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THE LUCIO-ALVARADO FORM OF LEPROSY¹
A CASE OBSERVED IN BRAZIL

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Rafael Lucio, in collaboration with Ignacio Alvarado, published in 1852 (8) a monograph on leprosy in which they distinguished three clinical forms of the disease, namely: nodular, anesthetic, and "manchada" (spotted) or "lazarine." They paid special attention to the last of these forms, giving a detailed clinical description of it which has not been greatly improved since then.

The characteristic lesions consist of eruptions of red, painful spots of the skin which appear in outbreaks lasting about 15 days. The preferential sites are the extremities, and only in advanced cases are they disseminated. The macules are generally flat, but sometimes they are infiltrated although they never form nodules. The center of the macule gradually turns darker, and finally becomes covered with a small, thin, dry crust which eventually drops off leaving an insignificant scar. On the larger and more inflammatory lesions there tend to appear dark, flaccid blisters. When such a blister ruptures it leaves a deep ulceration with an irregular border surrounded by an inflammatory zone. There may also appear, as a complication, especially on the legs, a secondary pyogenic infection and chronic cellulitis.

Other manifestations which occur to complete the symptomatologic pattern are: rhinitis; alopecia of the eyebrows, the eyelashes, and the body hairs; and frequent complications affecting the digestive and respiratory tracts and the circulatory system which generally occur with increasing severity. The prognosis is therefore poor, and death occurs earlier than in the other forms of the disease.

In spite of the masterly original description of Lucio and Alvarado, this form of leprosy was gradually forgotten, even in Mexico. A contributory cause of that effect was certainly the fact that certain authors [Pardo-Castello and associates (11, 12), and Rodriguez and Wade (13)] applied the name "lazarine leprosy," the synonym of "spotted

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leprosy," to ulcerated tuberculoid leprosy and the various clinical pictures with ulcerations, necrotic areas and even simple blisters.³

The credit for rediscovering the Lucio and Alvarado form of leprosy goes to Latapí who, in a series of articles (4-7), endeavored to place it in modern leprology and to show that it is not only seen in Mexico but also in other countries. Thus in Costa Rica, Romero *et al.* (14) reported that 70 per cent of the cases of leprosy seen there are lepromatous, and that of them 44.8 per cent (31.5% of all cases) are of the Lucio form. At the same time nodular cases are relatively rare, which makes it difficult to obtain material for the preparation of lepromin.

Latapí pointed out that the fundamental anatomic and clinical features of this morphologic type is a diffuse, generalized cutaneous infiltration, especially of the face and hands, which is succulent and myxedematoid at the beginning and atrophic and ichthyosiform later on. Over this diffuse lepromatosis, without nodules, there develop in an episodic manner the red and painful macules whose evolution is described above. These lesions were called by Latapí the "Lucio phenomenon," or "necrotizing erythema," by analogy with erythema multiforme or erythema nodosum, since they represent a variety of lepra reaction produced by a process of necrotizing vascularitis, multiple and acute.

We have, then, a picture of "diffuse lepromatosis, pure and primary, with multiple necrotizing vascularitis." Another noteworthy fact is the aggravation of the symptoms by active therapies such as antisyphilitic treatment, iodides and toxoids, and especially chaulmoogra oil, which seems even to hasten death. On the contrary, sulfone treatment gives excellent results.

Articles by other authors have contributed to completion of the picture of the Lucio and Alvarado leprosy. Escalona (1), and later Gonzalez Urueña (2), described a special type of cephalic alopecia. Martinez Baez (9) studied the histopathology, noting the basic lepromatous structure with acute vascularitis and perivascular lesions leading to necrosis. Gonzalez Chavez (2) reported on the scarcity of bacilli in the "necrotizing erythema" lesions, and their abundance in the nasal mucosa and the diffuse infiltrate. He also found the serologic tests for syphilis almost constantly to give positive, and strong, results

³ First place among these offenders should probably be given to Zambaco-Pacha, whose contributions to the literature had influence several decades ago. In a book entitled "Les Lépreux ambulants de Constantinople," a monograph published in 1897 in Paris, a whole chapter (Chap. VIII, pp. 261-277) was devoted to Lèpre ulcéreuse ou Lazarine. There is no indication in the text that Zambaco was aware of the publication of Lucio and Alvarado. He explained that the condition he described was ". . . ainsi dénommée de ce que le frère de Magdaleine, Lazare, que l'on confond à tort avec le ressuscité, en aurait été atteint. C'est par la même croyance universelle qu'il a été institué patron des lépreux, et que, sous les croisades, un ordre de religieux, invoquant ce tutélaire, a été créé pour soigner les ladres, qualification synonyme de lépreux."—EDITOR.

Medina (¹⁰) observed a special response to the lepromin antigen. There occurs 4-6 hours to 2 or more days after the injection, the reproduction on a small scale of the Lucio phenomenon at the site of injection.

Latapí and Chevez Zamora (⁷) made a complete clinical and histopathologic study of the condition. The microscopic changes are essentially those of lepromatous leprosy, the noteworthy features being the diffuse character of the granulomatous, Virchow-cell infiltrate, without nodule formation, and also the vascular neoformation and thrombosis of the dermal arterioles with consequent necrosis. In the bullous episodes there is a severe edematous process. Attempting to explain the Medina reaction, these authors suggested a mechanism of bacterial hypersensitivity of the Shwartzman-Sanarelli type. This would not, then, be a specific allergic reaction but a phenomenon apparently precipitated by a synergic bacterial mechanism dependent upon an association of the Hansen bacillus with pyogenic cocci. The histology differs from what is seen in the Fernandez reaction, there being a predominance of polymorphonuclear leucocytes and a relative abundance of eosinophils in the cellular infiltrate, with edema and necrosis of the collagen, the process terminating with formation of an intradermal abscess.

Latapí and Chevez Zamora have reported that the frequency of the Lucio form of leprosy is 15 to 20 per cent in the clinical material in Mexico City, and they suggested that its prevalence in other countries is probably not much lower. Actually, when Latapí visited Belo Horizonte in 1958, on the occasion of the leprosy seminar sponsored by WHO, he identified with us a case of that clinical form in a patient of the Santa Isabel Colony. That patient is the subject of this report, the first case reported from Brazil.

CLINICAL OBSERVATIONS

B. R., female, aged 16 years, white, domestic, native of Alto Garça, Mato Grosso, Brazil, was admitted to the Santa Isabel Colony July 30, 1953. She was barely 11 years old at the time, and with very little education, so no information could be obtained about the onset and course of the disease. She spoke of frequent episodes of epistaxis. One of her seven brothers is also interned in the Santa Isabel Colony. The following notes are of findings at the time of writing, more than five years after the patient was admitted.

Dermatologic examination.—Face and earlobes show erythema and diffuse infiltration, and there is subtotal superficial alopecia (Fig. 1). The skin of the forearms is

DESCRIPTION OF PLATE

FIG. 1. Face of patient with pure diffuse lepromatous leprosy. Note absence of nodule formation. Alopecia of eyebrows and eyelashes.

FIG. 2. Reproduction of the Lucio phenomenon seen 6 hours after the injection of lepromin (reaction of Medina). The Fernandez and Mitsuda reactions were negative.

FIGS. 3. and 4. Violaceous and purpuric macules with a reticular pattern, presenting livid, racemose, small ulcerations due to necrotizing vasculitis (Lucio phenomenon, or "necrotizing erythema"). Diffuse lepromatous infiltration without formation of nodules.



shiny, with at the elbows crusted ulcerous lesions and cicatricial residuae of old eruptions. The skin of the lower extremities is infiltrated, edematous, of the "glossy skin" type, showing violaceous and purpuric vascular macules disposed in an edident reticular pattern, resembling livedo racemosa (Figs. 3 and 4). In several places there are small, irregular ulcerations, with or without hemorrhagic crusts, as well as scars of healed lesions. The rest of the clinical examination reveals nothing noteworthy.

Complementary examinations.—Acid-fast bacilli: nasal mucosa, 2+; earlobes, 4+; lesions of the left leg and left arm, negative. Mitsuda reaction: negative. Chest fluorography: normal.

Treatment and progress.—In the period from March 16, 1954 to February 11, 1955, sulfone therapy was given: 50 ampules of Promin and 50 tablets of Diasone. No other data were found in her record, and she stated that she had received very little treatment in the last four years. In the meantime she continued to have periodical outbreaks of "necrotizing erythema."

The Mitsuda test was made on the flexor surface of the forearm, and 6 hours later there was reproduced at the site of the injection one of the many lesions exhibited by the patient (Fig. 2). This lesion persisted, but there was complete absence of infiltration; in other words, the Mitsuda reaction was negative. Also noteworthy is the negative bacteriology of the cutaneous lesions, a fact previously reported by Gonzalez Chavez.

SUMMARY

A case of "pure and primary diffuse lepromatosis with multiple necrotizing vascularitis" (Lucio and Alvarado leprosy) is reported, the first to be recorded in Brazil. The outstanding reports on the subject are reviewed with respect to the most important morphologic and histopathologic aspects of this variety of the lepromatous type.

RESUMEN

Se presenta un caso de "lepromatosis difusa pura y primaria con vascularitis necrotizante múltiple" (lepra de Lucio y Alvarado), el primero que se haya comunicado en el Brasil. Se repasan las reseñas más destacadas sobre la materia, con respecto a las más importantes fases morfológicas e histopatológicas de esta variedad de la forma lepromatosa.

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