Bacillary Angiomatosis in Immunocompetent Patient with Atypical Manifestations

Fariba Iraji, Mohsen Pourazizi, Bahareh Abtahi-Naeini, Mohsen Meidani, and Parvin Rajabi

Abstract

Bacillary angiomatosis is an infectious disease caused by two Gram-negative bacilli; this disease usually affects immunosuppressed hosts with a history of cat scratch. We report a rare case of bacillary angiomatosis in an immunocompetent 26-year-old woman with no history of exposure to cats, and with atypical clinical features (very pruritic vascular papules and nodules with ulceration and hemorrhage on the right arm and fingers). She was successfully treated with clarithromycin for 3 months. Bacillary angiomatosis must be kept in mind in the differential diagnosis of any papules and nodules in cases of unknown etiology and also in immunocompetent patients and HIV-negative individual.

Keywords: Bacillary angiomatosis, bartonella, clarithromycin, immunocompetent

What was known?

Bacillary angiomatosis is an infectious disease caused by two Gram negative bacilli. This disease usually affects immunosuppressed hosts.

Introduction

Bacillary angiomatosis (BA), an uncommon disease, was first described in 1983 as a disseminated infection in patients immunosuppressed by HIV infection.[1] It is also rarely encountered in patients with other immunocompromising conditions and even in immunocompetent people.[2] We report a rare case of BA in an immunocompetent 26-year-old woman with no history of exposure to cats, with atypical clinical features (very pruritic vascular papules and nodules with ulceration and hemorrhage).

Case Report
A 26-year-old healthy woman had developed papules and nodules on the right arm 6 months ago. Despite treatment with short course systemic antibiotics the lesions had enlarged and gradually extended to the forearm and hand and also to her fingers. She was referred to us for the evaluation of the eruption. The patient was not febrile and denied having been scratched by any cat. She had had no previous illness nor received any immunosuppressive drugs. On physical examination, multiple tender erythematous angiomatous papules and nodules in a grouped pattern were seen on the extensor aspect of the right arm that extended to the distal part in varying sizes of 1 to 2 cm in diameter [Figure 1]. The remainder of her skin was essentially normal, with no signs of a blistering or erythematous background. There was revealed painful axillary lymphadenopathy on general examination. There was no sign of a mucous membrane lesion or hepatosplenomegaly. It became evident that she also suffers from severe pruritus and occasionally bleeding in the papules. Routine laboratory blood tests, CD4 count were normal. ELISA tests for HIV were negative. No evidence of other immunosuppressive status was revealed by various systemic examinations. Histopathological examination showed an epidermal collaret with mild hyperkeratosis with focal parakeratosis and crust formation. In the dermis, proliferation of blood vessels and inflammatory cells were seen. The vascular component in the upper dermis was constituted by thin-walled vessels lined by plump, and deeper are small and thick-walled vessels [Figure 2]. The inflammatory cells were composed of lymphocytes and some eosinophils and neutrophils. In some areas eosinophilic granular material was present. The silver stain revealed a small number of bacilli like organisms [Figure 3]; we diagnosed the patient's eruption as BA. A cutaneous lesion did not respond within 4 weeks of adequate oral erythromycin, so the patient was treated with oral clarithromycin 500 mg twice daily (second-line of treatment) and in order to control the sever with pruritus doxepin 25 mg once daily which resulted in significant improvement over a 3 month treatment period [Figure 4]. The patient is now, 3 months after treatment, living her normal life without experiencing any problems or showing any signs of the recurrence of the disease.

![Figure 1](bacillary_angiomatosis.jpg)

**Figure 1**
Bacillary angiomatosis. Before treatment: Clinical appearance of the bacillary angiomatosis: Angiomatous papule and nodules (a) arm and (b) fingers

![Figure 2](bacillary_angiomatosish.jpg)

**Figure 2**
Bacillary angiomatosis. The vascular component in upper dermis constituted by thin-walled vessels lined by hobnail endothelial cell (vascular proliferation Hematoxylin and eosin stain H and E, ×400)
Discussion

BA is a skin infection in patients immunosuppressed by HIV infection. It is also rarely encountered in patients with other immunocompromising conditions and even in immunocompetent people.[2] The skin lesions are reddish or brown papules usually in large numbers that resemble Kaposi's sarcoma. There are only a few case reports about BA in immunocompetent individuals.[3,4,5,6,7] Our patient in this report was immunocompetent without any history of recurrent infections or usage of immunosuppressive drugs, leukemia or lymphoma. One third of the patients with BA have no known exposure to cats, suggesting that BA may also be acquired by other ways besides direct contact with these animals.[8] Only 20% of the patients with BA due to H. henselae report a preceding cat scratch or bite, compared to the 90% of those with cat-scratch disease.[9,10] Because BA can have extensive visceral involvement, even if only cutaneous BA is evident, visceral disease should be suspected.[11] Therefore, it is necessary that in the case of each patient with angiomatosis lesions, BA also be considered in the differential diagnosis in order to prevent the development of potentially dangerous complications. Warthin-Starry staining reveals these deposits to be dense masses of short bacilli; the method also exposes argyrophilic bacilli. Clinical and histopathological findings in our patient were sufficient for the diagnosis of BA. Routinely, diagnosis has been based on the viewing of the bacilli in histological sections stained with silver dyes or through the Warthin-Starry method, as in our case. The major differential diagnoses are Kaposi’s sarcoma, multiple pyogenic granuloma, multiple cherry angioma, angiookeratoma, and epithelioid hemangioma.[3,4]

Immunohistochemical demonstration of HHV-8 is of great help in the histologic differential diagnosis of Kaposi’s sarcoma. A useful diagnostic tool is the demonstration of HHV-8 in samples of patients that is negative in BA. Until recently, revealing the virus was only possible
by in situ hybridization. This has been simplified by the development of a commercially available monoclonal antibody against the latent nuclear antigen (LNA-l) of the virus that can be used in routine practice,[12] the result of which was negative in the case of our patient. The most efficient treatment consists of erythromycin or doxycycline, for 8 to 16 weeks. However, the treatment of choice is oral erythromycin. Response to this treatment varies from case to case. A second option consists of clarithromycin or azithromycin. Response to this treatment varies from case to case. Some cases respond to treatment rapidly, within several days,[5] Others require a prolonged treatment, maybe even for several months.[3,4,6] Our patient was treated with oral clarithromycin 500 mg twice daily (because not response to erythromycin), and doxepin to control the severe pruritus; this resulted in significant improvement in a 3 month treatment period without any recurrence. In conclusion, although, the most common risk factor for development of BA is HIV infection but our case such as rare case reports demonstrates that BA must be kept in mind in the differential diagnosis of any papules and nodules in cases of unknown etiology, also in immunocompetent patients and HIV-negative individual. BA should be kept in mind in the differential diagnosis of any angiomatous papules in cases of unknown etiology.

What is new?

We report a very rare case of bacillary angiomatosis in an immunocompetent 26-year-old woman with no history of exposure to cats, and with atypical clinical features (very pruritic vascular papules and nodules with ulceration and hemorrhage on the right arm and fingers).

Go to:

Footnotes

Source of support: Nil

Conflict of Interest: Nil.

Go to:

References