Volume 72, Number 1
Printed in the U.S.A.
(ISSN 0148-916X)

Lepromatous Leprosy with Extensive Unusual Ulcerations and Cachexia. Is It the First Case of Lucio's Phenomenon from Iran?[1]

ABSTRACT

We report a 33-year-old Iranian woman with widespread ulcerative lesions in the setting of lepromatous leprosy. We think that the sudden appearance of the characteristic necrotic lesions in the absence of fever and other systemic manifestation, and in accordance with epidermal necrosis and the presence of large numbers of AFB in the endothelium are all in favor of the diagnosis of Lucio's phenomenon for this patient. To our knowledge this is the first patient who may have had this phenomenon reported from the Middle East.

RÉSUMÉ

Nous rapportons ici le cas d'une femme iranienne de 33 ans qui a présenté des lésions disséminées d'ulcérations cutanées dans le contexte d'une lèpre lépromateuse. Nous pensons que la soudaine apparition de lésions nécrotiques caractéristiques, en l'absence de fièvre et d'autres manifestations systémiques, et en tenant compte de la nécrose épidermique et de la présence de très nombreux bacilles AAR dans l'endothélium, est en faveur du diagnostic de phénomène de Lucio pour cette patiente. A notre connaissance, il s'agit du premier cas rapporté au Moyen-Orient qui a probablement été victime de ce phénomène.

RESUMEN

Presentamos el caso de una mujer Iraní de 33 años con lesiones ulcerativas diseminadas asociadas a lepra lepromatosa. Pensamos que la aparición súbita de las lesiones necróticas características, la ausencia de fiebre y otras manifestaciones sistémicas, la necrosis epidérmica y la presencia de grandes números de bacilos ácido-resistentes, indican un caso de fenómeno de Lucio en esta paciente. Hasta donde sabemos, este es el primer caso de un paciente con fenómeno de Lucio en el Medio Este.

TO THE EDITOR:

Northern Iran was an endemic area for leprosy until a few years ago and almost all of the clinical variants of leprosy have been seen in this area. We report here a case of lepromatous leprosy with unusual extensive ulcerations that we think might be the first case of Lucio's phenomenon from the Middle East.

A 33-year-old woman with a long history of skin lesions was referred to our service after her lesions had started to become generalized and necrotic, a few days previous. She was inhabitant of a far located rural area. Her father was a known case of lepromatous leprosy and had been treated with dapsone as monotherapy during 1970's and later with standard multiple drug therapy (MDT) for leprosy, but the patient had never visited a dermatologist nor received any antileprosy medications.

On admission to hospital, the patient was cachectic and in poor general condition. An obvious leonine face with nearly complete loss of eyebrows and eyelashes, and extensive ecchymotic patches in association with deep ulcerative lesions were seen on her body, especially on the extremities (Figs. 1 and 2). On laboratory examinations, a full blood count showed anemia with Hb 4.8 g/dl. The white cell count was normal, but platelet count was raised to 7×10^5/dl. The erythrocyte sedimentation rate was 72 mm/hr. Rheumatic factor and C-Reactive Protein were weekly positive but other laboratory tests including, renal function tests, liver enzymes, electrolytes, and coagulation tests were all in the normal range.

Slit skin smears from different locations showed multiple acid-fast bacilli (AFB),

[1] Received for publication 26 March 2003. Accepted for publication 22 December 2003.

Reprint requests to: Omid Zargari, M.D., Hafez St., Booali Bldg., Rasht 41378, Iran.
E-mail: ozargari@iranderma.com

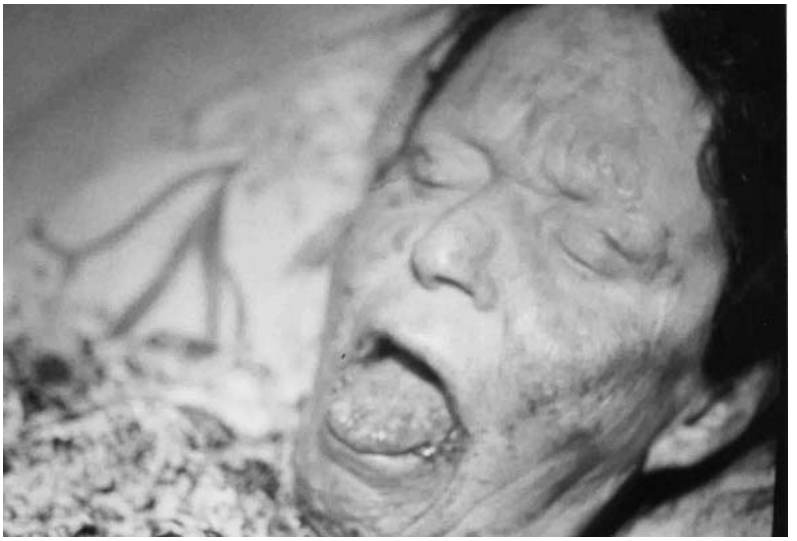

FIG. 1. Diffuse infiltrative lesions on the face in association with significant loss of eyebrows.

and a skin biopsy from an ecchymotic patch revealed swollen endothelial cells with fibrinoid necrosis and a mixed inflammatory infiltrate with nuclear dusts and an atrophic epidermis with focal necrosis. In the dermis, enormous numbers of AFB were seen with Ziehl-Neelsen staining. AFB were also present in the form of clumps in the vessel walls. We started supportive care and a standard antileprosy therapy including dapsone 100 mg daily, clofazimine 50 mg daily and rifampicin 600 mg monthly for the patient. A combination of systemic antibiotics including a 3rd generation cephalosporin and vancomycin was also started after the patient became febrile. During the admission, the patient's hemoglobin dropped and she developed hamaturia and pancytopenia and unfortunately died at the 35th day of admission because of severe sepsis.

Lucio's phenomenon is a severe necrotizing reaction occurring in the diffuse, lepromatous leprosy. Lucio and Alvardo first described this phenomenon in 1852. Latapi and

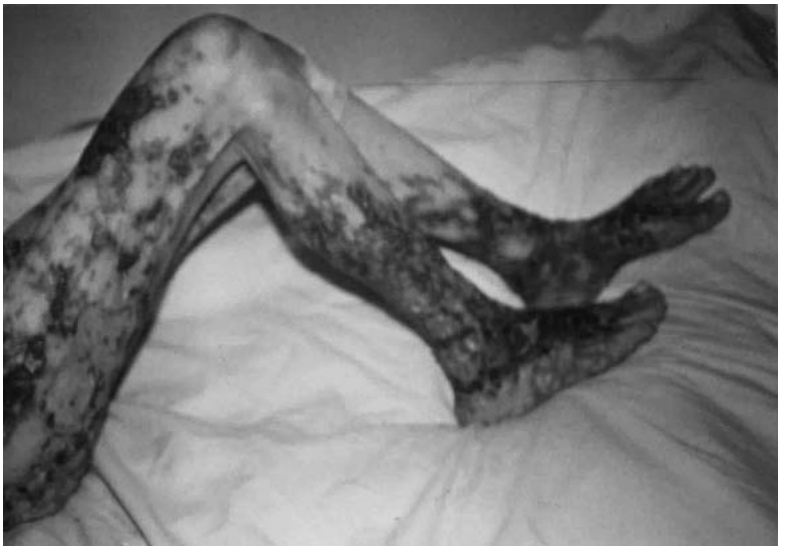

FIG. 2. Extensive ecchymotic patches and deep ulcerative lesions on the extremities.

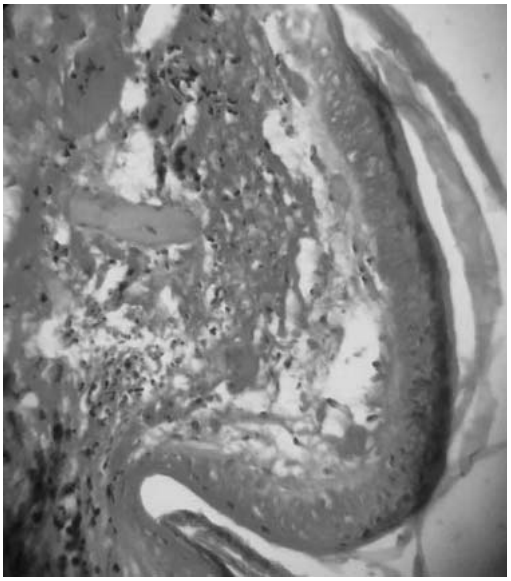

FIG. 3. Swollen endothelial cells with fibrinoid necrosis and a mixed inflammatory infiltrate (Hematoxylin-Eosin ×100).

Chevez later denoted this reaction in 1948 as Lucio's phenomenon ([3]). This type of reaction is most commonly seen in Mexico and Central America ([3, 6, 8]), and is rare outside America, although Saoji, *et al*. reported two cases of this phenomenon from India ([9]), and Ang, *et al.* recently reported two Chinese men with fatal Lucio's phenomenon ([1]).

Painful macules or plaques progressing to ulcers characterize Lucio's phenomenon. Features of the underlying lepromatous leprosy commonly described include diffuse thickening of facial skin, maderosis, and destructive rhinitis ([6]). Anemia, lymphadenopathy, splenomegaly, hypoalbuminemia, hypocalcemia and polyclonal gammopathy are among the other reported manifestations ([4, 6]).

Rea, *et al*. have hypothesized that patients with Lucio's phenomenon have an exceptionally deficient defense mechanism, allowing unrestricted proliferation of AFB in endothelial cells, facilitating contact between bacterial antigens and circulating antibody and leading to infarction. Also, this nadir of resistance allows unimpeded dissemination of AFB, accounting for the clinical features of diffuse non-nodular leprosy ([7]). Most cases of Lucio's phenomenon have been reported to have a leukocytoclastic vasculitis as the underlying pathologic abnormality, although some cases of Lucio's phenomenon may be caused by vascular damage due to direct invasion of *Mycobacterium leprae* and not necessarily by leukocytoclastic vasculitis ([4]). Deposits of mixed-type cryoglobulins

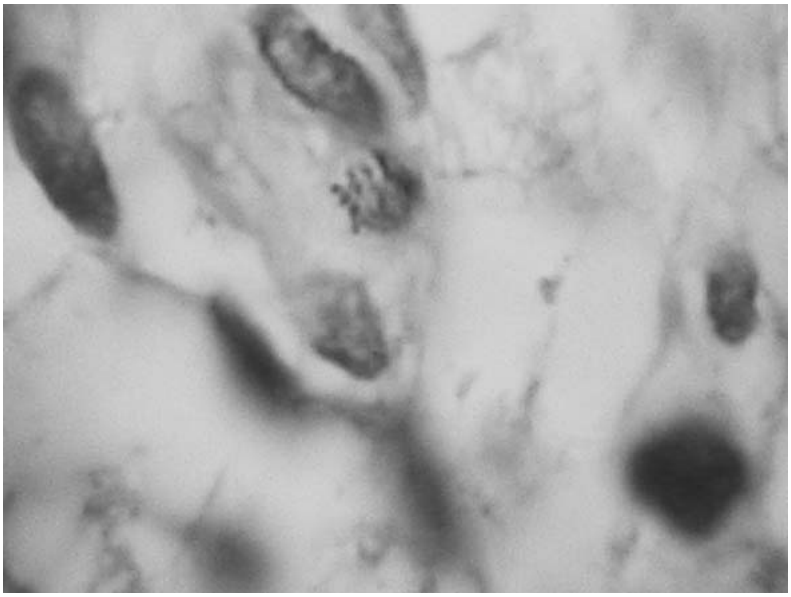

FIG. 4. Aggregates of acid-fast bacilli in the endothelial walls (Ziehl-Neelsen ×400).

(IgG, IgA, IgM, C3, and C1q) have been observed in dermis vessels affected by vasculitis of the Lucio's phenomenon type, suggesting a mechanism mediated by deposits of immune complexes ([5]).

Lucio's phenomenon occurs in patients with undiagnosed and untreated leprosy, whereas the erythema nodosum leprosum (ENL) may occur in any type of lepromatous leprosy, and frequently occurs after starting the treatment. The main clinical differences between the Lucio's phenomenon and ENL are that the former is an ulcerative reaction occurring in the absence of cutaneous nodules, whereas the latter usually present as tender cutaneous nodules that rarely ulcerate ([1]). Absence of fever, leukocytosis, tenderness, and other systemic presentations such as arthritis, neuritis, and iridocyclitis, and failure to respond to thalidomide are among the other distinct features of Lucio's phenomenon ([6, 10]). Histologically, Lucio's reaction can be distinguished from ENL by epidermal necrosis and by necrotizing vasculitis manifesting necrosis in the walls of superficial vessels and severe, focal endothelial proliferation of mid-dermal vessels. Furthermore, the numbers of AFB are much more in the lesions of Lucio's phenomenon in comparison with The ENL ([7]). In addition to ENL, in the differential diagnosis of Lucio's phenomenon, vasculitis of other origins should be considered, such as the antiphospholipid antibody thrombotic syndrome associated with lepromatous leprosy ([2]).

Although we were not able to assay cryoglobulins, cryofibrinogens, and antiphospholipid antibodies in this patient, normal coagulation tests including partial thromboplastin time make this syndrome improbable for this case.

—Javad Golchai, M.D.,

Associate professor,

—Omid Zargari, M.D.,

Assistant professor,

—Arash Maboodi, M.D.

Resident of Dermatology
Department of Dermatology
Guilan University of Medical Sciences
Rasht-Iran

—Aptin Maboodi, M.D.

Resident of Internal Medicine
Iran University of Medical Scineces
Tehran-Iran

—Siamak Granmayeh, M.D.

Afrah Pathobiology Center. Rasht-Iran

REFERENCES

1. Ang, P., Tay, Y. K., Ng, S. K., and Seow, C. S. Fatal Lucio's phenomenon in 2 patients with previously undiagnosed leprosy. J. Am. Acad. Dermatol. **48** (2003) 958–961.
2. Bakos, L., Correa, C. C., Bergmann, L., Bonamigo, R. R., and Muller, L. F. B. Antiphospholipid antibodies thrombotic syndrome misdiagnosed as Lucio's phenomenon. Int. J. Lepr. Other Mycobact. Dis. **64** (1996) 320–323.
3. Latapi, F., and Chevez, Z. A. The spotted leprosy of Lucio: an introduction to its clinical and histological study. Int. J. Lepr. **16** (1948) 421–430.
4. Pursley, T. V., Jacobson, R. R., and Apisarnthanarax, P. Lucio's phenomenon. Arch. Dermatol. **116(2)** (1980 Feb) 201–204.
5. Quismorio, F. P., Rea, T. H., Chandor, S., Levan, N., and Friou, G. Lucio's phenomenon: an immune complex deposition syndrome in lepromatous leprosy. Clin. Immunol. Immunopathol. **9** (1978) 184–193.
6. Rea, T. H., and Levan, N. E. Lucio's phenomenon and diffuse non-nodular lepromatous leprosy. Arch. Dermatol. **1114** (1978) 1023–1028.
7. Rea, T. H., and Ridley, D. S. Lucio's phenomenon: a comparative histology study. Int. J. Lepr. Other Mycobact. Dis. **47** (1979) 161–166.
8. Romero, A., Ibarra, A. B., and Fallas, M. Clinical study of lepromatous leprosy in Costa Rica. Mt. J. Lepr. **17** (1949) 27–33.
9. Saoji, V., and Salodkar, A. Lucio leprosy with lucio phenomenon. Indian J. Lepr. **73(3)** (2001) 267–272.
10. Souza, C. S., Roselino, A. M., Figueiredo, F., and Foss, N. T. Lucio's phenomenon: clinical and therapeutic aspects. Int. J. Lepr. Other Mycobact. Dis. **68(4)** (2000) 417–425.