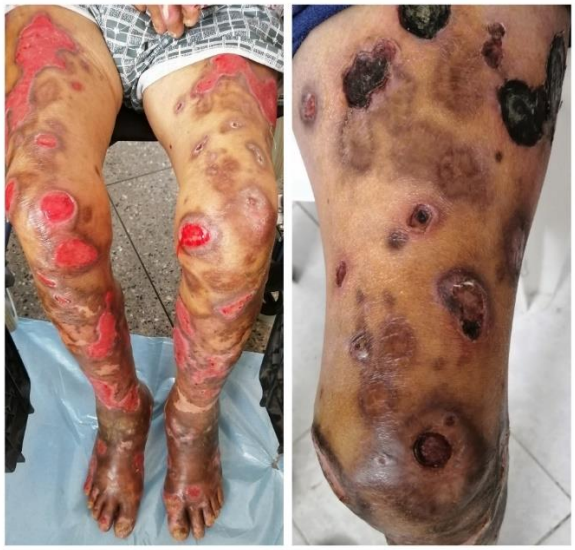

Figure 1: Symmetrical involvement of the lower limb showing large denuded areas secondary to ulceration, necrosis and areas of cicatrization.

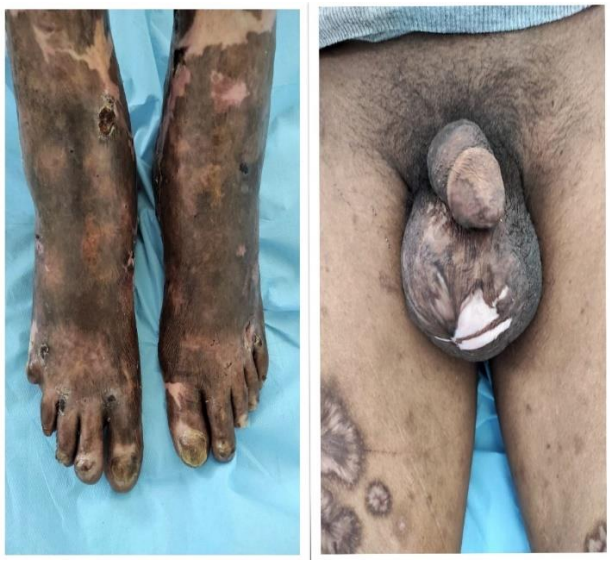

Figure 2: Ulceration and necrosis of the feet, amputation of the distal phalange of the left little toe and onychodystrophy. Bilateral epididymo-orchitis with achromic macules.

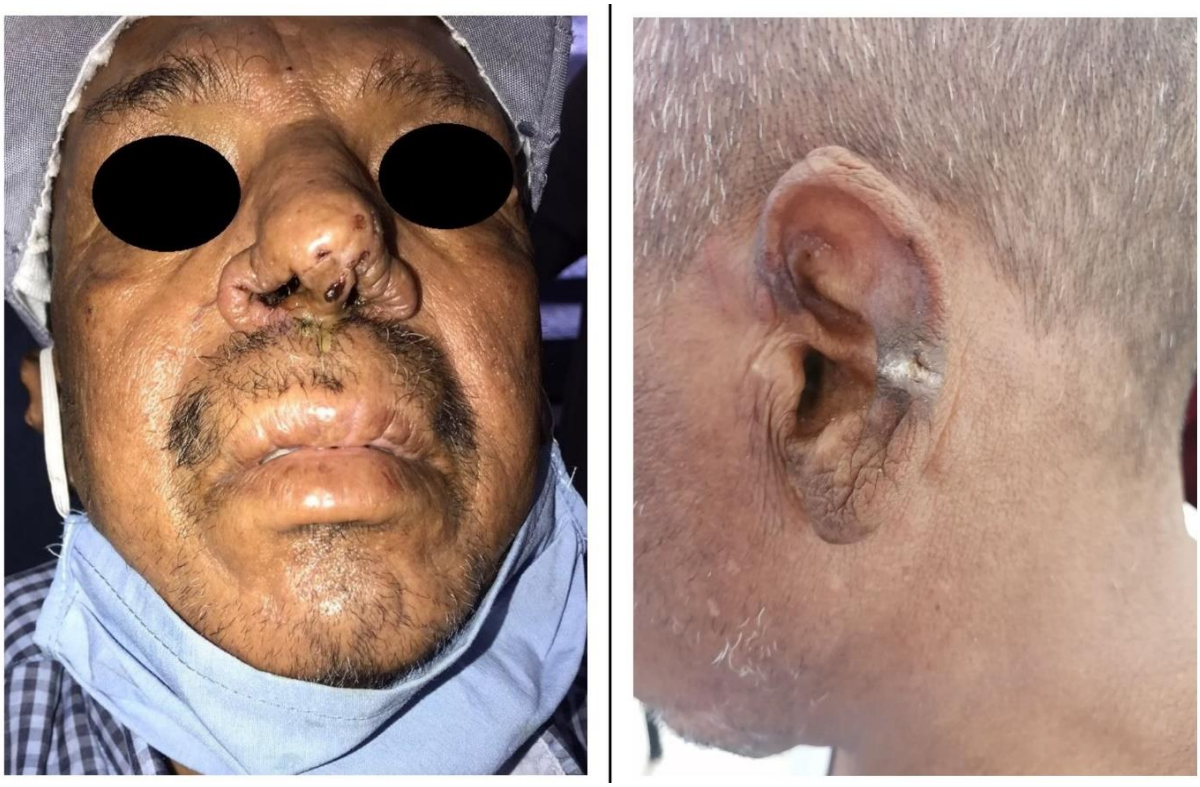

Figure 3: Diffuse facial involvement with shiny skin, madarosis and infiltration, involvement of the ear with necrotic lesions associated with crusting.

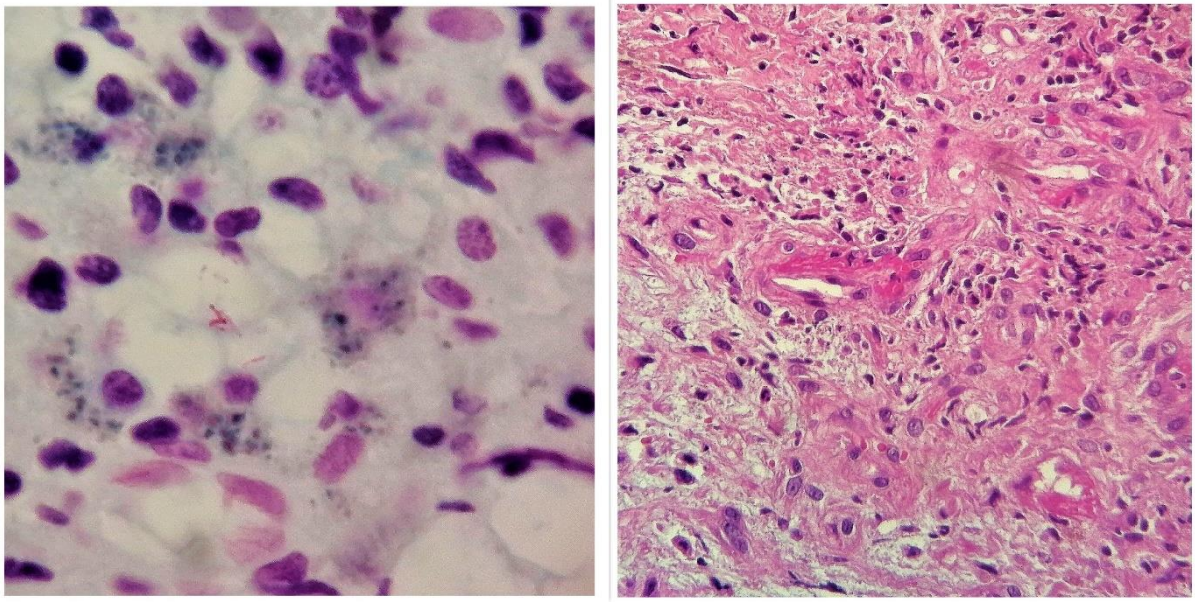

Figure 4: In the left histology of nasal endoscopy biopsy depicts wade fite stain demonstrating the presence of (red) bacilli. In the right, histology of skin biopsy. H and E stain endothelial proliferation of the medium-sized vessels of the mid-dermis, neutrophilic infiltration and necrotizing vasculitis of the small vessels of the superficial dermis.

Discussion

Morocco achieved the goal of leprosy elimination as a public health problem in 1991. To continue working towards eradication of leprosy, single dose rifampicin chemoprophylaxis was introduced for household contacts of registered and newly diagnosed leprosy patients in 2012 with reduction in case detection of 16% per year [2]. Hence, we report a case of non-treated diffuse lepromatous leprosy with multiple spontaneous skin ulcerations. Despite of epidemiological control and the lack of histological evidences that characterize LP, the clinical manifestations and the inefficacy of antibiotics cleared out the path to reach the diagnosis making it the first reported case of LP in Morocco. In leprosy, ulcers tend to occur on the extremities due to trophic mechanisms in relation to severely sensory-impaired skin [3]. Ulcers can also occur as Type 1 or Type 2 leprosy reactions as well as LP, reflecting the host's immune responses to leprosy bacillus [3]. Initially described in Mexico and rarely reported in Africa [1], LP may present as a new aspect in an already established, yet untreated leprosy patient. It can also be the preliminary symptom introducing the lepromatous version of the disease. Rarely it can present in a fully treated diffuse lepromatous leprosy patient in the absence of bacilli even years later [1]. The skin manifestations classically occur as generalized dryness and shininess with a myxedematous appearance called "lepra bonita". There may also be hair loss involving any area of the body. The ongoing vasculitic process causes erythematous spots leading to bulla, epidermal necrosis and ulceration. The affection can be diffuse with a predilection for the extremities [3,4]. Lesions may be destructive in superficial regions as pinna. Systemic symptoms are sometimes associated. In a Brazilian cohort, five of the 12 cases were male with an age range from 27 to 67years old; death occurred in four cases indicating that lethality is not uncommon [5]. The pathophysiology is poorly understood. The clinical feature is thought to be the results of bacterial colonization of the capillary endothelium resulting in the formation of immune complexes, necrotizing vasculitis of superficial, medium size vessels and dermal necrosis seen in histological staining [4,6]. The presence of cryoglobulinemia in lepromatous leprosy as in our patient is frequent and is due to complex phenomenon.

The level of circulating immune complexes may parallel disease activity [7]. Due to its rarity, the treatment of LP is not well-codified. The pure form may only necessitate the usual MDT without requiring thalidomide or systemic steroids to be included, though these have also been opted to be used in severe cases [8].

Conclusion

Leprosy diagnosis may be challenging due to various clinical manifestations and long incubation period. Leg ulcers are a rare presentation and doctors should be aware and consider this condition in their differential diagnosis.

REFERENCES

1. Herath S, Navinan MR, Liyanage I, et al. Lucio's phenomenon, an uncommon occurrence among leprosy patients in Sri Lanka. BMC Res Notes. 2015; 8: 672.
2. Thomas Hambridge, Shri Chandran. *Mycobacterium leprae* transmission characteristics during the declining stages of leprosy incidence: A systematic review. 2021.
3. Laurence DM, Spanoudi KI, Liliane Marot, et al. Leg ulcers as an atypical presentation of *Mycobacterium leprae* infection. Europ J Dermatol. 2018; 28: 105-106.
4. Faye O, Dicko A, Traoré B, et al. Diffuse necrotic ulcerations revealing lepromatous leprosy with lucio's phenomenon. Dermatol Case Rep. 2017; 2: 136.
5. Rocha RH, Emerich PS, Diniz LM, et al. Lucio's phenomenon: Exuberant case report and review of Brazilian cases. An Bras Dermatol. 2016; 91: 60-63.
6. Azulay-Abulafia L, Pereira Spinelli L. Lucio phenomenon: Vasculitis or occlusive vasculopathy? Der Hautarzt; Zeitschrift fur Dermatologie, Venerologie, und verwandte Gebiete. 2006; 57: 1101-1105.
7. Bonomo L, Dammacco FM. Cryoglobulinemia in lepromatous leprosy: An immune complex phenomenon. Int J Lepr Other Mycobact Dis. 1971; 39: 554-555.
8. Peixoto AB, Portela PS, Leal FR, et al. Lucio's phenomenon. Case study of an exceptional response to treatment exclusively with multibacillary multidrug therapy. An Bras Dermatol. 2013; 88: 93-96.