DISCUSSION

In this patient, inflammatory processes associated with fibrosis from papillary dermis with the presence of Leishmania was observed in cutaneous lesions. In macular lesions of PKDL, chronic inflammation is characterized by infiltrates of histiocytes, lymphocytes, and a few plasma cells, and Leishmania is also observed. However, there are no consistent infiltrates in PKDL lesions. The clinical manifestations of PKDL are immunologically mediated with features of a Th2 response in the skin and a systemic Th1 response, resulting in skin abnormalities in patients who are otherwise well without features of systemic leishmanial infection. In this patient, the clinical progression from visceral disease with subsequent relapses to para-kala-azar dermal leishmaniasis and later isolated (post-kala-azar) lymphadenopathy without obvious clinical evidence of visceral disease suggests a similarly developing, but abnormal and inadequate systemic immune response. In immunosuppressed patients, in particular those who are HIV infected, relapses are common, as some degree of developing antileishmanial immunity is needed to prevent a relapse of VL. With each relapse, the treatment becomes more difficult. This patient had a similar clinical syndrome, which justified the decision for maintenance treatment that to date seemed to be successful. There is no consensus regarding the preferred regimen; single-dose administration of pentavalent antimonial, Lamb (AmBisome ©) or pentamidine has been used, often in cycles of 3–4 weeks.

This case illustrates the need for follow-up of patients with VL, in particular to monitor for PKDL or para-kala-azar dermal leishmaniasis as experience elsewhere indicates that as the rash often remains unnoticed and self-cures, these patients often do not report to the clinic and may play a role in transmission.

DISCUSSION

This case illustrates some of the more uncommon issues that may arise in VL in Brazil and that have not been well described. Despite adequate treatment with a full course of pentavalent antimonial, this patient relapsed 3 times; during the third episode, he presented with PKDL confirmed by skin biopsy, while at the same time, he presented with systemic infection with demonstrable parasites in a bone marrow aspirate. This should therefore be called para-kala-azar dermal leishmaniasis that has not yet been described in VL in South America where VL is caused by Leishmania infantum. While in VL there is a predominantly Th2 response with absent immune response against Leishmania, after successful treatment Th1 responses develop that indicate cure and immunity. In this patient, inflammation and fibrosis was found in the papillary dermis with the presence of Leishmania parasites. As in macular lesions, predominantly lymphocytes and macrophages may be found; plasma cells are often scanty, but there is no consistent pattern. Leishmania parasites may be difficult to demonstrate in macular lesions.

The clinical manifestations of PKDL are immunologically mediated with features of a Th2 response in the skin and a systemic Th1 response, resulting in the skin abnormalities in a patient who is otherwise well without features of systemic leishmanial infection. In this patient, the clinical progression from visceral disease with subsequent relapses to para-kala-azar dermal leishmaniasis and later isolated (post-kala-azar) lymphadenopathy without obvious clinical evidence of visceral disease suggests a similar developing but abnormal and inadequate immune systemic response.

In immunosuppressed patients, in particular those who are HIV infected, relapses are common, as some degree of developing antileishmanial immunity is needed to prevent a relapse of VL. With each relapse, the treatment becomes more difficult. This patient had a similar clinical syndrome, which justified the decision for maintenance treatment that to date seemed to be successful. There is no consensus regarding the preferred regimen; single-dose administration of pentavalent antimonial, Lamb (AmBisome ©) or pentamidine has been used, often in cycles of 3–4 weeks.