DISSEMINATED CUTANEOUS LEISHMANIASIS DUE TO LEISHMANIA GUYANENSIS: CASE OF A PATIENT WITH 425 LESIONS

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Abstract. Disseminated cutaneous leishmaniasis is characterized by the presence of a large (≥10) number of lesions at several anatomic sites (head, limbs, and trunk). Most of the lesions are small, papular, and appear simultaneously with or secondarily to one or several ulcerated lesions of localized cutaneous leishmaniasis. We report the first case of disseminated cutaneous leishmaniasis in French Guiana. It concerns a 24-year-old woman who tested negative for human immunodeficiency virus (HIV). The disease began with three lesions that became ulcerated. One week later, multiple papulo-nodular lesions appeared. We counted a total of 425 lesions. Leishmania were observed in the lesions. The species involved was L. guyanensis, which has never been described in a case of disseminated cutaneous leishmaniasis. The patient was rapidly cured by a single course of pentamidine. Disseminated cutaneous leishmaniasis should be distinguished from other types of leishmaniasis with multiple lesions. These include anergic diffuse cutaneous leishmaniasis, post–kala-azar leishmaniasis, and leishmaniasis associated with HIV infection.

INTRODUCTION

Certain forms of cutaneous leishmaniasis result in large numbers of lesions. This is the case for anergic diffuse cutaneous leishmaniasis, post–kala-azar cutaneous leishmaniasis, and cutaneous leishmaniasis arising in a context of immunodeficiency, such as infection with human immunodeficiency virus (HIV).

Brazilian scientists have identified another form of cutaneous leishmaniasis, which was named disseminated cutaneous leishmaniasis (DCL).1–3 This form can be differentiated from classic localized cutaneous leishmaniasis (LCL) by the large (≥10) number of lesions, the clinical type of the major elementary lesions (papular and or acneiform), and the weaker response to classic treatments. These non-ulcerated secondary lesions are thought to be formed following the dissemination of the protozoan in the lymph and blood from the initial lesions. The species involved are Leishmania braziliensis and L. amazonensis.2

In French Guiana, LCL is particularly frequent, with an annual incidence of approximately 0.2%.4 Leishmania guyanensis is responsible for most of the cases.4 We present a patient who developed a form of cutaneous leishmaniasis due to L. guyanensis that met the criteria for DCL.

CASE REPORT

A 24-year-old woman with no particular medical history was hospitalized in the Dermatology Department of the Centre Hospitalier de Cayenne in February 1997. She lived in Cayenne, but for two years, she had regularly accompanied her husband into a forested area to help him to develop a small farm, which involved cutting down trees.

Upon questioning, the patient described the appearance in December 1996 of three cutaneous lesions on her right hand, left breast, and back. About a week later, a profusion of lesions appeared, covering almost her entire body. These lesions gradually increased in size, while the initial lesions became ulcerated. She consulted a general practitioner in January 1997 who prescribed oxacilline. However, she did not respond to this treatment and she was then referred to a dermatologist who admitted her to the Dermatology Department of the Centre Hospitalier de Cayenne.

Clinical examination on admission showed the patient to be in good general condition without fever. She presented cutaneous lesions, most of which were papulo-nodular, infiltrated, and slightly squamous. Her entire body was affected, with the exception of the scalp, palms of the hands, and soles of the feet. We counted a total of 425 lesions. The largest lesions, located on her back (Figure 1), on the middle finger of her right hand, and on her left breast (Figure 2), were ulcerated and had scabs. Palpation revealed signs of painless, non-inflamatory lymphangitis around certain lesions, and left inguinal adenopathy. The mucous membranes, particularly those within the nose, were free of lesions.

Smears were performed from scrapings of an ulceration on the left breast, of a nodule on the forehead, and of a papulous lesion of the right thigh. Direct examination of skin smears stained with May-Grünwald-Giemsa dye showed the presence of numerous Leishmania amastigotes, either scattered or within macrophages, in all three smears. Histologic examination showed a diffuse non-granulomatous inflammation of the dermis containing mostly lymphocytes, histiocytes, and plasma cells, and many intrahistiocytic forms of leishmaniasis, primarily concerning the subpapillary venular plexus. A strain of Leishmania was isolated from cultures. Isoenzyme analysis showed that this strain was L. guyanensis.

The C-reactive protein concentration was 84 mg/L (normal < 5 mg/L). The blood eosinophil count was 680/mm³ and CD4 and CD8 lymphocyte counts were normal. The patient tested negative for HIV-1. A myelogram showed no Leishmania.

The patient was treated with two deep intramuscular injections of pentamidine isethionate (Pentacarinat®; Aventis, Paris, France), 48 hours apart, with a dose of 4 mg/kg of pentamidine base used for each injection. One month after treatment, all lesions had healed. A checkup in June 2002 (five years later) showed no recurrence.

The patient’s husband was also examined in February 1997. This 34-year-old man was found to have an isolated ulceronecrotic lesion of the neck. Smears confirmed a diagnosis of localized cutaneous leishmaniasis (presence of Leishmania). Given the uncomplicated presentation, culture and biopsy were not performed. Clinical cure was obtained in one month with the same pentamidine treatment as that given to his wife.
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FIGURE 1. Multiple papular lesions and an ulceration on the back of the patient.

FIGURE 2. Ulceration of a finger and the left breast (initial lesions) of the patient.

DISCUSSION

Disseminated cutaneous leishmaniasis has been described in the northern and northeastern regions of Brazil.1–3 In these regions, DCL has been reported in 0.2–1.9% of cutaneous leishmaniasis cases, depending on the series.2 The dissemination of lesions from initial lesions is observed three days to eight weeks after the appearance of the initial lesions.2–3 The number of lesions is generally between 10 and 800.2–3 Treatment with N-methyl-glucamine antimoniate (Glucantime®; Aventis) cures leishmaniasis within a year in 40% of patients with DCL.2,3 Our case had the characteristics of DCL: multiples lesions, mostly papulous, full expression of the disease within a few weeks, presence of ulcerations at the beginning of the disease, and a good therapeutic response. One course of pentamidine isethionate (Pentacarinate®), which is an effective treatment for LCL in French Guiana,5 cured our patient with no recurrence after five years of follow-up. The only discordant feature was the absence of granuloma on histopathologic examination, but granuloma is not always present in lesions of DCL.2

The differential diagnoses we made involved the three other forms of leishmaniasis in which a large number of lesions occur. The first was anergic DCL. This rare form of cutaneous leishmaniasis is secondary to an underlying deficit in cellular immunity specific for certain species of Leishmania.6 In South America, this form is caused by L. amazonensis. Multiple papulo-nodular non-ulcerated lesions appear progressively over several years of evolution. This clinical form is refractory to treatment. The second was post–kala-azar dermal leishmaniasis. This form is encountered principally in India.7 The cutaneous lesions accompany or follow visceral leishmaniasis caused by L. donovani. The third was cutaneous leishmaniasis associated with immunodeficiency, such as HIV infection. An increasing number of cases of cutaneous leishmaniasis with profuse lesions, and sometimes with visceral involvement, are being reported, particularly in patients with HIV infections.8–10

In our patient, the diagnosis of DCL can be rejected by several arguments: the appearance of the lesions was far too rapid for DCL; the isolated species was not L. amazonensis, the only species known to cause DCL in French Guiana; and treatment led to complete cure without recurrence in the five years of follow up. Similarly, post–kala-azar was rejected on epidemiologic and clinical grounds (no L. donovani in French Guiana and no visceral leishmaniasis in our patient). Finally, in the absence of clinical or biologic signs of immunodepression and given the HIV-negative status, the diagnosis of leishmaniasis associated with immunodeficiency was rejected.

The dissociated clinical presentation between our patient and her husband suggests that most lesions were caused by hemolymphatic dissemination from the initial lesions. Indeed, the multiple lesions of the patient are probably not due to hundreds of phlebotomine bites because if that was the case the husband would also be expected to have numerous lesions. In addition, upon questioning the patient did not recall multiple painful insect bites.

A number of arguments suggest that dissemination of the lesions in DCL is determined by the immunogenetic background of the patient rather than the specific virulence of the parasite itself; the absence of clustered cases of DCL, the fact that our patient’s husband after probable simultaneous contamination by the same parasite strain only developed LCL, and the description of a new species implicated in DCL (L. guyanensis) in addition to L. braziliensis and L. amazonensis.

In conclusion, this is the first case of DCL to be reported in French Guiana. The species isolated from our patient was L. guyanensis. This observation shows that this species can also cause the disseminated form of the disease, as has been shown for L. amazonensis and L. braziliensis.2

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REFERENCES


